Imaging of intestinal vasculitis focusing on MR and CT enterography: a two-way street between radiologic findings and clinical data

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Abstract
Diagnosis of intestinal vasculitis is often challenging due to the non-specific clinical and imaging findings. Vasculitides with gastrointestinal (GI) manifestations are rare, but their diagnosis holds immense significance as late or missed recognition can result in high mortality rates. Given the resemblance of radiologic findings with some other entities, GI vasculitis is often overlooked on small bowel studies done using computed tomography/magnetic resonance enterography (CTE/MRE). Hereon, we reviewed radiologic findings of vasculitis with gastrointestinal involvement on CTE and MRE. The variety of findings on MRE/CTE depend upon the size of the involved vessels. Signs of intestinal ischemia, e.g., mural thickening, submucosal edema, mural hyperenhancement, and restricted diffusion on diffusion-weighted imaging, are common in intestinal vasculitis. Involvement of the abdominal aorta and the major visceral arteries is presented as concentric mural thickening, transmural calcification, luminal stenosis, occlusion, aneurysmal changes, and collateral vessels. Such findings can be observed particularly in large- and medium-vessel vasculitis. The presence of extra-intestinal findings, including within the liver, kidneys, or spleen in the form of focal areas of infarction or heterogeneous enhancement due to microvascular involvement, can be another radiologic clue in diagnosis of vasculitis. The link between the clinical/laboratory findings and MRE/CTE abnormalities needs to be corresponded when it comes to the diagnosis of intestinal vasculitis.

Keywords: Magnetic resonance enterography, Computed tomography enterography, Vasculitis, Intestines

Key points
• While gastrointestinal presentations are uncommon in vasculitis, their timely diagnosis is critical.
• Patients may present with a wide variety of non-specific findings on MRE/CTE.
• Extraintestinal findings are useful that should be covered in the MRE/CTE field.
• Imaging features need to be linked with the clinical and laboratory data.

Background
Vasculitis is a term used for a heterogeneous group of conditions resulting in inflammation and injury of blood vessels in a variety of organs [1]. While this condition is rare, it can potentially have fatal outcomes due to resulting necrosis and ischemia. Therefore, there would be a need for early diagnosis and therapeutic interventions. Due to the non-specific clinical and imaging findings, diagnosis of vasculitis is often challenging. This is due to its similarities with other conditions, such as infections,
malignancies, connective tissue diseases, and thromboembolic disorders [2–4]. A clinical history of systemic diseases, along with constitutional symptoms, including fever, weight loss, and fatigue, and most importantly, a dramatic response to immunosuppressive therapy could suggest a potential diagnosis of vasculitis.

A wide variety of vasculitides can present with abdominal manifestations, which could be the first presentation of a systemic disease [4–6]. While vasculitis of the gastrointestinal (GI) tract is rare, its diagnosis holds immense significance as late or missed recognition can result in high morbidity and mortality rates [5]. Abdominal pain, nausea, vomiting, diarrhea, GI bleeding (commonly presented as occult blood in stool, hematochezia, melena, and hematemesis), and weight loss are the most common GI presentations. Rarely, patients may develop serious complications, including intestinal perforation, ischemia, or peritonitis [7]. The wide spectrum of clinical and imaging findings in vasculitis, which may be non-specific and overlapping with other conditions, raises a diagnostic challenge.

Patients with vasculitis of the GI tract may undergo computed tomography/magnetic resonance enterography (CTE/MRE) to assess intestinal pathologies responsible for the non-specific clinical manifestations [7]. In addition to intestinal findings, CTE and MRE can often detect extra-intestinal pathologies [8–10]. The most common radiographic features in the GI tract, seen in different types and various stages of vasculitis, are submucosal edema or hemorrhage, mesenteric edema, bowel dilatation, and findings of acute or chronic mesenteric ischemia. Eventually, the ischemia may result in bowel perforation or stricture [4, 11].

Given the resemblance of radiologic findings with other entities such as infection, inflammatory bowel disease (IBD), and mesenteric ischemia secondary to vascular thromboembolism, diagnosis of vasculitis is often overlooked on MRE or CTE [12]. It is a diagnostic challenge and could not be simply differentiated from other disorders based on imaging findings alone. Therefore, a multidisciplinary team approach and histopathologic analysis are often required to make a definitive diagnosis. Radiologists must be aware of the diverse imaging patterns in intestinal vasculitis since they might be the first caregivers suggesting the proper diagnosis preventing misdiagnosis [12]. To the best of our knowledge, there is no comprehensive review describing the imaging characteristics of intestinal vasculitis on MRE/CTE.

Herein, we present radiologic findings of GI vasculitis with a focus on CTE and MRE by representative clinical cases. We highlight the diagnostic clues in clinical features and laboratory findings, helping in establishing the diagnosis. Lastly, a practical algorithm is provided to assist in the diagnosis of intestinal vasculitis and to differentiate it from other common GI disorders.

Classification of vasculitis: an overview

Vasculitides are classified as (1) primary (idiopathic), with most likely autoimmune causes, and (2) secondary, which can stem from malignancy, infection, connective tissue diseases, drugs, or environmental exposures. The revised Chapel Hill Consensus Conference nomenclature system classifies vasculitides primarily according to the predominant type of vessel involvement and etiology. The classification is into seven categories: large-, medium-, small-, and variable-vessel vasculitis, single-organ vasculitis, vasculitis associated with systemic disease, and vasculitis associated with probable etiology (Fig. 1) [13]. Clinical presentation and imaging features are typically related to the size and location of involved vessels and the extent of the disease.

While the exact pathophysiology of vasculitis is not elucidated, the inflammation caused by infiltration of immune cells (lymphocytes, neutrophils, and monocytes/macrophages) in different layers of the vessel walls, release of pro-inflammatory cytokines, deposition of immune complexes, or presence of auto-antibodies can trigger vascular injury. Subsequent to the inflammation, conditions such as stenosis, occlusion, and rarely aneurysms can develop within the vessel (Fig. 2). Aneurysms commonly result from the destruction of the media layer and are associated with a risk of spontaneous hemorrhage and mural thrombosis. Obliteration or stenosis of the arterial lumen leads to decreased perfusion in abdominal organs, and in case vasa vasorum is involved, it can result in intestinal ischemia. Furthermore, diffuse intimal calcification may occur as a result of chronic vascular inflammation [14].

Technical considerations

The 2017 consensus statement of joint European Society of Gastrointestinal and Abdominal Radiology (ESGAR) and European Society of Pediatric Radiology (ESPR) describes routine technical considerations for cross-sectional small bowel and colonic imaging [15]. Three main additional technical points should be incorporated into the routine MRE and CTE protocols when intestinal vasculitis is suspected:

(a) Arterial phase should be added to the routine postcontrast portal or enteric phase, which enhances visualization of arterial structures and vasculitis-associated intestinal ischemia [16, 17];

(b) As concurrent involvement of solid abdominal organs may be the only key to distinguish intestinal vasculitis from other entities, it is important to expand the field of scan as far as possible to cover extra-intestinal organs while avoiding a drop in image quality.
Large-vessel vasculitides predominantly involve the great vessels, such as the aorta and its major branches. Takayasu arteritis and giant cell arteritis (GCA) are the most prevalent vasculitis in this subgroup. Considering the lack of specific diagnostic biomarkers, imaging modalities, including Doppler ultrasound, CT or MR angiography, digital subtraction angiography, and 18F-fluorodeoxyglucose positron emission tomography (PET), play a crucial role in diagnosing large-vessel vasculitis [1, 18, 19]. GI involvement is far more common in Takayasu arteritis than GCA. Radiologic features may be similar to mesenteric ischemia caused by thromboembolism or non-occlusive causes [20, 21]. The other differential diagnoses include atherosclerosis, connective tissue disorders, such as Marfan syndrome, Ehlers–Danlos syndrome, and Loeys–Dietz syndrome [12, 22].

**Takayasu arteritis**

Takayasu arteritis is an autoimmune-mediated granulomatous inflammation of the aorta, its major branches, and pulmonary arteries, most commonly affecting young Asian women [23]. Lesions are typically close to the origin of the primary aortic branch [24]. At the disease onset or during acute phases, the presentations can be non-specific, including malaise, fever, weight loss, anorexia, myalgia, or arthralgias. Abdominal pain was observed in approximately 16% of patients [25]. However, more characteristic signs and symptoms develop with disease progression, including reduced or absent peripheral pulses, arterial bruits, limb claudication, blood pressure discrepancies between the arms due to stenotic or occlusive lesions, and hypertension [23].

GI involvement is rare in Takayasu arteritis. The intestine, spleen, and, rarely, the liver may undergo ischemic changes due to stenosis or occlusion of large- and medium-size GI arteries [6, 25]. Notably, in a cohort of 79 patients with Takayasu, only one patient developed mesenteric ischemia [25]. Approximately one-third of patients have involvement of the abdominal aorta or mesenteric circulation [26]. Kermani et al., in a cohort of 125 patients with Takayasu, found involvement of the abdominal aorta, mesenteric artery, and renal arteries in imaging of approximately 38%, 35%, and 20% of the patients, respectively [27]. The vascular involvement included arterial stenosis, thrombosis, and, rarely,
aneurysms. Aortic dissection is very rare, and less than ten cases have been reported involving the abdominal aorta [28]. Notably, several studies have reported a higher prevalence of IBD among patients with Takayasu arteritis with evidence of genetic overlap [29, 30].

The intestinal findings on the CTE/MRE may include segmental intestinal circumferential mural thickening, abnormal wall enhancement, and submucosal edema. The edema and enhancement reduce dramatically following corticosteroid treatment. Ischemic changes caused by vasculitis can be manifested by a diffuse long segment GI involvement [25]. Bowel dilation and mesenteric vascular engorgement are among other GI findings [12]. Extra-intestinal findings may include concentric mural thickening, transmural calcification, luminal stenosis, occlusion, aneurysmal changes, and collateral vessels formation in the abdominal aorta and its major branches, such as the celiac and superior mesenteric arteries. These vascular pathologies can also be seen on other cross-sectional imaging modalities. On contrast-enhanced CT, arterial mural enhancement can be found during the arterial phase, which is intensified during the late phase. However, since the measurement of mural enhancement may be hindered by the overshining intravascular contrast material, the black blood MRI technique is recommended for early detection of vessel wall inflammation [24]. Following contrast administration, the intima is poorly enhanced while the media-adventitia layer is intensely enhanced, resulting in a “double-ring” enhancement pattern (Fig. 3) [24, 31–33]. The possibility of underlying Takayasu arteritis should be considered when mural thickening or irregularity of the aorta and main branches is seen in younger age patients. Intestinal involvement in this disease may only cause a non-specific bowel wall thickening. Rapid response of bowel abnormality to steroid treatment which can be confirmed in follow-up imaging is a potential diagnostic clue.

