CASE REPORT

Acute massive congestive ischaemic colitis related to inferior mesenteric arteriovenous malformation

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INTRODUCTION

Ischaemic colitis (IC) is the most prevalent type of gastrointestinal ischaemia (50–60% of all episodes). Its real incidence is probably underestimated because many patients suffer only mild or transient damage that remains undiagnosed. IC has a female gender predilection and mostly affects elderly patients, with 90% of them being over 60 years of age. Patients with IC frequently present with co-morbid conditions.

The aetiologies of IC are numerous and lead to a diminished perfusion of the colon, which may result in reversible mucosal injury or even full-thickness necrosis. Fortunately, most cases (80–85%) only result in non-gangrenous segmental IC and resolve with medical management, without a specific cause being found.2

IC is frequently classified as occlusive or non-occlusive. Occlusive IC can not only be attributed to thrombosis or emboli in large vessels, numerous and various chronic diseases of small vessels, trauma, various surgical procedures and iatrogenic causes (endoscopy, colonoscopy, barium enema, etc.) but also to venous causes resulting from venous thrombosis or venous hypertension.2

Non-occlusive causes comprise numerous life-threatening situations resulting in systemic hypoperfusion, situations of colonic dilatation or obstruction, iatrogenic causes related to a long list of drugs and long-distance running.

We hereby report a typical but very unusual case of acute congestive IC related to the decompenation of a very uncommon arteriovenous malformation (AVM) in the inferior mesenteric artery (IMA) territory.

CLINICAL PRESENTATION

A 45-year-old male patient was admitted to the emergency department with a 24-h history of heavy abdominal pain. The patient had previously experienced a prolonged episode of constipation and had been treated with laxatives. He was extremely obese and had a previous history of severe ischaemic cardiopathy complicated by episodes of ventricular tachycardia and auricular fibrillation. At admission, the pain was focalized in the hypogastrium and the left iliac fossa. These areas were painful to palpation with rebound. The patient also presented a small ecchymotic umbilical hernia. He was apyretic and had recurrent episodes of abundant mucoid stools. There was no rectal bleeding or melena. Laboratory tests were unremarkable except for a mild inflammatory syndrome with a C-reactive protein protein level ranging from 92 mg l⁻¹ at admission to 215 mg l⁻¹ 4 days later.
INVESTIGATIONS AND IMAGING FINDINGS

An unenhanced abdominal multidetector CT (MDCT; not illustrated) scan was first performed because the patient was allergic to iodinated contrast media. A marked homogeneously circumferential hypodense thickening of a very long segment of the left colon extending from the descending colon to the rectosigmoid junction was found. Prominent vessels and massive fat stranding were depicted in the mesosigmoid fat. A small amount of ascites were present. There were no colonic diverticula and the diagnosis of colonic diverticulitis was excluded. The first retained diagnosis was IC or inflammatory colonic disease.

Careful optical colonoscopy (Figure 1) was performed the same day and demonstrated an extremely oedematous, cobblestone appearance of the colonic mucosa causing narrowing of the lumen, especially at the level of the descending colon. There was neither blood nor ulceration. Limited biopsies were performed because the mucosa bleed easily. Histopathology demonstrated congestive and oedematous colonic mucosa with no trace of acute or chronic inflammatory process and no sign of infection.

A complementary contrast-enhanced abdominal MDCT (64 row MDCT, 140 kV, auto mA modulation, 0.6 s/rot, 55 mm/rot, thickness 1.2 mm, pitch 1.375:1, reconstruction spacing 0.7 mm) was performed 48 h later after premedicating with oral steroid antiallergic (Figures 2 and 3). Enhancement of the vessels was suboptimal, probably because of the morbid obesity and cardiac insufficiency of the patient. Nevertheless, the homogeneously circumferential hypodense thickening of the left colon was confirmed and better imaged. There was a sharp cutoff of the thickening at the level of the left phrenicocolic ligament and massive congestive fat stranding of the mesosigmoid, the greater omentum and the umbilical hernia. The prominent vessels were identified as unusually large and serpiginous varicosities running not only through the mesosigmoid fat but also within the colonic wall, especially at the level of the phrenicocolic ligament.

These varicosities converged to a dilated vein running through the transverse mesocolon to penetrate the venous splenoportal confluence at a right angle. An accessory venous drainage also runs to the left renal vein. The inferior mesenteric vein was constitutionally absent.

