Imaging Findings of the Unusual Presentations, Associations and Clinical Mimics of Acute Appendicitis

Akut Apandisitin Alışılmadık Sunum, Birliktelik ve Klinik Taklitlerinin Görüntüleme Bulguları

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ABSTRACT

There are many kinds of unusual presentations or associations and clinical mimics of acute appendicitis, and definitive diagnosis requires knowledge of the imaging findings in some cases. The unusual presentations and associations of acute appendicitis included in this study are perforated appendicitis, acute appendicitis occurring in hernias, acute appendicitis with cystic endosalpingiosis, intussusception of appendix, and acute appendicitis with pregnancy. We also present uncommon gastrointestinal, urinary and gynecologic clinical mimics of acute appendicitis including anomalous congenital band, duplication cysts, giant Meckel’s diverticulitis, inflammatory fibroid polyp, renal artery thrombosis, spontaneous urinary extravasation and OHVIRA syndrome. Familiarity with these entities may improve diagnostic accuracy and enable the quickest and most appropriate clinical management.

Keywords: Abdomen, acute appendicitis, mimics, computed tomography, magnetic resonance imaging, diagnosis

ÖZ

Akut apandisitte olağandışı sunumlar veya birliktelikler ve klinik taklitlerin birçok türü vardır ve kesin teşhis bazı odağında görüntüleme bulguları hakkında bilgi gerektirir. Bu çalışmada perfor akut apandisit, hernilerde akut apandisit, kistik endosalpingiosis akut apandisit, apandisit anomalous congenital band, inflamatuar fibroid polyp, renal arter trombozu, spontan üriner ekstravazasyon ve OHVIRA sendromu dahil olmak üzere nadir görülen gastrointestinal, idrar ve jinekolojik klinik akut apandisit taklitlerini sunuyoruz. Bu varlıklara aşinalık tanısal doğruluğu artırabilir ve en hızlı ve en uygun klinik tedaviyi sağlayabilir.

Anahtar Kelimeler: Karın, akut apandisit, taklitleri, bilgisayarlı tomografi, manyetik rezonans görüntüleme, tanı

Introduction

Acute appendicitis (AA) is one of the most common abdominal emergencies. It is defined as an inflammation of the inner lining of the vermiform appendix that spreads to its other parts. It is usually caused by obstruction of the appendicular lumen from a variety of causes such as appendicolith, lymphoid hyperplasia of the appendix wall, foreign bodies, parasites, neoplasia, and metastasis. The diagnosis is often made based on clinical, laboratory, and cross-sectional imaging findings. The main imaging findings include the presence of a dilated, thick-walled, blind-ending, tubular structure with a diameter exceeding 6 mm, periappendiceal inflammation, and prominent mucosal enhancement, with or without an appendicolith [1]. However, the definitive diagnosis of the disease can be challenging in unusual presentations or associations, and there are many rare abdominal and pelvic diseases that can mimic AA, complicating its diagnosis [2].

In this pictorial essay, we present a few examples of unusual presentations or associations, and clinical mimics of the disease from our archive with a brief review to familiarize the readers with these imaging appearances. It should be noted that there are many other acute appendicitis mimics that are not mentioned in this text, such as appendicitis with tumors, ovarian pathologies, or gastrointestinal system infections.

Normal anatomy of the appendix

The appendix is derived from the cecal diverticulum as a diverticular outpouching on the antimesenteric side of the caecal midgut loop during the six weeks of gestation. Since the appendix grows in length, but its diameter does not grow as fast as the cecum, it is long and worm-shaped,
or vermiform at birth. Although the human appendix arises from the medial-posterior end of the cecum as a thin tube, the position of the appendix can be varied such as retrocecal, retrocolic, or pelvic. The appendix averages 6-8 cm in length, but can have a variable length. It is supplied by the appendicular artery, a terminal branch of the ileocolic artery from the superior mesenteric artery, and the venous blood drains through the correspondent veins into the superior mesenteric vein. Lymphatic drainage is into the ileocolic lymph nodes along the course of superior mesenteric artery [3].

**Normal imaging appearance**

Ultrasonography, which is a valuable first pass modality for evaluation of the appendix, reveals the organ as a compressible tubular structure. Multidetector computed tomography with multiplanar reformatted images with or without intravenous or oral contrast material is used at many institutions as the initial tool for evaluation of abdominal pain, which may lead to the detection of various pathologies including appendix. When needed, magnetic resonance imaging should be performed with a 1.5 T or a great unit and protocols should include diffusion-weighted imaging and dynamic contrast-enhanced imaging.