**Giant cell arteritis**

Giant cell arteritis (GCA) is a granulomatous vasculitis with a higher prevalence among women older than 50
and patients of northern European descent. GCA has a higher tendency to involve carotid, vertebral, and temporal arteries [34]. The disease manifestations include new-onset headache, jaw claudication, visual loss, constitutional symptoms, anorexia, and polymyalgia.

Abdominal involvement is extremely rare in GCA, especially when compared with Takayasu arteritis [35]. Kermani et al. reported mesenteric artery, abdominal aorta, and renal artery lesions in 17%, 6%, and 10% of patients, respectively [27]. So far, fewer than 15 cases of mesenteric ischemia have been reported [6], some with obstruction and infarction of the small intestine [36] or infarction of the sigmoid colon [37]. The abdominal aorta was involved in nearly half of GCA patients with aortitis, with approximately one-third presenting with abdominal pain [38]. Nevertheless, a recent longitudinal study found that abdominal aorta dilation in GCA was comparable to controls [39]. Acute abdominal pain in patients with GCA, which usually develops 6–7 years after disease onset, might also be an alarming sign for aneurysm or dissection [6, 40]. Strikingly, we did not identify any investigation on MRE or CTE findings of GCA in the literature.

**Medium-vessel vasculitis**

Medium-sized vessels, mainly splanchnic arteries and their branches, are predominantly involved in medium-vessel vasculitis. The onset of vasculitis in medium vessels is more acute and necrotizing than in large-vessel [1]. This group includes polyarteritis nodosa and Kawasaki disease. GI involvement is more common in polyarteritis nodosa, with up to 95% of patients presenting with abdominal pain [1]. Radiologic findings include discovery of microaneurysms, arterial stenosis/occlusion, signs of intestinal ischemia, bleeding or perforation, rupture of hepatic, splenic, and/or renal (micro-) aneurysms, and rarely segmental hepatic or splenic infarction. Conventional angiography is the most reliable imaging technique utilized to investigate splanchnic vascular abnormalities [41, 42].

**Polyarteritis nodosa**

Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis affecting medium and small vessels like in the kidneys without glomerulonephritis. It can occur in patients of any age, gender, or ethnicity; however, the peak incidence is at the 5th–6th decade of life. It involves the kidneys, skin, musculoskeletal system, nerves, and GI
system [43]. The inflammation commonly affects all layers of vessels, resulting in stenosis, occlusion, or aneurysm. Therefore, tissue ischemia or hemorrhage may be observed in various organs, leading to acute life-threatening complications. Theoretically, histological confirmation of segmental fibrinoid necrosis of medium-sized vessels is required for the diagnosis. Nevertheless, the diagnosis can also be established based on clinical, laboratory (absence of antineutrophil cytoplasmic antibody (ANCA)), and radiologic findings. Angiography is the preferred imaging modality for detecting microaneurysm formations in visceral arteries [1, 43, 44].

PAN can cause heterogeneous GI manifestations, with non-specific abdominal pain occurring in 35–95% of the patients [4, 6, 11]. Other GI presentations are nausea, vomiting, diarrhea, gastroduodenal ulcers (mainly in the jejunum), hematochezia, hematemesis, and melena. Mesenteric vasculitis is observed in 50–60% of patients, which can lead to GI ischemia [26] or (rarely) aneurysm formation [45]. Serious abdominal complications include GI bleeding caused by ischemic mucosal ulcerations, bowel infarct or perforation, as well as perforation of microaneurysms, and organ ischemia due to visceral artery involvement [6, 46]. The severe manifestations rarely develop (<5%) and can be a predictor of high mortality [11, 46]. Intestinal ischemia has even been reported as the initial presentation of the systemic disease [47]. PAN is the most common systemic vasculitis associated with gallbladder disease, with nearly 4% of patients developing cholecystitis [11]. While hepatic and splenic complications are rare, the patients may experience occlusion of hepatic veins (Budd–Chiari syndrome) and segmental liver or spleen ischemia [6, 46]. The intestinal findings on CTE/MRE include segmental bowel mural thickening, submucosal edema, and abnormal hyperenhancement with striated pattern (Fig. 4). The involved intestinal region may also show diffusion restriction on DWI sequence. Luminal stenosis, mural irregularity and thickening, and aneurysmal dilatation can be detected in visceral arteries, especially in the superior mesenteric artery (SMA) branches (Fig. 5). Visceral infarction and ruptured aneurysm may also be observed on contrast-enhanced CT [26, 48]. While microaneurysms (of renal, mesenteric, and/or splenic arteries) are the hallmark of PAN, they are not often directly detectable on enterography [49]. However, heterogeneous liver enhancement can indicate microvascular abnormalities (Fig. 4). Besides, the addition of an arterial phase in the imaging protocol may improve the detection of the microaneurysms. Since the kidney is the most commonly affected organ, its end-organ damage can present as striated nephrogram indicating renal infarct [49, 50]. Hydronephrosis resulting from detrusor muscle spasm is another urinary system complication [4, 51].

Differential diagnoses of splanchnic vascular abnormalities in PAN include segmental arterial mediolysis (SAM), fibromuscular dysplasia (FMD), and mycotic aneurysm. Intestinal involvement in PAN may mimic other inflammatory conditions like Crohn’s disease (CD). A clinical history of underlying systemic disorders, a dramatic response to immunosuppressive therapy, and most importantly abnormalities of visceral arteries could suggest a potential diagnosis of vasculitis. In the absence of bowel abnormality, involvement of branch points in mesenteric vessels and sparing of the main renal artery are useful clues to distinguish vasculitis from SAM and FMD, respectively. In a young patient with clinical manifestation of mesenteric ischemia, a thorough inspection of visceral arteries should be done in the arterial phase of CTE/MRE to find segmental arterial wall thickening, stenosis, or aneurysmal dilatation, especially in the presence of constitutional symptoms, elevated levels of inflammatory markers showing a systemic inflammatory response (such as erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP)). It is important to exclude vasculitis when mesenteric ischemia is not located in the classic watershed territory of bowel ischemia.

Kawasaki disease
Kawasaki disease is a self-limiting acute necrotizing vasculitis affecting the medium and small vessels in almost all organs. Infants and children under five years of age and of Asian ethnicity are most commonly affected. Adults are scarcely affected, and they commonly have incomplete forms of the disease. GI manifestations include abdominal pain and/or distension, vomiting, diarrhea, paralytic ileus, jaundice, hepatomegaly, gallbladder hydrops, and, far less frequently, serious complications such as pancreatitis, GI obstruction, or pseudo-obstruction [6, 52–54]. In the largest series of patients with adult-onset Kawasaki disease, 56% had GI manifestations [55]. Presentation of GI involvement can be associated with a poor prognosis [52]. Notably, we did not identify any investigation on MRE or CTE features in Kawasaki disease.

Small-vessel vasculitis
Small-vessel vasculitis predominantly involves small intraparenchymal vessels. This group based on pathogenesis is classified into two sub-groups; (1) antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and (2) immune complex-associated vasculitis. Current imaging techniques due to limited resolution are unable to directly visualize small-vessel vasculitis. Consequently, imaging is used to assess the damaging effects on affected organs [56, 57]. Common findings on CTE/MRE include intestinal submucosal edema or
hemorrhage, mesenteric edema, bowel dilatation, mesenteric fat haziness, and findings of acute or chronic mesenteric ischemia. Rarely, bowel perforation or stricture may occur as a consequence of inflammation. Differential diagnoses based on imaging include non-occlusive mesenteric ischemia, infectious enteritis, eosinophilic enteritis, IBD, radiation, and chemotherapy-induced enteritis.

**Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis**

Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) can occur at any age and includes granulomatosis with polyangiitis, microscopic polyangiitis, and eosinophilic granulomatosis with polyangiitis [58]. While AAV is a small-vessel vasculitis, it may cause inflammation of medium-sized vessels as well. A wide variety of organs, including the respiratory system, kidneys, nervous system, and GI tract, can be involved in AAV [59].

**Granulomatosis with polyangiitis (Wegener’s granulomatosis)**

Granulomatosis with polyangiitis (GPA) is a systemic necrotizing vasculitis of small- and medium-sized vessels with granulomatosis, which involves the upper and lower respiratory tracts and the kidneys. The condition is most common in the fourth or fifth decade of life [60]. Imaging can aid in determining the extent of disease, including pulmonary involvement [44]. The most common imaging findings in GPA are multiple and bilateral pulmonary nodules or masses [31, 61].

GI manifestations are present in 5–11% of patients with GPA. While any part of the GI tract might be involved, the abnormalities are commonly seen in the small and large bowel [6, 62]. GI manifestations include abdominal pain, ulceration (oral, esophageal, peptic, and intestinal), bloody diarrhea, peritonitis, and hematochezia. Rarely, patients may develop severe life-threatening intestinal involvement, including ischemia and perforation [6, 63].