Scrupulous analysis of volume rendering and maximum intensity projection clearly diagnosed an arteriovenous communication in the distal mesosigmoid (Figure 4). There was an abrupt increase in the caliber of the rectal vein just distal to the
malformation. Also, there was considerable increase in the cali-
ber of the IMA when compared with a view obtained 4 years
ago. A colour Doppler ultrasound (Figure 3b) confirmed major
varicosities not only along the thickened colonic wall but also
within the wall. The Doppler spectrum of these varicosities
showed a pulsatile arterial component, reinforcing the hypothe-
sis of an AVM.

TREATMENT
4 days after admission, a decision to first treat the patient with
selective embolization was proposed. Selective arteriography
(Figure 5) of the IMA first confirmed the diagnosis of an arterio-
capillary malformation with a rather low flow. Careful emboliza-
tion with ethylene vinyl alcohol copolymer (ONYX®, Micro Therapeutic Inc., Irvine, CA) was performed
(white arrows on d and e). Nevertheless, 3 days later, impor-
tant symptoms persisted. A new angio-CT scan with volume
rendering views (e) confirmed the persistence of an active
arteriovenous fistula (white arrowheads).

OUTCOME AND FOLLOW-UP
Gross anatomy of the resected specimen (Figure 6) confirmed
the extremely diffuse congestion not only of the sigmoid but also
of the entire mesosigmoid and the epiploic appendages. A trans-
verse section through the specimen and histopathology showed
congestion of the mesocolon with varicosities and major submu-
osal oedema with preservation of the mucosa itself. The post-
operative period was uneventful and the protective ileostomy
was closed 2 months later.

DISCUSSION
AVMs and arteriovenous fistulas (AVFs) of the gastrointesti-
nal tract are often grouped together in clinical studies because
their physiologic consequences are frequently exactly the
same. However, there are important distinctions.7 Primary
mesenteric AVMs are congenital and/or idiopathic and differ
from secondary or acquired AVFs that are commonly caused
by blunt or penetrating trauma (bullet or knife) or have iatro-
genic aetiologies.1,3–5

In the splanchnic network, AVMs and AVFs are rather rare
(about 200 cases reported). The involved arteries in decreasing
appearance evoking venous ischaemia. The mesocolon and
omental appendages were bloated. A complete resection of the
left descending and sigmoid colon, and upper rectum was per-
formed and completed by loop ileostomy for protection of the
low colorectal anastomosis.
AVMs or fistulas between the IMA and venous network recently identified only 26 primary or secondary cases described in the literature. In an extensive review, Athanasiou et al. recently identified only 26 primary or secondary cases described in the literature.

Except for rare cases related to penetrating trauma, secondary fistulas (AVFs) in the IMA territory usually occur following colonic surgery or during arterial catheterization. Transfixion sutures or inadvertent ligation simultaneously passing through IMA branches and veins may cause fistulas during sigmoidectomy or left hemicolectomy. Similarly, the rupture of a congenital arterial aneurysm in very close anatomic relation with a draining vein or veins. In an extensive review, Athanasiou et al. recently identified only 26 primary or secondary cases described in the literature.

In the reported case, the congenital origin of the AVM was the most probable cause. Indeed, the anamnesis of our patient was absolutely free of any previous surgical procedure or trauma. Moreover, embolization failed to successfully resolve the entity probably because multiple arteriovenous connections were present. The congenital nature of the AVM of our patient was also reinforced by the simultaneous presence of anatomic variants comprising agenesis of the inferior mesenteric vein and the presence of an exclusively mesenteric origin of the hepatic artery. Congenital malformations also often result in the formation of multiple fistulas (classical components of Osler–Weber–Rendu and Ehlers–Danlos syndromes).

The cause of the acute clinical decompensation of the AVM in our patient is not clear. A possible worsening of cardiac fibrillation or ischaemic cardiopathy could have promoted the degradation. The history of our patient is also consistent with the classical natural history of AVMs, which, although they are congenital, do not usually clinically reveal until adulthood.

The vast majority of patients with inferior mesenteric AVMs present with symptoms of portal hypertension such as variceal bleeding or ascites. Less commonly, they also present with signs of congestive IC such as abdominal pain, diarrhea and haematochezia. However, some patients may remain asymptomatic, with only incidental detection on imaging. The intensity of symptoms is extremely variable. These symptoms are flow dependant and may range from minimal signs to severe heart failure related to left-to-right shunt.