On imaging studies, the three anatomic parts of the appendix should be visualized as a blind-ending tubular structure with thin walls that measures less than 6-mm wall-to-wall diameter. Wall thickness is normally less than 2 mm. The base is attached to the wall of cecum about 2 cm below the ileocecal valve. The body is a thin, tubular part between the base and the tip, which is the distal blind end. The appendix may be filled with feces, air, or contrast material (Figure 1) [4].

**Unusual Presentations and Associations**

**Perforated appendicitis**

The diagnosis of perforated appendix is crucial as perforation increases the risk of complications after surgery. Defects in the appendiceal wall, extraluminal air locules or free intraperitoneal air, localised right iliac fossa abscess or phlegmon, and appendicolith outside the appendix or within the right iliac fossa abscess are the main imaging findings of this increasingly rare complication (Figure 2) [5]. It should be mentioned that the appendix may still be ruptured without these imaging findings (Figure 3). Perforated appendicitis may cause complications like abscess formation and peritonitis. The development of peritonitis secondary to perforation is more frequent in children due to rapid progression from inflammation to wall rupture. However, in adults, the inflammatory adhesions developing around the site of inflammation more frequently cause phlegmons and abscesses instead of a rapid evolution in peritonitis. Pylephlebitis and pylethrombosis, hydroureteronephrosis; gangrenous appendicitis, dynamic or mechanical bowel obstruction, and fistula with other contiguous organs such as bladder, vagina, uterus, and skin are the other complications that may be detected in the abdominal cavity following appendix perforation [6].

**Acute appendicitis occurring in hernias**

Acute appendicitis may occur in any type of abdominal hernia including the inguinal hernia sac (Amyand’s hernia), the femoral hernia sac (De Garengneot’s hernia), obturator hernia, umbilical hernia, Spigelian hernia, laparoscopic port site hernia, and incisional site hernia. An appendix incarcerated within a hernia makes it vulnerable to trauma and adhesions, further restricting it from sliding back into the abdominal cavity and increasing the risk of inflammation. Acute hernial appendicitis usually creates a diagnostic problem prior to surgery and most often the diagnosis is incarcerated or strangulated hernia. Other considerations may vary due to the type of hernia and may include variable pathologies such as Richter’s hernia, orchitis, omentocele, inguinal lymphadenitis, epididymitis, and hemorrhagic testicular
tumor. Multidetector CT and multiplanar MRI are excellent imaging modalities for elucidating a blind-ending tubular structure arising from the caecum that extends into the hernia sac (Figure 4) [7].

Acute appendicitis with cystic endosalpingiosis

Endosalpingiosis is defined as ectopic tubal epithelium. It usually occurs in the pelvic and abdominal peritoneum and may rarely involve the serosal surface of the vermiform appendix. Cystic endosalpingiosis is usually seen as a multilocular septated cystic mass in imaging studies. The walls of the cystic mass and septal structure may show contrast enhancement mimicking a tumor. Endosalpingiosis cannot cause pain. The association of acute appendicitis and cystic endosalpingiosis is incidental and it must be considered in the differential diagnosis of cystic appendiceal tumors (Figure 5). It is commonly diagnosed through histological examinations [8].

Intussusception of the appendix

Intussusception of the appendix is a rare disease that constitutes a diagnostic challenge. It may mimic acute appendicitis, may present with typical symptoms of intussusception, or may be totally asymptomatic. There may be partial invagination to caecum or the whole colon may be involved, and, furthermore, the appendix may be totally or partially inverted with or without a lead point. Physiological and anatomical changes associated with pregnancy may obscure or delay the correct diagnosis of AA. Abdominal ultrasonography has a high rate of non-visualisation of the appendix in gravid patients, and CT presents a potential hazard to the developing fetus due to ionizing radiation. However, MR imaging of the appendix is the safe and preferred modality when appendicitis is suspected in pregnant women. MR imaging findings of appendicitis include an appendiceal diameter greater than 7 mm, an appendiceal wall thickness greater than 2 mm, high-signal-intensity luminal contents on T2-weighted images, hyperintense periappendiceal fat stranding and fluid, and restricted diffusion in diffusion-weighted image (Figure 7) [11].

Rare Clinical Mimics

Anomalous congenital band

Congenital bands of the abdomen are a rare cause of acute abdomen in children and extremely rare in adults. Its location is variable and may be found between the bowel/mesentery/abdominal wall/liver and intraabdominal...
ligaments. The etiology of these bands remains unclear, since its location is not similar to that of remnants. Although they usually cause bowel obstruction symptoms due to compression or entrapment of a bowel, the presentation may suggest mesenteric infarction, perforated duodenal infarction, diverticulitis, cholecystitis, strangulated hernia, and AA. Although it is difficult to establish a preoperative imaging diagnosis, a congenital band should also be included in the differential diagnosis of patients with symptoms and signs of AA in the absence of previous surgery, which excludes postoperative intraabdominal adhesions or bands (Figure 8) [12].