**Fig. 4** Intestinal vasculitis in a 32-year-old female with PAN (polyarteritis nodosa) presenting with episodic abdominal pain, fever, anemia, and elevated liver enzymes and inflammatory markers. MRE was obtained. Coronal and axial T2-W images (A, B) show a single thickened segment of mid ileum (thick white arrows) with mild T2 hyperintensity more likely secondary to submucosal edema. This is associated with mild degree of upstream bowel distention. Coronal and axial post-contrast T1-W images (C, D) display mural hyperenhancement (thin white arrows) with a relatively striated pattern. DWI at a b-value of 800 s/mm² (E) shows restricted diffusion (white arrowhead), suggesting increased inflammatory cell density. The patient underwent double-balloon enteroscopy following this MRE, but it was complicated by bowel perforation at this segment. Then, surgical resection and ileostomy were performed. Histopathological assessment of resected segment revealed vasculitis consistent with polyarteritis nodosa (PAN). Heterogeneous enhancement of liver at arterial phase is also depicted (C), which raises suspicion for microvascular involvement (white dotted oval).
Prolonged immunosuppressive treatment is postulated as the primary underlying risk factor for intestinal necrosis and perforation [63, 64]. The severe manifestations typically develop in patients with extensive involvement of the other organs shortly after the diagnosis and initiating the immunosuppressive therapy [63, 64]. Other rare abdominal complications of GPA may include granulomatous colitis, granulomatous pancreatic mass, splenic or hepatic infarct or hemorrhage, and gastritis [6, 65, 66]. Multi-focal segmental circumferential mural thickening and abnormal hyperenhancement of the small and/or the large intestine can be detected on MRE/CTE (Fig. 6). Mesenteric vascular engorgement or unexpected large-vessel pathology (i.e., stenosis or dilation) and ascites can be observed as well [4, 51, 67]. Mesenteric fat haziness is another occasional finding, which stems from inflammation of the mesenteric vessels. In patients presenting with acute abdominal pain, signs of bowel perforation and/or necrosis, including focal intestinal pneumatosis and mild ascites, can be present. However, it should be noted that the immunosuppressive treatment might mask acute symptoms of intestinal ischemia (Fig. 6). The presence of pathologies in the kidneys can be a valuable diagnostic clue. Wedge-shaped T2-weighted hyperintensities and hypoenhancing lesions at the renal parenchyma with diffusion restriction on DWI are potential findings that can imply interstitial nephritis or renal ischemia (Fig. 7). Splenic involvement, including splenomegaly and splenic infarction, can also be detected [51]. In patients with bowel pathologies, concurrent abnormal laboratory results or imaging findings related to pulmonary or renal involvement and positive c-ANCA titer are helpful clues to differentiate GPA from other disorders (Fig. 8).

Microscopic polyangiitis
Microscopic polyangiitis (MPA) (also known as leukocytoclastic vasculitis) is a systemic autoimmune non-granulomatous vasculitis of the small vessels associated with the presence of ANCA, with a slightly higher prevalence in men [68]. Similar to PGA, kidneys and lungs are the primary organs that are involved. However, skin lesions (commonly present as palpable purpura), neurological manifestations, and GI involvement may also occur.
GI involvement is observed in 5–30% of the patients [6, 68]. Abdominal pain is the most frequent GI presentation, associated with nausea, vomiting, and diarrhea. Lower GI bleeding and ischemia are rare but life-threatening complications [46, 68–70], which can scarcely result in massive hemorrhage [71].

Radiologic features of bowel involvement are similar to other small-vessel vasculitides [4]. Correspondingly, intestinal concentric mural thickening and post-contrast hyperenhancement can be detected on enterography (Fig. 9). Engorgement of adjacent mesenteric vessels is one of the other potential findings. In acute cases, signs of bowel infarction or perforation can be rarely observed [4]. Similar to GPA, the presence of extra-intestinal renal and pulmonary pathologies can be an important diagnostic clue. Cutaneous lesions, including palpable purpura, especially on the extremities, may be the key clinical feature of leukocytoclastic vasculitis compared to similar conditions.

Eosinophilic granulomatosis with polyangiitis (Churg–Strauss)
Eosinophilic granulomatosis with polyangiitis (EPGA) is a necrotizing granulomatous small-vessel vasculitis characterized by adult-onset asthma and tissue eosinophilia. Patients can be divided into two groups: (1) ANCA-positive patients (30–40%) with predominant vasculitis manifestations, and (2) ANCA-negative patients with predominant eosinophilic presentations [72]. The most common signs and symptoms include asthma, lung infiltrates, neuropathy, and constitutional symptoms, while cardiac, skin, renal, and GI involvement are less frequent [72].

GI presentations can be observed in 30–50% of patients, including abdominal pain, diarrhea, melena, and hematochezia. No significant difference is found in the frequency of GI manifestations between ANCA-positive and ANCA-negative patients [72]. Vasculitis of the mesenteric artery is the most prevalent cause of GI involvement, resulting in bowel ischemia. Mucosal eosinophilic infiltration can be another underlying cause of ulceration, motility disorders, and obstructive symptoms [6]. Mucosal ulcers, which are typically limited to the small intestine, can be detected by endoscopy [72]. Severe complications, including bowel obstruction and stenosis, can occur in 22–45% of the patients [72]. Hepatobiliary complications are infrequent and include cholestasis and
liver infarction [73–75]. The potential underlying causes of hepatic involvement are fibrinoid necrosis of arteries, eosinophilic infiltration, and granulomatous reaction in the portal area. These conditions may be seen in addition to the irregular narrowing of the small hepatic arteries, which can be detected by angiography [74].

On MRE, intestinal mural hyperenhancement can be delineated, which may or may not be associated with
mural thickening (Fig. 10) [76]. Mural hyperenhancement can also be evident on CT images [72], which occurs as a result of mesenteric vasculitis or mucosal eosinophilic infiltrations [4]. Submucosal edema causes a striated bowel wall appearance with diffuse mucosal enhancement manifesting as a halo sign [51]. Signs of severe complications include bowel obstruction and stenosis [77–79], and as a result, bowel dilatation is a frequent CT finding [72]. Hepatic infarction may present as multiple wedge-shaped areas of hypoattenuation. Moreover, signs of eosinophilic abscess, such as intraparenchymal gas and fluid formation, may also be evident [80]. Approximately 70% of patients with EGPA present with pulmonary involvement, such as patchy, transient, and non-segmental opacities that have no predilection for any lung zone. Since intestinal vasculitis related to EGPA could only present as increased wall enhancement on MRE/CTE without gross mural thickening, attention should be paid beyond the GI tract searching for important clues such as infarction of the visceral organs or lung involvement, particularly in patients with a history of asthma and eosinophilia [81].

**Immune complex-associated vasculitis**

Similar to AAV, medium-sized vessels can be involved in addition to small vessels in immune complex-associated vasculitis. Activation of complements by the immune complexes leads to the attraction of neutrophils to the vessel wall and subsequently inflammation and vascular lesion. Radiologic findings are similar to other small-vessel vasculitides. This group includes immunoglobulin (Ig) A vasculitis and cryoglobulinemic vasculitis.

**IgA vasculitis (Henoch–Schönlein purpura)**

IgA vasculitis, formerly called Henoch–Schönlein purpura (HSP), is a systemic, leukocytoclastic vasculitis affecting small vessels. Being the most common vasculitis in children, IgA vasculitis in adulthood is less prevalent than in childhood. While the etiology is not well understood, genetic and environmental factors play a major role in disease pathogenesis. The most common manifestations are palpable purpura, arthralgia and/or arthritis, glomerulonephritis, and acute enteritis [82].

GI involvement can be observed in approximately two-thirds of patients. Abdominal pain is the most common...
GI presentation, followed by nausea, vomiting, melena, and/or rectorrhagia [83]. In 40–50% of the patients, the mesenteric circulation is involved [26]. Rarely (3–5%) patients may experience serious complications, including bowel infarct, perforation [84], obstruction, or irreducible intussusception [85]. Intestinal ischemia can even be the initial manifestation of the disease (29). Seldom (<5%) the hepatobiliary system can be involved as well [86].

On imaging, signs of bowel ischemia and edema can be observed, including segmental mural thickening and post-contrast focal mural hypoenhancement (Fig. 11). However, hemorrhage is commonly limited to mucosa and submucosa and is self-limiting [4, 87]. Importantly, glucocorticoids, which are used to alleviate clinical symptoms such as arthralgia, can mask pain and signs of bowel ischemia [88]. Descending duodenum and the terminal ileum are frequently involved [89]. Dramatic improvement after corticosteroid therapy is a characteristic feature of GI involvement [89]. Gallbladder wall thickening can also be observed in 25% of those with hepatobiliary involvement [86]. The diagnosis of HSP is mainly based on typical clinical signs and symptoms like skin rash, arthritis, colicky abdominal pain, GI bleeding, and hematuria. Imaging characteristics of the bowel wall are also helpful in diagnosis and management. Based on our clinical experience, while mural hyperdensity in non-contrast CTE images favors submucosal hemorrhage, focal hypoenhancement in post-contrast images suggests ischemic changes or edema associated with inflammation.

**Cryoglobulinemic vasculitis**

Cryoglobulins are abnormal immune system proteins that can deposit in the small vessel walls leading to cryoglobulinemic vasculitis. The skin, kidneys, joints, and peripheral nervous system are commonly involved. Type I cryoglobulinemic vasculitis is usually related to an underlying lymphoproliferative disorder. Types II (monoclonal) and III (polyclonal) are typically associated with rheumatoid factor activity. The main underlying causes of cryoglobulins production include lymphoproliferative disorders, systemic lupus erythematosus (SLE), Sjögren syndrome, and rheumatoid arthritis [90].

GI involvement is rare but can become disastrous. These manifestations include abdominal pain, melena, and more severe complications, such as intestinal perforation, intestinal ischemia, acute cholecystitis, and pancreatitis [91]. Liver involvement can be observed in up to 60% of the patients [6].

**Fig. 10** Intestinal vasculitis and perforation in a 32-year-old male with a history of asthma presenting with abdominal pain and eosinophilia. MRE was obtained. Coronal and axial post-contrast T1-W images (A, B) display increased mural enhancement at the terminal ileum without thickening (thick white arrows). Three months later (C, D), the patient presents to the ER with acute abdominal pain without definite diagnosis or treatment. Chest X-ray (C) shows subdiaphragmatic free gas (thick black arrow). He underwent an emergency laparotomy. Intraoperative photograph (D) revealed focal bowel perforation at the distal ileal segment. After segmental bowel resection, an ileostomy was performed. Eight months after surgery (E, F), he was admitted for RUQ pain and dyspnea. Axial chest CT image (E) demonstrates bilateral GGO and consolidations with lobular distribution (white dotted oval). Axial contrast-enhanced CT image (F) shows multiple wedge-shaped areas of hypoattenuation in the liver (thin white arrows), consistent with hepatic infarction. Intraparenchymal gas formation is also evident at the infarcted segment, suggestive for abscess formation (black dotted oval). Small-vessel vasculitis was confirmed based on clinical/laboratory data and pathology results suggestive of Churg–Strauss syndrome. Unfortunately, he died secondary to sepsis.
Variable-vessel vasculitis

The vasculitis does not have any predominant vessel size or type involvement. This group includes Behçet’s disease and Cogan syndrome [13]. In the latter syndrome, only a few cases of mesenteric vasculitis have been reported [92]. The location, extent, and size of the involved vessels determine the radiologic and clinical signs. Differential diagnoses of MRE/CTE findings are non-occlusive mesenteric ischemia, infectious enteritis, eosinophilic enteritis, IBD, radiation enteritis, and chemotherapy-induced enteritis.