Congestive IC is one of the most serious complications (50% of cases) of inferior mesenteric AVM or AVF. The clinical classic triad includes diarrhoea, haematochezia (not present in our patient) and abdominal pain. Guidelines for the diagnostic management of IC have recently been published. A combination of modalities comprising essentially MDCT—the cardinal imaging modality for patients with suspected IC—followed by colonoscopy with biopsies for a more specific diagnosis in suspected cases constitute the recommended diagnostic sequence. Contrast-enhanced MDCT assesses the distribution and phase of IC. Secondary colonoscopy with minimal insufflation follows, except in cases of gangrene, irreversible ischaemic damage or perforitis. A recent study recently concluded that MRI, only using pre-contrast images, could be used as a valid substitute for more invasive procedures in the diagnosis and follow-up of acute IC.

Wall thickening is the most common CT manifestation of IC. The wall is usually hypodense from mural oedema. Nevertheless, wall thickening is a non-specific sign and there is a wide degree of overlap with other colonic diseases such as inflammatory bowel disease and infectious colitis. Mesenteric fat stranding and ascites are also common but non-specific signs. When ischaemia progresses to infarction, free perforation, peritonitis, pneumato-sis, dilatation of the colon and portal or mesenteric gas may be seen. Direct colonoscopic visualization of a cyanotic bowel wall with submucosal oedema or haemorrhage, necrotic mucosa with ulcers, or infarction can confirm the diagnosis.

The mechanism of congestive IC in the IMA territory may be caused by the variable combination of hypoperfusion of the mucosa—owing to a steal phenomenon during which the arterial blood flow through AVM bypasses the capillary bed of the rectosigmoid—and congestive submucosal oedema due to venous hypertension—clearly diagnosed during sigmoidoscopy and confirmed by CT in our case. Acute congestive IC directly related to an AVM of the IMA territory as presented here is a very uncommon event.

The congested viscera and, especially, the sigmoid colon can constitute a painful clinically palpable mass. This condition was not found in our very obese patient. Nevertheless, the
sigmoid appeared extremely painful and incompressible during ultrasound examination.

Lower gastrointestinal bleeding can result from congestion of the bowel mucosa, direct fistula rupture within the lumen, necrosis owing to IC or bleeding haemorrhoids. Upper gastrointestinal bleeding resulting from oesophageal varices is rarer.

The treatment of inferior mesenteric AVMs or AVFs is complex and needs co-operation of the medical, radiological and surgical teams because the treatment needs case-specific solutions. Embolization is considered less invasive and potentially safer.\(^5\) Percutaneous endovascular arterial embolization of the feeding artery at the artery–venous junction is the technique of choice.\(^6\) It is extremely effective and has a low risk of complications in moderate-flow fistulae. Complications not only comprise serious IC but also recurrence, especially if there is more than one feeding artery. Embolization is not recommended in fistulas with large vessels because of the increased risk of extensive arterial thrombosis and ischaemia. Migration of the coils into the portal system has also been described for blood vessels of diameter \(>8\) mm at high flow rates. Surgery would appear preferable in these situations. In our patient, two attempts at embolization failed to resolve the symptoms and extensive surgery was necessary.

**DIFFERENTIAL DIAGNOSIS**

The first retained diagnosis after emergency unenhanced abdominal MDCT was IC or inflammatory colonic disease. Colonic diverticulitis was immediately excluded because of the unusual length of the affected colonic segment and the absence of colonic diverticula. Drug-induced colitis was excluded by the anamnesis. Histopathology of the biopsies performed during optical colonoscopy confirmed a congestive and oedematous colonic mucosa with no trace of acute or chronic inflammatory process (ulcerative colitis or Crohn’s disease) and no sign of infection. Finally, contrast-enhanced abdominal MDCT and colour Doppler ultrasound were decisive for the diagnosis of the AVM causing congestive IC.

**LEARNING POINTS**

1. The precise aetiology of IC remains unclear in many clinical cases. Numerous occlusive or non-occlusive causes exist. In the group of occlusive aetiologies, congestive ischaemia resulting from venous congestion owing to an AVM or fistula is extremely unusual.
2. Only 26 cases of AVMs and fistulas in the territory of the IMA have been reported until 2014. IC is the most common complication with a prevalence of 50%. The classic clinical triad includes diarrhoea, haematochezia and abdominal pain.
3. Fistulas essentially develop after a surgical or an interventional radiology procedure, or trauma. On the contrary, AVMs are congenital but classically remain clinically silent until adulthood.
4. The combination of abdominal MDCT and focalized colour Doppler ultrasound was effective for the diagnosis in the reported case.
5. The treatment of these vascular entities is complex and needs case-specific solutions. Co-operation of the medical, radiological and surgical teams is then required.

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