Duplication cysts
Alimentary tract duplication cysts are uncommon congenital anomalies containing a normal gastrointestinal mucosa and smooth muscle layer. Duplications have two types, either cystic or tubular attached to the gastrointestinal tract, and they share the same blood supply. Clinical symptoms may vary depending on their type, site, and size and may include pain, distension, palpable mass, vomiting, and bleeding. They may also present with complications such as perforation, intussusception, bowel obstruction, and volvulus. Duodenal duplication cysts and tubular jejunal duplication cysts, which directly communicate with the bowel lumen, are extremely rare entities causing acute right lower abdominal pain mimicking AA in adults (Figure 9, 10) [13].

Meckel’s diverticulitis
A Meckel diverticulum is a vestigial remnant of the omphalomesenteric (vitellointestinal) duct that communicates between the yolk sac and midgut lumen of the developing fetus. Thus, it is a true diverticulum that includes all three coats of the small intestine. It may range from 1 to 12 cm in length and is found at an average distance of 60 cm from the ileo-cecal valve. They may include embryonic remnants such as ectopic gastric mucosa and pancreatic tissue. The inflamed Meckel’s diverticulum is usually seen as a blind-ending pouch of variable size and mural thickness that arises from the antimesenteric side of the distal ileum with surrounding mesenteric inflammation in imaging studies. The location of the diverticulum may vary from the right lower quadrant to the mid abdomen. It may occur with hemorrhagic, mechanical, infectious, or tumoral complications. The diagnosis is most difficult in the setting of secondary intestinal obstruction (Figure 11) [14].

Inflammatory fibroid poly
Inflammatory fibroid tumor or Vanek’s tumor of the ileum may simulate clinical findings of acute appendicitis, but imaging findings easily rule that out. It is a rare benign lesion of the gastrointestinal tract, and the most common location is the antrum of the stomach. Clinical presentation varies by location and size, and an ileal location can present with abdominal pain, lower gastrointestinal bleeding, anemia, and (rarely) small bowel obstruction due to intussusception. Determination of an intraluminal mass between 2 and 5 cm with intussusception and mechanical intestinal obstruction is the key finding for preoperative diagnosis on imaging (Figure 12) [15].

Spontaneous urinary extravasation
Spontaneous urinary extravasation is defined as a non-traumatic urinary leakage from the collecting system due to an excessive sudden increase in the intraluminal pressure as a result of obstruction and may be together with perirenal and retroperitoneal urinoma formation. It is an uncommon complication of obstructive uropathy and usually results from a stone in the uretero-vesical junction.
Other causes may include extrinsic ureteral compression by tumors, pelviureteric junction obstruction, vesicoureteric junction obstruction, instrumentation, and trauma. Clinical presentation may range from mild flank pain, nausea, and vomiting to acute abdomen, and symptoms may mimic pyelonephritis, duodenal ulcer, biliary colic, cholecystitis, and appendicitis. The most useful imaging modality to identify spontaneous urinary extravasation is abdominal computed tomography. Delayed images usually show extravasation of the contrast medium and provide information regarding the perforation site (Figure 13) [16].

Renal artery thrombosis and renal infarction
Acute renal infarction due to right renal artery thrombosis is rare and usually present with abrupt flank or abdominal pain accompanied by nausea, vomiting, and fever. The laboratory findings include leukocytosis besides hematuria, proteinuria, and elevated lactate dehydrogenase. The differential diagnosis of the disease is extensive, and emergent surgical or nonsurgical conditions causing acute abdominal pain such as AA should be ruled out [17]. A contrast-enhanced CT is the best way to recognize the disease (Figure 14).

In conclusion, we have briefly reviewed the imaging appearances of unusual presentations and coexisting pathologies of AA together with its uncommon clinical mimics. In patients with suspected AA, clinicians and radiologists should remain vigilant for rare presentations, associations and mimics in order to make a prompt correct diagnosis and to avoid unnecessary or complicated surgical interventions.

OHVIRA Syndrome
Uterus didelphys with obstructed hemivagina and ipsilateral renal agenesis (OHVIRA syndrome) is a very uncommon developmental urogenital malformation. It results from complete failure of fusion of the müllerian ducts and their normal differentiation to form a cervix and uterus during the 8th week of gestation. It usually presents after menarche with remittent pelvic pain and a palpable pelvic mass due to hematocolpos. It may present with acute severe abdominal pain in the right lower quadrant of the abdomen [18]. Magnetic resonance imaging is the best imaging modality to confirm the diagnosis and provides excellent images demonstrating iso/high T1W signal and high T2W signal that indicates pelvic fluid collection contiguous with the endocervix along with didelphic uterus and an absent kidney on the affected side (Figure 15).
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