Behçet disease

Behçet’s disease is a systemic inflammatory vasculitis affecting vessels of all sizes and also multiple organs. Ocular lesions, oral aphthous ulcers, genital ulcers, and skin lesions are the characteristic manifestations, which can be associated with vascular, neurological, and GI involvement. The prevalence of Behçet’s disease is higher in the Mediterranean, Middle East, and the Far East regions (along the ancient Silk Route) and is associated with the distribution of HLA-B51. The disease onset is commonly at the third or fourth decades of life [93].

The prevalence of GI manifestations is widely varied in different geographic regions, ranging from 1.4 to 60% [94–96]. GI involvement is more frequent in the Far East than in the Middle East and Europe. Abdominal pain, usually in the right lower quadrant, is the most common presentation, followed by diarrhea and GI bleeding [96]. In Behçet’s disease, GI complications typically have a relapsing nature [97]. The ileocolic region, including the ileocecal area, ileum, or different colonic segments, is most commonly involved [98, 99]. Bleeding and perforation are among serious complications; however, in chronic cases, closed perforation might develop. Acute lower GI bleeding has been reported in approximately 11–25% of those with GI manifestations, and it might be the initial GI presentation [100, 101]. In addition to intestinal involvement, thrombosis of the hepatic vein or inferior vena cava can result in hepatic complications, particularly Budd–Chiari syndrome. Very rarely, large-vessel involvement might result in the formation of abdominal aorta aneurysms [102].

On imaging, irregular circumferential mural thickening with homogeneous mural enhancement is the most common finding of bowel involvement. Behçet’s disease can significantly mimic CD on MRE. Deep penetrating ulcers and restricted diffusion on DWI in the involved segment could also be observed (Fig. 12) [20, 103]. Unlike CD, there is no specific predilection to the mesenteric side in Behçet’s disease with less surrounding mesenteric inflammatory changes (Fig. 13). Peker and colleagues reported a specificity of 100% and sensitivity of 57% for polypoid patterns and homogeneous mural enhancement findings in distinguishing Behçet’s from CD [104]. While pathologies of more proximal ileal segments favor small bowel CD, ileocecal involvement favors Behçet’s disease. Additionally, patients with Behçet’s disease may have a shorter length of the involved segment than CD [104]. History of recurrent oral and genital ulcers and uveitis is also important to make the diagnosis. Mural enhancing saccular pseudoaneurysms of the abdominal aorta may also be detected [40].
Vasculitis associated with systemic diseases

According to the revised Chapel Hill Consensus Conference nomenclature system, vasculitides secondary to connective tissue and autoimmune diseases are classified as vasculitis associated with systemic diseases [13]. While small vessels are commonly involved, medium and large vessels may also be affected. SLE, rheumatoid arthritis, systemic sclerosis (scleroderma), and antiphospholipid antibody syndrome are among diseases associated with vasculitis that can have concurrent GI involvement [6, 105–107].

Systemic lupus erythematosus (SLE)

SLE is a complex systemic disease with a female predominance resulting from immune complex deposition and production of auto-antibodies in various organs. The prevalence of vasculitis in patients with SLE ranges from 11 to 36%. Lupus vasculitis has various clinical manifestations as a result of involving vessels of all sizes [108, 109]. However, small-sized vessels are more frequently affected, and mesenteric vasculitis, also known as lupus enteritis, is far less common, with a prevalence of 0.2% to 9.7%, mostly involving the SMA [110]. SMA involvement can result in ileal and jejunal ischemia [108]. Lupus mesenteric vasculitis commonly presents with acute abdominal pain with diffuse localization [110]. Additionally, thrombosis of the mesenteric arteries may also occur even in the absence of vasculitis [111]. Rarely, GI manifestations, such as intestinal ischemia, can be the initial presentation of the systemic disease [47]. Notably, secondary Sjögren’s syndrome may occur in approximately 14%–17.8% of SLE patients (Fig. 14) [112].

Imaging is an indispensable tool in the assessment of lupus enteritis. Multi-focal segmental circumferential wall thickening, submucosal edema, and mucosal ischemic changes are among the potential intestinal findings on MRE/CTE. Bowel wall thickening commonly caused by edema can result in a “thumb printing sign” presentation that can be found in bowel ischemia [113, 114]. The diffuse circumferential wall thickening with submucosal edema gives rise to a “target sign” or “double-halo” finding, which can help in distinguishing SLE from other pathologies [115, 116]. The local hypervascular appearance of the adjacent mesentery, often seen in CD, also known as the “Comb sign,” is one of the other findings on CT of patients with lupus enteritis. Collectively, comb sign and/or target signs are present in approximately 70% of the patients with lupus enteritis [117].

The genitourinary system is frequently involved in SLE presenting as hydronephrosis, cystitis, and lupus nephritis, which could be used to reach the diagnosis (Fig. 15). Vasculitis should always be considered in patients with...
enteritis accompanied by single- or multi-organ infarcts (Fig. 16). Lupus enteritis must be suspected in a young patient with evidence of serositis (pleural effusion, pericardial effusion, or ascites). Imaging appearance of lupus enteritis can mimic CD, although the presence of ascites is not common in CD and should raise suspicion of serositis. Bowel ischemic changes may present mucosal hypoenhancement and serosal hyperenhancement on MRE suggestive for early to intermediate stage bowel ischemia compared to transmural necrosis seen at late-stage ischemia [21]. Similar imaging features might be reported in antiphospholipid antibody syndrome, which is characterized by thrombosis and/or recurrent early pregnancy loss and is highly associated with SLE. However, abdominal vessel thrombosis can be a key diagnostic clue (Fig. 17).

Systemic sclerosis (Scleroderma)
Systemic sclerosis is an autoimmune disease leading to fibrosis and disfiguration of the skin, along with dysfunctioning lungs, kidneys, heart, and GI tract [118]. Without histopathological investigation, it is difficult to distinguish between systemic sclerosis-related vasculopathy, which is non-inflammatory, and concurrent vasculitis [105]. However, whether due to vasculopathy or vasculitis, GI manifestations are very common in systemic sclerosis, with involvement of the oral cavity, esophagus, stomach, small and large intestine, liver, and pancreas. In nearly 10% of the patients, GI symptoms are the initial presentation of the systemic disease. The small intestine is the second most frequently involved organ, although the majority of patients with intestinal pathologies remain asymptomatic [119].

Small bowel involvement includes stiffness and atrophy of the smooth muscles leading to reduced small bowel mobility, chronic or acute intestinal pseudo-obstruction, small intestinal bacterial overgrowth, pneumatosis cystoides intestinalis, and jejunal diverticula. Smooth muscle atrophy and fibrosis of the intestinal wall with dominant inner circular muscle layer involvement compared to the outer longitudinal layer result in a characteristic finding in systemic sclerosis, namely “hidebound sign,” which is an increased number of bowel folds stacked together in spite of luminal distention without an increase in interfold distance [120]. Intestinal mural thickening with a

Fig. 13 Intestinal vasculitis in a 26-year-old female with known Behçet’s disease and positive HLA-B5 complaining of episodic RLQ pain. MRE was obtained. Coronal T2-W image (A) demonstrates irregular circumferential mural thickening of the cecum and proximal ascending colon (thick black arrows) without mesenteric side predilection or pericolic infiltration. Coronal and axial post-contrast T1-W images (B, C) show marked enhancement of thickened colonic wall (thick white arrows) mildly extending to ileocecal valve (thin white arrow) with low-grade mural hyperenhancement in the adjacent terminal ileum (white arrowhead). Axial DWI image (D) obtained at a b-value of 800 s/mm² reveals restricted diffusion in the thickened cecum (white dotted oval).
**Fig. 14** Intestinal ischemia in a 31-year-old female with known SLE-Sjögren overlap syndrome presenting with fever, anemia, and LUQ pain. MRE was obtained. Coronal T2-W and post-contrast T1-W images (A, B) display mural thickening, mucosal hypoenhancement, and serosal hyperenhancement of descending colon adjacent to the splenic flexure (thick white arrows) suggestive for mucosal ischemic changes. Coronal T2-W and post-contrast T1-W images (C, D) display mural thickening and increased mural enhancement of the cecum and ascending colon (Thin white arrows). Axial T2-W image and corresponding DWI sequence (E, F) show concentric mural thickening of the proximal ascending colon, demonstrating restricted diffusion (white arrowheads), in favor of ischemic changes or increased inflammatory cells. Coronal post-contrast T1-W image (G) reveals subcapsular areas of splenic hypoenhancement (white dotted oval), indicative of infarction. Pale and fragile mucosa is seen in colonoscopic view (H) of the involved colonic splenic flexure.

**Fig. 15** Intestinal vasculitis in a 42-year-old female with a known history of SLE and hypothyroidism presenting with generalized abdominal pain, fever, anemia, and elevated inflammatory markers. She also complained of dysuria, hematuria, and frequency. MRE was obtained. Axial T2-W images (A, B) show bilateral hydronephrosis (thin white arrows) and multi-focal segmental circumferential mural thickening involving several jejunal and ileal loops (thick white arrows). Axial T2-W image (C) displays an under distended urinary bladder containing a Foley catheter showing diffuse wall thickening (thick black arrows). Coronal T2-W and post-contrast T1-W images (D, E) demonstrate multi-segmental mural thickening involving several jejunal and ileal loops showing increased enhancement due to active nature of inflammation (black arrowheads). Submucosal edema is also noted (thin black arrow). Axial T2-W and post-contrast T1-W images (F, G) also reveal involvement of rectosigmoid colon (white dotted ovals). Small amount of free fluid is noted at the abdominopelvic cavity (black asterisks).
hypointense appearance on T2-weighted imaging indicates mural fibrosis (Fig. 18). Additionally, asymmetric small bowel wall fibrosis can result in intestinal sacculcation, which may present as multiple wide-mouthed outpouchings involving all intestinal wall layers [120]. Diffuse dilation of the small intestine (particularly jejunum) may occur, mainly upstream to the fibrotic bowel strictures. Colonic dilation may develop following acute colonic pseudo-obstruction, also known as Ogilvie syndrome (Fig. 19). Moreover, pneumatosis cystoides intestinalis, defined as air-filled cysts within the intestinal wall, can be detected on enterography. Rupture of these intraluminal cysts can result in benign or sterile pneumoperitoneum.

Subcutaneous calcification is a valuable extra-intestinal finding suggestive of an underlying systemic cause (e.g., scleroderma) for intestinal disease (Fig. 20). Meanwhile, esophageal dilatation and rarely hepatobiliary pathologies (nearly in 1.5% of patients), including hepatic duct obstruction due to vasculitis and as manifestations of primary biliary cirrhosis, can be detected as extra-intestinal pathologies [119].

Rheumatoid arthritis
Rheumatoid arthritis-associated vasculitis is extremely rare and can occur in patients with long-standing erosive rheumatoid arthritis [107]. Small- and medium-sized vessels are usually involved. GI manifestations may develop in 10–38% of patients with rheumatoid arthritis-associated vasculitis. These presentations include segmental or extensive intestinal infarction, ileal stricture [121, 122], ischemic ulcers of the intestine, and bowel perforation. Rarely other extra-intestinal findings, including hepatomegaly, intrahepatic hemorrhage, and pancreatic necrosis, can be evident [123].

Single-organ vasculitis (SOV)
This condition rarely occurs in the abdomen, and involvement of the abdominal organs is commonly part of systemic vasculitis. However, SOV in the GI tract has been reported in the stomach, small and large intestine, and most commonly appendix. Abdominal pain is the most frequent manifestation. Bowel infarction, perforation, bleeding, and solid-organ infarction are serious complications of GI SOV [124, 125]. The imaging findings include abnormalities of the corresponding arteries, intestinal wall thickening, bowel infarction, and infarcted areas within solid organs, including the spleen and liver [124].

Frequent mimickers of vasculitis on MRE/CTE
Inflammatory bowel disease (IBD)
IBD, particularly CD, is one of the mimickers of AAV or Behçet’s disease. The common findings on MRE/CTE

Fig. 16 Intestinal vasculitis and renal infarction in a 29-year-old female with known SLE presenting with fever, malaise, generalized abdominal pain, dyspnea, and elevated ESR. An abdominopelvic CT scan with IV and oral contrast was done. There are subpleural patchy areas of GGO at the left lower lobe suggestive of lupus pneumonitis (thick black arrows in A). Small pericardial effusion and thickening (thin white arrows in B) are seen in favor of serositis. There is a wedge-shaped subcapsular hypodensity in the spleen (white dotted oval in C), indicative of splenic infarction. Multiple bilateral wedge-shaped renal parenchymal infarcts are also depicted (thick white arrows in D). Diffuse circumferential wall thickening of ileal loops (white arrowheads in E and F) is evident, representing lupus enteritis.
include multi-focal intestinal mural thickening, striated mural hyperenhancement, and engorgement of mesenteric vessels. The predominant involvement of the bowel mesentery in CD compared to vasculitis can be an essential discriminating factor. Other helpful clues suggestive of CD are the fibrofatty proliferation of the mesentery and pseudodiverticula formation (Fig. 21). Notably, IBD can occasionally be associated with vasculitis [126]. Therefore, in addition to the clinical presentation, endoscopic and histologic investigations may become necessary to differentiate these entities [127].

**Eosinophilic enteritis**

There is an important controversy as to whether eosinophilic gastroenteritis is categorized as the pathogenic spectrum of hypereosinophilic syndrome or EGPA [128]. Imaging findings of eosinophilic enteritis are nonspecific, with mural thickening being a common finding. Other radiologic presentations include nodularity, luminal narrowing, and inflammation in the adjacent mesentery [129]. The previous history of allergy and presence of eosinophilia are important diagnostic clues for eosinophilic enteritis. Similar to vasculitis, corticosteroid therapy results in dramatic alleviation of the signs and symptoms [130].

**Graft-versus-host disease (GVHD)**

GVHD is a life-threatening complication of allogeneic stem cell or bone marrow transplant (HSCT) involving multiple organ systems, including the GI tract, and frequently is associated with skin involvement. Imaging findings are commonly observed in the GI tract and hepatobiliary system and are non-specific. These include ascites, periportal edema, intestinal wall thickening, mucosal hyperenhancement, bowel dilatation, and submucosal edema, leading to a tubular featureless appearance of the small intestine, called the ribbon sign.
The main differential diagnoses include neutropenic enterocolitis, infectious enterocolitis, or radiation enteritis. The location and extent of bowel involvement, clinical history, laboratory findings, and stool studies may aid in distinguishing GVHD from intestinal vasculitis.
Segmental arterial mediolysis (SAM)
SAM is a non-atherosclerotic and non-inflammatory arteriopathy more commonly affecting middle-aged and elderly patients. It results from lysis of the smooth muscle of the vascular outer layer. Imaging findings in SAM are similar to vasculitis and include aneurysmal dilation, dissection, stenosis, occlusion, and mural thrombosis of splanchnic arteries, particularly in SMA. However, SAM typically involves only the splanchnic arteries and spares other vessels like renal arteries and mesenteric arterial bifurcations from damage [12, 135]. Normal levels of inflammatory or immune markers can also help to differentiate SAM from vasculitis.

Infectious enteritis
Enterography is usually not able to detect infectious enteritis at the early stages. However, chronic infectious diseases, including tuberculosis and cytomegalovirus (CMV), are usually noticeable on CTE and MRE. Findings on enterography include segmental circumferential wall thickening, hyperenhancement, mural edema, and enlargement of the mesenteric lymph nodes [120, 136]. Infectious enteritis should always be considered, particularly in patients with immunodeficiency. Clinical history, laboratory data, and a trial of antibiotic therapy are helpful to confirm the diagnosis. Infectious enteritis is always a challenging differential diagnosis for intestinal vasculitis.

Angioedema
Angioedema is the swelling of the body following protein extravasation due to increased vascular permeability. The most common causes of angioedema include hereditary or acquired deficiency of C1-esterase inhibitor and medications, such as angiotensin-converting enzyme (ACE) inhibitors [137]. Angioedema commonly presents with cutaneous manifestations; however, in some cases, intra-abdominal involvement may occur without superficial manifestations. Patients with GI involvement commonly present with non-specific manifestations, including abdominal pain, nausea, and vomiting; therefore, imaging plays a key role in diagnosis [138]. Findings on CT include circumferential or asymmetric bowel wall thickening, mural stratification or halo sign due to submucosal edema, prominent mesenteric vessels, ascites, and hyperenhancement [137, 139]. Additionally, MR imaging shows submucosal edema. A thorough evaluation of patient and
family history and review of medication usage history in addition to the assessment of complement markers can aid in the differentiation of angioedema from vasculitides [137].

Diagnostic strategy

Figure 23 presents a flowchart for inference about a potential diagnosis of intestinal vasculitis on MRE or CTE. Imaging features are often non-specific, including mural thickening, submucosal edema, mural hyper/hypoenhancement, and rarely stricture or perforation. Positive laboratory biomarkers, multi-organ involvement, and the presence of vascular abnormalities, especially in a young/middle-aged patient without evidence of atherosclerosis, raise the clinical suspicion of vasculitis. Since intestinal vasculitis often mimics CD, a careful inspection must be done searching for fistula/abscess, mesenteric fat wrapping, or dominant involvement of the adjacent mesenteric border. Moreover, rapid response to corticosteroids, acute presentation, multi-segmental involvement at unusual sites, i.e., esophagus, duodenum, and rectum, or diffuse long segmental bowel involvement without skip may also be suggestive of vasculitis. Infectious enteritis is another differential, particularly in immunocompromised patients. As a challenging issue, any suspicion of infectious enteritis may postpone life-saving treatment with corticosteroid or immunosuppressive agents in patients with vasculitis. Diagnosis of intestinal vasculitis (Fig. 24) relies on a combination of history, clinical presentation, imaging, laboratory findings, and histopathology. The radiologists need to be aware of the findings suggestive of the diagnosis of intestinal vasculitis to avoid potential missed or late diagnosis. Table 1 summarizes clinical clues and enterography findings of common vasculitides involving the GI tract.

Limitations

While this review provides a comprehensive summary of imaging findings of intestinal involvement in vasculitis with the demonstrative cases, it faces several limitations. The sparse data on MRE and CTE findings of vasculitis were the major limitation of the literature, which might have affected this study. We did not identify any report in the literature about the MRE and CTE of some vasculitides, such as Takayasu arteritis, GCA, Kawasaki disease, and HSP. This review highlighted this gap in the literature warranting further investigation. Moreover, we could not include the images from a limited number of cases such as GCA, Kawasaki disease, and cryoglobulinemic vasculitis due to the very low frequency of these cases with intestinal involvement.

Conclusion

While gastrointestinal presentations are uncommon manifestations in vasculitis, their timely diagnosis holds immense importance. Establishing the diagnosis is hindered by the heterogeneous and non-specific manifestations of the vasculitides and usually requires a combination of clinical, imaging, laboratory, and histopathological data. MRE and CTE may be utilized in diagnostic investigations of patients presenting with GI involvement of a previously undiagnosed or diagnosed vasculitis. As previously described in detail, several key technical points on MRE/CTE protocols should be considered in these patients. In order to make the correct diagnosis, the imaging features need to be linked with the relevant clinical and laboratory data in a two-way street.
## Table 1
Vasculitides with detected complications on enterography

| Vasculitis                      | Clinical clues                                                                 | Imaging pearl for the radiologists                                                                 | Mimickers on MRE/CTE                                                                 |
|--------------------------------|--------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------|
| **Large-vessel vasculitis**    |                                                                                  |                                                                                                |                                                                                      |
| Takayasu arteritis             | Age at onset ≤ 40 y                                                            | Involvement of the aorta and the origin of major visceral arteries, presenting with concentric mural thickening, transmural calcification, luminal stenosis, occlusion, aneurysmal changes, and collateral vessels formation | Mesenteric ischemia due to other etiologies                                         |
|                               | Claudication of the extremities                                                 | Double ring vascular enhancement pattern                                                             | Atherosclerosis                                                                      |
|                               | Decreased brachial artery pulse                                                |                                                                                                   | Connective tissue disorders                                                          |
|                               | Blood pressure difference > 10 mm Hg between the arms                          |                                                                                                   | Marfan syndrome                                                                      |
|                               | Bruit over the subclavian arteries or aorta                                    |                                                                                                   | Ehlers-Danlos syndrome                                                               |
|                               | Abdominal bruit in 14%                                                         |                                                                                                   | Loeys-Dietz syndrome                                                                 |
| Giant cell arteritis           | Patient age > 50 years                                                         | Signs of mesenteric ischemia                                                                        |                                                                                      |
|                               | New-onset headache                                                             |                                                                                                   |                                                                                      |
|                               | Temporal artery abnormality (tenderness or decreased pulsation)                |                                                                                                   |                                                                                      |
|                               | ESR ≥ 50 mm/h                                                                  |                                                                                                   |                                                                                      |
|                               | Abnormal temporal artery biopsy results                                         |                                                                                                   |                                                                                      |
|                               | Abdominal pain may indicate aorta aneurysm or dilation                          |                                                                                                   |                                                                                      |
| **Medium-vessel vasculitis**   |                                                                                  |                                                                                                |                                                                                      |
| Polyarteritis nodosa           | Disease-associated weight loss ≥ 4 kg                                           | Involvement of mesenteric and visceral arteries, e.g., stenosis, vessel irregularity, aneurysmal dilation, and arterial mural thickening, with a predilection for superior mesenteric artery (SMA) branches | Segmental arterial medialysis (SAM)                                                  |
|                               | Livedo reticularis                                                             | Segmental bowel mural thickening                                                                      | Fibromuscular dysplasia (FMD)                                                        |
|                               | Testicular pain or tenderness                                                   | Submucosal edema and mural hyperenhancement with a striated pattern                                  | Mycotic aneurysm                                                                     |
|                               | Myalgia, weakness, or leg tenderness                                           | Visceral infarction (liver, spleen, kidneys, and intestine)                                           |                                                                                      |
|                               | Mono- or polyneuropathy                                                        | Spontaneous abdominal hemorrhage secondary to a ruptured aneurysm                                     |                                                                                      |
|                               | Diastolic blood pressure > 90 mm Hg                                            | Heterogeneous liver or renal enhancement due to microvascular abnormalities                           |                                                                                      |
|                               | Elevated serum levels of creatinine or blood urea nitrogen                      |                                                                                                   |                                                                                      |
|                               | Presence of hepatitis B reactants in serum                                      |                                                                                                   |                                                                                      |
|                               | Biopsy of a small- or medium-sized artery containing neutrophils or mixed leukocyte infiltrate |                                                                                                   |                                                                                      |
| Vasculitis                                    | Clinical clues                                                                 | Imaging pearl for the radiologists                                                                 | Mimickers on MRE/CTE                                                                 |
|----------------------------------------------|---------------------------------------------------------------------------------|----------------------------------------------------------------------------------------------------|-------------------------------------------------------------------------------------|
| Small-vessel vasculitis                      |                                                                                  |                                                                                                    | Non-occlusive mesenteric ischemia                                                   |
| ANCA-associated vasculitis                   |                                                                                  |                                                                                                    | Infectious enteritis                                                                 |
| Granulomatosis with polyangitis (Wegener’s   | Nasal and oral inflammation: oral ulcers or bloody nasal discharge             | Intestinal ischemia presenting with multi-focal segmental circumferential mural thickening and     | Eosinophilic enteritis                                                              |
| granulomatosis)                              | Microhematuria                                                                   | abnormal hyperenhancement                                                                         | Inflammatory bowel disease                                                           |
|                                              | Granulomatous inflammation of arterial walls or extravascular tissue            | Abnormal chest radiographic findings: multiple and bilateral pulmonary nodules fixed infiltrates,   | Radiation and chemotherapy induced enteritis                                         |
|                                              |                                                                                  | or masses or alveolar hemorrhage                                                                   |                                                                                      |
|                                              |                                                                                  | Renal pathologies, i.e., ischemia and interstitial nephritis                                      |                                                                                      |
|                                              |                                                                                  | Visceral infarction (liver, spleen, and kidneys)                                                   |                                                                                      |
|                                              |                                                                                  |                                                                                                    |                                                                                        |
| Eosinophilic granulomatosis with polyangitis  | Asthma                                                                          | Submucosal edema and mural hyperenhancement with a striated pattern with or without thickening    |                                                                                        |
| (Churg-Strauss)                              | Eosinophilia > 10%                                                              | (halo sign)                                                                                       |                                                                                        |
|                                              | Mono- to polyneuropathy                                                          | Bowel dilatation, stenosis, or obstruction                                                         |                                                                                        |
|                                              | Nonfixed pulmonary infiltrates                                                   | Hepatobiliary complications, i.e., cholestasis and liver infarction                                |                                                                                        |
|                                              | Paranasal sinus abnormality                                                      | Pulmonary involvement (Non-Fixed parenchymal opacities)                                            |                                                                                        |
|                                              | Extravascular eosinophils                                                       |                                                                                                    |                                                                                        |
|                                              |                                                                                  |                                                                                                    |                                                                                        |
| Microscopic polyangiitis                     | Rapid progressive glomerulonephritis and/or alveolar hemorrhages                 | Concentric mural thickening                                                                        |                                                                                        |
|                                              | Histopathologic findings of small vessel vasculitis or necrotizing glomerulonephritis | Post-contrast T1-weighted hyperenhancement                                                          |                                                                                        |
|                                              | Symptoms suggestive of small vessel involvement                                  | Engorgement of mesenteric vessels                                                                  |                                                                                        |
|                                              | Skin lesions (commonly presented as palpable purpura)                           | Signs of bowel infarction or perforation                                                           |                                                                                        |
|                                              | Neurological manifestations                                                     | Renal pathologies (striated pattern)                                                               |                                                                                        |
|                                              |                                                                                  | Pulmonary pathologies                                                                              |                                                                                        |
|                                              |                                                                                  |                                                                                                    |                                                                                        |
| Immune complex-associated vasculitis         |                                                                                  |                                                                                                    |                                                                                        |
| IgA vasculitis                               | Age ≤ 20 years at disease onset                                                 | Signs of bowel ischemia and edema, e.g., segmentement mural thickening and post-contrast focal     |                                                                                        |
|                                              | Palpable purpura                                                                | mural hypo-enhancement                                                                             |                                                                                        |
|                                              | Acute abdominal pain                                                            | Self-limiting mucosal and submucosal hemorrhage                                                     |                                                                                        |
|                                              | Biopsy showing granulocytes in the walls of small vessels (presence of 2 or more)| Signs of bowel perforation, obstruction, or irreducible intussusception                           |                                                                                        |
|                                              | Arthralgia and/or arthritis                                                     | Gallbladder wall thickening                                                                         |                                                                                        |
|                                              | Glomerulonephritis                                                              |                                                                                                    |                                                                                        |
| Vasculitis                          | Clinical clues                                                                 | Imaging pearl for the radiologists                                                                 | Mimickers on MRE/CTE                                                                 |
|-----------------------------------|-------------------------------------------------------------------------------|--------------------------------------------------------------------------------------------------|----------------------------------------------------------------------------------|
| Variable-vessel vasculitis        | Mandate criteria: Recurrent oral aphthosis                                      | Predominant ileocecal involvement                                                              | Non-occlusive mesenteric ischemia                                                |
| Behçet disease                    | Minor criteria: Ocular lesions, Recurrent genital aphthosis, Skin lesions,     | Irregular circumferential mural thickening with homogeneous mural enhancement                   | Infectious enteritis                                                            |
|                                   | Positive pathergy test                                                         | Deep penetrating ulcers and restricted diffusion on DWI in the involved section                  | Eosinophilic enteritis                                                          |
|                                   | Predominant ileocecal involvement                                             | No specific predilection to the mesenteric side                                                | Inflammatory bowel disease                                                      |
|                                   | Irregular circumferential mural thickening with homogeneous mural enhancement | No surrounding mesenteric inflammatory changes                                                  | Radiation and chemotherapy-induced enteritis                                    |
|                                   | Deep penetrating ulcers and restricted diffusion on DWI in the involved section |                                                                                                 |                                                                                  |
|                                   | No specific predilection to the mesenteric side                                |                                                                                                 |                                                                                  |
|                                   | No surrounding mesenteric inflammatory changes                                 |                                                                                                 |                                                                                  |
|                                   | Non-occlusive mesenteric ischemia                                             |                                                                                                 |                                                                                  |
|                                   | Infectious enteritis                                                          |                                                                                                 |                                                                                  |
|                                   | Eosinophilic enteritis                                                         |                                                                                                 |                                                                                  |
|                                   | Inflammatory bowel disease                                                    |                                                                                                 |                                                                                  |
|                                   | Radiation and chemotherapy-induced enteritis                                  |                                                                                                 |                                                                                  |
| Vasculitis associated with systemic diseases | Multi-focal bowel wall thickening not confined to a single vascular territory |                                                                                                 |                                                                                  |
| Systemic Lupus Erythematosus (SLE)| Submucosal edema and mural hyper enhancement                                 |                                                                                                 |                                                                                  |
|                                   | Target sign (diffuse circumferential wall thickening with submucosal edema)    |                                                                                                 |                                                                                  |
|                                   | Evidence of serositis, e.g., ascites                                          |                                                                                                 |                                                                                  |
|                                   | Comb sign (The hypervascular appearance of the mesentery)                     |                                                                                                 |                                                                                  |
|                                   | Genitourinary involvement, e.g., hydrenephrosis, cystitis, and lupus nephritis |                                                                                                 |                                                                                  |
|                                   | Signs of solid-organ infarction, including wedge-shaped renal and splenic infarcts |                                                                                                 |                                                                                  |
|                                   | Systemic sclerosis                                                           |                                                                                                 |                                                                                  |
|                                   | Skin thickening of the fingers                                                |                                                                                                 |                                                                                  |
|                                   | Fingertip lesions                                                             |                                                                                                 |                                                                                  |
|                                   | Telangiectasia                                                                |                                                                                                 |                                                                                  |
|                                   | Abnormal nailfold capillaris                                                   |                                                                                                 |                                                                                  |
|                                   | Pulmonary arterial hypertension and/or Interstitial lung Disease               |                                                                                                 |                                                                                  |
|                                   | Raynaud’s phenomenon                                                          |                                                                                                 |                                                                                  |
|                                   | anti-centromere and/or anti-topoisomerase antibody positive                    |                                                                                                 |                                                                                  |
|                                   | Benign or sterile pneumatosis                                                  |                                                                                                 |                                                                                  |
|                                   | Hidebound sign (increased number of bowel folds stacked together despite luminal disten- |                                                                                                 |                                                                                  |
|                                   | tion without an increase in interfold distance                                |                                                                                                 |                                                                                  |
|                                   | Diffuse dilation of the small intestine (particularly jejunum)                |                                                                                                 |                                                                                  |
|                                   | Intestinal mural thickening, indicating mural fibrosis                        |                                                                                                 |                                                                                  |
|                                   | Multiple wide-mouthed outpouchings involving all intestinal wall layers        |                                                                                                 |                                                                                  |
|                                   | Subcutaneous calcifications                                                   |                                                                                                 |                                                                                  |
Fig. 23 A practical flowchart for potential diagnostic work up of intestinal vasculitis following MRE or CTE.
Abbreviations
AAV: Antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis; ACE: Angiotensin-converting enzyme; ANCA: Antineutrophil cytoplasmic antibody; CD: Crohn’s disease; CMV: Cytomegalovirus; CRP: C-reactive protein; CTE: Computed tomography enterography; EPGA: Eosinophilic granulomatosis with polyangiitis; ESGAR: European Society of Gastrointestinal and Abdominal Radiology; ESPR: European Society of Pediatric Radiology; ESR: Erythrocyte sedimentation rate; FMD: Fibromuscular dysplasia; GCA: Giant cell arteritis; GI: Gastrointestinal; GPA: Granulomatosis with polyangiitis; GVHD: Graft-versus-host disease; HSP: Henoch–Schönlein purpura; IBD: Inflammatory bowel disease; Ig: Immunoglobulin; MPA: Microscopic polyangiitis; Myo: Myocardial ischemia; NAD: Natural killer T cells; NOD/SCID: NOD/Lt scid beige; OSA: Obstructive sleep apnea; PAN: Polyarteritis nodosa; PET: Positron emission tomography; SAM: Segmental arterial mediolysis; SLE: Systemic lupus erythematosus; SMA: Superior mesenteric artery; SOV: Single-organ vasculitis.

Author contributions
MA contributed to conceptualization, investigation, resources, writing—original draft, and writing—review and editing. SM was involved in conceptualization, investigation, writing—original draft, and writing—review and editing. HK, AHD, and AS contributed to conceptualization and writing—review and editing. ARR was involved in conceptualization, investigation, resources, writing—original draft, writing—review and editing, and supervision. All authors read and approved the final manuscript.

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Competing interests
All authors have no competing interests.

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References
1. Saadoun D, Vautier M, Cacoub P (2021) Medium- and large-vessel vasculitis. Circulation 143(5):267–282
2. Broncano J, Vargas D, Bhalla S, Cummings KW, Raptis CA, Luna A (2018) CT and MR imaging of cardiothoracic vasculitis. Radiographics 38(4):997–1021
3. Konttinen YT, Rotar Z, Pettersson T, Nordstrom DC, Bacon P, Petersen J (2006) Roadmap to vasculitis. Acta Reumatol Port 31(1):15–36
4. Ha HK, Lee SH, Rha SE et al (2000) Radiologic features of vasculitis involving the gastrointestinal tract. Radiographics 20(3):779–794
5. Zhang X, Furth EE, Tondon R (2020) Vasculitis involving the gastrointestinal system is often incidental but critically important. Am J Clin Pathol 154(4):536–552
6. Soowamber M, Weizman AV, Pagnoux C (2017) Gastrointestinal aspects of vasculitides. Nat Rev Gastroenterol Hepatol 14(3):185–194
7. Anderson E, Gakhari N, Stull C, Caplan L (2018) Gastrointestinal and hepatic disease in vasculitis. Rheum Dis Clin North Am 44(1):1–14

Fig. 24 The diagnosis of intestinal vasculitis is based on a combination of the clinical manifestation and history, imaging, laboratory, and histopathology findings.
8. Elsayes KM, Al-Hawary MM, Jagdish J, Ganesh HS, Platt JF (2010) CT enterography: principles, trends, and interpretation of findings. Radiographics 30(7):1955–1970
9. Tolan DJM, Greenhalgh R, Zealley IA, Halligan S, Taylor SA (2010) MR enterographic manifestations of small bowel Crohn disease. Radiographics 30(2):367–384
10. Fidler JL, Guimaraes L, Einstein DM (2009) MR Imaging of the small bowel. Radiographics 29(6):1811–1825
11. Pagnoux C, Seror R, Henegar C, et al. (2010) French Vasculitis Study G: clinical features and outcomes in 348 patients with polyarteritis nodosa: a systematic retrospective study of patients diagnosed between 1963 and 2005 and entered into the French Vasculitis Study Group Database. Arthritis Rheum 62(2):616–626
12. Ghodasara N, Liddell R, Fishman EK, Johnson PT (2019) High-value multidetector CT angiography of the superior mesenteric artery: what emergency medicine physicians and interventional radiologists need to know. Radiographics 39(2):559–577
13. Jennette JC, Falk RJ, Bacon PA et al (2013) 2012 revised international chapel hill consensus conference nomenclature of vasculitides. Arthritis Rheum 65(1):1–11
14. Nienhuis PH, van Praagh GD, Glaudemans AWJM, Brouwer E, Slart RHJA (2021) A review on the value of imaging in differentiating between large vessel vasculitis and atherosclerosis. J Pers Med 11(3):779–794
15. Taylor SA, Avni F, Cronin CG et al (2017) The first joint ESGAR/ESPR consensus statement on the technical performance of cross-sectional small bowel and colonic imaging. Eur Radiol 27(6):2570–2582
16. Kanasaki S, Furukawa A, Fumoto K, et al. (2018) Acute mesenteric ischemia: multidetector CT findings and endovascular management. Radiographics 38(3):945–961
17. Furukawa A, Kanasaki S, Kono N, et al. (2009) CT diagnosis of acute mesenteric ischemia from various causes. AJR Am J Roentgenol 192(2):408–416
18. Nastri MV, Baptista LP, Baroni RH, et al. (2004) Gadolinium-enhanced three-dimensional MR angiography of Takayasu arteritis. Radiographics 24(3):773–786
19. Litmanovich DE, Yıldırım A, Bankier AA (2012) Insights into imaging of Takayasu’s arteritis. Radiographics 32(2):367–384
20. Chung SY, Ha HK, Kim JH et al (2001) Radiologic findings of Behçet syndrome involving the gastrointestinal tract. Radiographics 21(4):911–924
21. Davarpanah AH, Ghamari Khameneh A, Khosravi B, Mir A, Saffar H, Radmand AR (2021) Many faces of acute bowel ischemia: overview of radiologic staging. Insights Imaging 12(1):56
22. Sug MD, Menias CO, Lubner MG, et al. (2018) CT findings of acute small-bowel entities. Radiographics 38(5):1352–1369
23. de Souza AW, de Carvalho JF (2014) Diagnostic and classification criteria of Takayasu arteritis. J Autoimmun 48–49:79–83
24. Spira D, Kötter I, Ernemann U, et al. (2010) Imaging of primary and secondary inflammatory diseases involving large and medium-sized vessels and their potential mimics: a multitechnique approach. AJR Am J Roentgenol 194(3):848–856
25. Schmidt J, Kermani TA, Bacani AK, et al. (2013) Diagnostic features, treatment, and outcomes of Takayasu arteritis in a US cohort of 126 patients. Mayo Clin Proc 88(8):822–830
26. Koster MJ, Warrington KJ (2017) Vasculitis of the mesenteric circulation. Best Pract Res Clin Gastroenterol 31(1):85–96
27. Kermani TA, Crowson CS, Muratore F, Schmidt J, Matteson EL, Warrington KJ (2015) Extra-cranial giant cell arteritis and Takayasu arteritis: how similar are they? Semin Arthritis Rheum 44(6):724–728
28. Merem E, Hatiem E, Sergio EC, Khadja BEH, Ittimade N, Nabil MB (2020) Chronic dissection of the abdominal aorta as a rare complication of Takayasu disease. Radiol Case Rep 15(11):2188–2191
29. Kilic L, Kalyoncu U, Karadag O, et al. (2016) Inflammatory bowel diseases and Takayasu’s arteritis: coincidence or association? Int J Rheum Dis 19(8):814–818
30. Terao C, Matsumura T, Yoshifuji H et al (2015) Takayasu arteritis and ulcerative colitis: high rate of co-occurrence and genetic overlap. Arthritis Rheumatol 67(8):2226–2232
31. Hur JH, Chun EJ, Kwag HJ, et al. (2017) CT features of vasculitides based on the 2012 International Chapel Hill Consensus Conference Revised Classification. Korean J Radiol 18(5):786–798
32. Gotway MB, Arazo PA, Macedo TA, et al. (2005) Imaging findings in Takayasu’s arteritis. AJR Am J Roentgenol 184(6):1945–1950
33. Khandelwal N, Kalra N, Garg MK, et al. (2011) Multidetector CT angiography in Takayasu arteritis. Eur J Radiol 77(2):369–374
34. Younger DS (2019) Giant cell arteritis. Neurol Clin 37(2):335–344
35. Gribbison KB, Ponce C, Carette S et al (2020) Patterns of arterial disease in Takayasu arteritis and giant cell arteritis. Arthritis Care Res (Hoboken) 72(11):1615–1624
36. Annamalai A, Francis ML, Ratnatunga SK, Resch DS (2007) Giant cell arteritis presenting as small bowel infarction. J Gen Intern Med 22(1):140–144
37. Trimble MA, Weisz MA (2002) Infarction of the sigmoid colon secondary to giant cell arteritis. Rheumatology 41(1):108–110
38. Espitia O, Bloniz G, Urbanski G et al (2021) Symptomatic aortitis at giant cell arteritis diagnosis: a prognostic factor of aortic event. Arthritis Res Ther 23(1):14
39. Jud P, Verheyen N, Dejaco C et al (2020) Prevalence and prognostic factors for aortic dilatation in giant cell arteritis – a longitudinal study. Semin Arthritis Rheum 51(4):911–918
40. Restrepo CS, Ocazionez D, Suni R, Vargas D (2011) Aortitis: imaging spectrum of the infectious and inflammatory conditions of the aorta. Radiographics 31(2):435–451
41. Stanson AW, Friese JL, Johnson CM, et al. (2001) Polyarteritis nodosa: spectrum of angiographic findings. Radiographics 21(1):151–159
42. Hekali P, Rajander H, Pajari R, Stennman S, Somer T (1991) Diagnostic significance of angiographically observed visceral aneurysms with regard to polyarteritis nodosa. Acta Radiol 32(2):143–148
43. Hernandez-Rodriguez J, Alba MA, Prieto-Gonzalez S, Cid MC (2014) Diagnosis and classification of polyarteritis nodosa. J Autoimmun 48–49:848–89
44. Guggenberger KV, Bley TA (2020) Imaging in vasculitis. Curr Rheumatol Rep 22(8):34
45. Harada M, Yoshida H, Ikeda H et al (1999) Polyarteritis nodosa with mesenteric aneurysms demonstrated by angiography: report of a case and successful treatment of the patient with prednisolone and cyclophosphamide. J Gastroenterol 34(6):705–720
46. Passam FH, Diamantis ID, Perisinaki G, et al. (2004) Intestinal ischemia as the first manifestation of vasculitis. Semin Arthritis Rheum 34(1):431–441
47. Khandelwal N, Kalra N, Garg MK, et al. (2011) Multidetector CT angiography in Takayasu arteritis. Eur J Radiol 77(2):369–374
48. Vinemoth MJ, Lenz A, Adam G, Francois CJ, Bannas P (2020) Radiologic imaging in large and medium vessel vasculitis. Radiol Clin North Am 58(4):765–779
49. Ozaki K, Miyayama S, Ushio Y, Matsu O (2009) Renal involvement of polyarteritis nodosa: CT and MR findings. Abdom Imaging 34(2):265–270
50. Naeem M, Menias CO, Cail AJ, et al. (2021) Imaging spectrum of granulomatous diseases of the abdomen and pelvis. Radiographics 41(3):783–801
51. Fabi M, Cornalesi E, Pierantoni L, et al (2018) Gastrointestinal presentation of Kawasaki disease: a red flag for severe disease? PLoS One 13(9):e0202659
52. Colomba C, La Placa S, Saporito L, et al. (2018) Intestinal involvement in Kawasaki disease. J Pediatr 202:186–193
53. Kostner A, Seve P, Dauphin C, et al (2016) Kawasaki disease in adults: observations in France and literature review. Autoimmun Rev 15(3):242–249
56. Singhal M, Gupta P, Sharma A (2019) Imaging in small and medium vessel vasculitis. Int J Rheum Dis 22(Suppl 1):78–85
57. Jennette JC, Falk RJ (1997) Small-vessel vasculitis. N Engl J Med 337(21):1512–1523
58. Watts RA, Mahr A, Mohammad AJ, Gatenby P, Basu N, Flores-Suarez LF (2015) Classification, epidemiology, and clinical subgrouping of anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis. Nephrol Dial Transplant 30(Suppl 1):i14–22
59. Hunter RW, Welsh N, Farrah TE, Gallacher PJ, Dhaun N (2020) ANCA associated vasculitis. BMJ 369:n1070
60. Comarmond C, Cacoub P (2014) Granulomatosis with polyangiitis (Wegener): clinical aspects and treatment. Autoimmun Rev 13(1):1121–1125
61. Ananthakrishnan L, Sharma N, Kanne JP (2009) Wegener's granulomatosis in the chest: high-resolution CT findings. AJR Am J Roentgenol 192(3):676–682
62. Storeolum B, Gran JT, Koldingssen W (1998) Severe intestinal involvement in Wegener's granulomatosis: report of two cases and review of the literature. Br J Rheumatol 37(4):387–390
63. Sato H, Shima K, Sakata H, Ohtoh T (2019) Granulomatosis with polyangiitis with intestinal involvement successfully treated with rituximab and surgery. BMJ Case Rep 12(8):e230355
64. Yildirim AC, Koçak E, Yildiz P, et al. (2010) Multiple intestinal perforation in a patient with Wegener's granulomatosis: a case report and review of the literature. Gastroenterol Clin Biol 34(12):712–715
65. Valeneva Ya, Golemanov B, Tzolova N, Mitova R (2013) Pancreatic mass in Wegener's granulomatosis with polyangiitis: a case report and literature review. Clin Rheumatol 23(2):152–159
66. Chirinos JA, Tamariz LJ, Lopes G, et al. (2004) Large vessel involvement in Wegener's granulomatosis: report of two cases and review of the literature. Clin Rheumatol 23(2):152–159
67. Greco A, De Virgilio A, Rizzo M, et al. (2015) Microscopic polyangiitis: Advances in diagnostic and therapeutic approaches. Autoimmun Rev 14(9):837–844
68. Eriksson P, Segelmak Mark, Hallbook O (2018) Frequency, diagnosis, treatment, and outcome of gastrointestinal disease in granulomatosis with polyangiitis and microscopic polyangiitis. J Rheumatol 45(4):529–537
69. Fukushima M, Inoue S, Chno Y, et al. (2013) Microscopic polyangiitis complicated with ileal involvement detected by double-balloon endoscopy: a case report. BMC Gastroenterology 13:42
70. Ueda S, Matsumoto M, Ahn T, et al. (2001) Microscopic polyangiitis complicated with massive intestinal bleeding. J Gastroenterol 36(4):264–270
71. Trivoli G, Ternier B, Vaglio A. Eosinophilic granulomatosis with polyangiitis: understanding the disease and its management. Rheumatology (Oxford) 2020, 59(Suppl 3):ii84–ii94.
72. Harada M, Os J, Shibata M, et al. (2012) Churg-Strauss syndrome manifesting as cholestasis and diagnosed by liver biopsy. Hepatol Res 42(9):940–944
73. Otani Y, Anzai S, Shibuya H, et al. (2003) Churg-Strauss syndrome (CSS) manifested as necrosis of fingers and toes and liver infarction. J Dermatol 30(11):810–815
74. Sironen RK, Seppa A, Kosma VM, Kuopio T (2010) Churg-Strauss syndrome: a clinical case and review of the literature. Eur J Gastroenterol Hepatol 22(6):676–682
75. Tursen U, Guler A, Boyvat A (2003) Evaluation of clinical findings according to sex in 2313 Turkish patients with Behcet's disease. Int J Dermatol 42(5):346–351
76. Balta I, Akbay G, Kalkan G, Eksioglu M (2014) Demographic and clinical features of 521 Turkish patients with Behcet's disease. Int J Dermatol 53(5):564–569
77. Lee CR, Kim WH, Cho YS, et al. (2001) Colonoscopic findings in intestinal involvement in Behcet's disease: comprehensive review of multisystemic involvement. Radiographics 21(5):e31
78. Hayatani I, Hatemi G, Areli MS, et al. (2016) Characteristics, treatment, and long-term outcome of gastrointestinal involvement in Behcet's syndrome: a stroboscopic observational study from a dedicated multidisciplinary center. Medicine (Baltimore) 95(16):e3348
79. Park J, Cheon JH, Park YE, et al. (2017) Risk factors and outcomes of acute lower gastrointestinal bleeding in intestinal Behcet's disease. Int J Colorectal Dis 32(5):745–751
80. Roeyen G, Van Schil PE, Vannmaele RG et al (1997) Abdominal aortic aneurysm with lumbar vertebral erosion in Behcet’s disease. A case report and review of the literature. Eur J Vasc Endovasc Surg 13(2):242–246
103. Ha HK, Lee HJ, Yang SK et al (1998) Intestinal Behçet syndrome: CT features of patients with and patients without complications. Radiology 209(2):449–454
104. Peker E, Erden A, Erden İ, Düzgün N (2018) Intestinal Behçet disease: evaluation with MR enterography—a case-control study. AJR Am J Roentgenol 211(4):767–775
105. Kao L, Weyand C (2010) Vasculitis in systemic sclerosis. Int J Rheumatol 2010:385938
106. Norden DK, Ostrov BE, Shafritz AB, Von Feldt JM (1995) Vasculitis associated with antiphospholipid syndrome. Semin Arthritis Rheum 24(4):273–281
107. Sharma A, Dhoooria A, Aggarwal A, Rathi M, Chandran V (2016) Connective tissue disorder-associated vasculitis. Curr Rheumatol Rep 18(6):31
108. Barile-Fabris L, Hernandez-Cabrera MF, Barragan-Garfias JA (2014) Lupus mesenteric vasculitis: a delayed diagnosis. J Investig Med High Impact Case Rep 5(4):2324709617734246
109. Ju JH, Min JK, Jung CK et al (2009) Lupus enteritis. Radiographics 29(4):1069–1086
110. Mok CC (2005) Investigations and management of gastrointestinal and hepatic manifestations of systemic lupus erythematosus. Best Pract Res Clin Rheumatol 19(5):741–766
111. Pasoto SG, Adriano de Oliveira Martins V, Bonfa E (2019) Sjogren’s syndrome: a delayed diagnosis. J Investig Med High Impact Case Rep 5(4):2324709617734246
112. Cicero G, Blandino A, D’Angelo T, et al. (2018) Magnetic resonance enterography appraisal of lupus enteritis: a case report. Radiol Case Rep 13(5):915–919
113. Terra C, Ramos-Andrade D, Sá-Marques I, Brito J, Caseiro-Alves F, Curvo-Semedo L (2021) Duodenal imaging on the spotlight: from A to Z. Insights Imaging 12(1):94
114. Huang YT, Chung TW, Wang JJ (2017) Target sign in lupus enteritis. QJM Intern Med 110(4):245–246
115. Byun JY, Ha HK, Yu SY, et al. (1999) CT features of systemic lupus erythematosus in patients with acute abdominal pain: emphasis on ischemic bowel disease. Radiology 211(1):203–209
116. Brewer BN, Kamen DL (2018) Gastrointestinal and hepatic disease in systemic lupus erythematosus. Rheum Dis Clin N Am 44(1):165–175
117. Madani G, Katz RD, Haddock JA, Denton CP, Bell JR (2008) The role of radiology in the management of systemic sclerosis. Clin Radiol 63(9):959–967
118. McFarlane IM, Bhamra MS, Kreps A et al (2018) Gastrointestinal manifestations of systemic sclerosis. Rheumatology (Sunnyvale) 8(1):235
119. Amzallag-Bellenger E, Oudjit A, Ruiz A, Cadiot G, Soyer PA, Hoeffel CC (2012) Effectiveness of MR enterography for the assessment of small-bowel diseases beyond Crohn disease. Radiographics 32(5):1423–1444
120. Lubnner MG, Menias CO, Agtons M, et al. (2017) Imaging of abdominal and pelvic manifestations of graft-versus-host disease after hematopoietic stem cell transplantation. Radiology 258(3):660–671
121. Hoeffel C, Crema MD, Belkacem A, et al. (2006) Multi-detector row CT: spectrum of diseases involving the ileocecal area. Radiographics 26(S1):1373–1390
122. Ishigami K, Averill SL, Pollard JH, McDonald JM, Sato Y (2014) Radiologic manifestations of angioedema. Insights Imaging 5(3):365–374
123. Savino MR, Mittal PK, Miller FH (2017) MR imaging of intestinal angioedema related to angiotensin-converting enzyme inhibitors: Report of three cases and review of literature. Clin Imaging 43:122–126
124. De Backer AI, De Schepper AM, Vandeveerne JW, Schoeters P, Michelsen P, Stevens WJ (2001) CT of angioedema of the small bowel. AJR Am J Roentgenol 176(3):649–652

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