

The Ultrasonographic Findings of Appendiceal Diseases: Benign and Malignant

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Abstract: The appendix is a small pouch-like sac with no fixed anatomical position, narrowing entrance to caecum predisposes to both benign and malignant obstruction in nature. In recent years, ultrasonography plays an important part in diagnosis of appendiceal diseases. This article aims to describe ultrasonographic assessment of different kinds of appendiceal diseases. We provide an overview of the literature about the ultrasonographic features of appendiceal diseases.

Key words: Appendix; Ultrasound; Appendicitis; Appendiceal neoplasms

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The appendix is a small pouch-like sac that forms a long blind-ending off the cecum below the ileocecal valve. There is no fixed anatomical location about the tip of the appendix. Retrocaecal appendix is considered to be predominant among adults [1]. The function of the appendix remains unknown. It was once thought to be a degenerating structure that helps digestion. However, recent theories suggested that the appendix was a "safe house" for commensal bacteria and had a role in regulating the homeostasis of the intestinal lumen [2,3]. The narrow entrance of the appendix to caecum is prone to both benign and malignant obstruction in nature. Improvement in modern ultrasonographic techniques breaks barriers and presents ultrasonography as the most common and widespread tool in both areas of screening and diagnosis of a variety of abdominal diseases. Here, we are aimed to reveal the ultrasonic features of appendiceal diseases.

Normal appendix

In an ultrasound examination, it may not be easy to identify a normal appendix. The appendix layer from the inside to the outside appears as a linear hyperechoic luminal surface, a hypoechoic mucosa, a hyperechoic submucosa, a hypoechoic muscular lamina propria, and an echogenic serosa layer (Fig. 1) [4]. The findings of

normal appendix vary depending on the contents of the cavity and the presence or absence of lymphoid tissue proliferation. Normal cavity contents include residual excreta, liquids and gases. On ultrasound, a central shadowing echogenicity with acoustic shadows may indicate fecalith [5]. Moreover, the intestinal gas of the appendix appears hyperechoic and acoustic shadows can be found later [4]. The fluid in the cavity usually exhibits

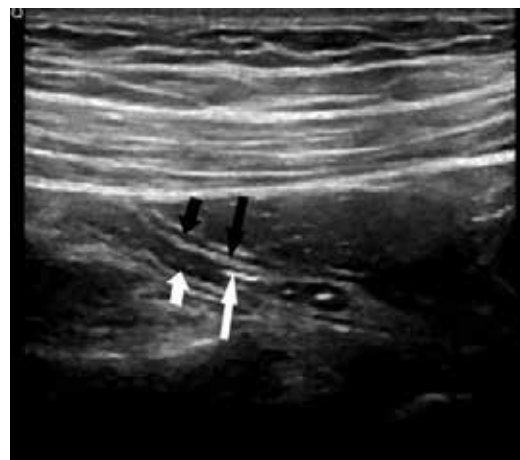


Figure 1 Layers of normal appendiceal wall. Long white arrow and long black arrow indicated echogenic luminal interface and echogenic submucosal layer. Short white arrow and short black arrow indicated hypoechoic mucosa and muscularis propria respectively.

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anechoic.

The normal appendix is easily compressed. A gentle pressure compressed by an ultrasonic transducer on the abdominal wall, the circular appendix is transformed into an oval shape in a transverse plane. This phenomenon is called graded compression. The maximum outer diameter of the normal appendix measured in cross section is typically no more than 5 mm. However, the maximum diameter may increase to more than 6 to 7 mm when the appendix is dilated by retained excretion or due to lymphatic tissue hyperplasia. At this point, the swollen appendix should be distinguished from appendicitis.

Appendicitis

Normal adult appendicitis

Nearly 50%–80% of cases of acute appendicitis caused by luminal obstruction and mucosal infiltration are due to bacteria in the cavity [6]. However, approximately one-third of patients with appendicitis do not show typical signs and symptoms [7–9].

Ultrasound has a sensitivity of 78% to 83% for acute appendicitis and a specificity of 83% to 93% [10,11]. In the 1980s, ultrasound graded compression technique for the diagnosis of appendicitis was firstly proposed [12]. This technique requires the examiner to apply a

slight force with the transducer to move the pneumatized intestine that normally covers the actual appearance of the abdominal cavity.

The appendix may be an immobile, incompressible, and dilated tubular structure in the right lower abdomen. The diameter of the inflamed appendix is the most important indicator of appendicitis. The examiner should note that measurements should be taken when the appendix is compressed. However, the traditional threshold of 6 mm is still controversial. One study showed that 23% of normal appendices were greater than 6 mm [13]. Therefore, some agencies used 7 mm as the diagnosis threshold, which seemed more appropriate in recent research [14]. In the transverse view, the inflamed appendix was rounded, causing a "target" sign. In addition, patients may have focal tenderness at the point of compression of the ultrasound probe, showing a positive "sonographic McBurney's sign" [15]. Imaging of inflammatory adipose tissue around the appendix can be used to diagnose appendicitis. It may appear as an echogenic and incompressible area. Perforation as a complication is determined by the presence of fluid or abscess around the appendix. In addition, a study reported that the loss of normal echogenic submucosa was the most useful feature to distinguish complicated appendicitis from uncomplicated appendicitis [16] (Fig. 2).

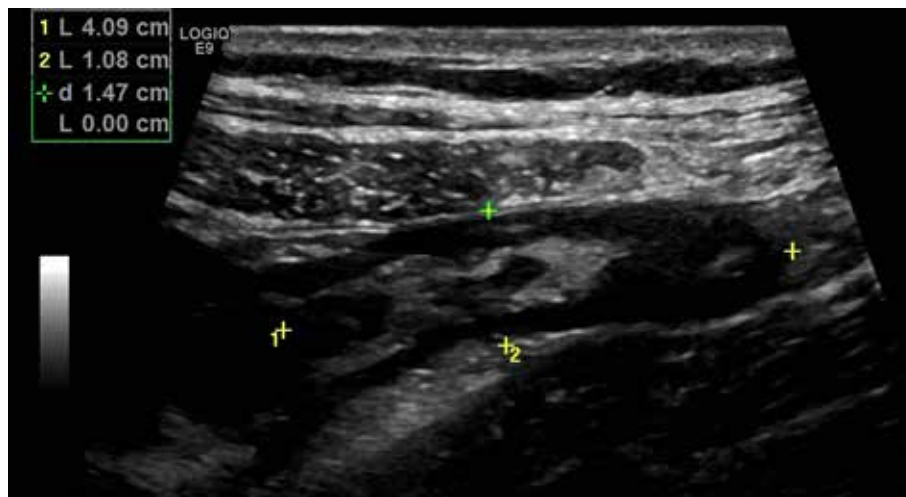


Figure 2 Sonogram from a 56-year-old male patient with appendicitis without perforation showing the loss of the submucosal layer of the appendix.

When the diameter is ambiguous, an increase in the vascular distribution of the appendix in the Doppler examination may be helpful in diagnosing appendicitis. However, when the appendix is gangrenous or partially necrotic, it may be found that the blood vessel distribution is reduced [14].

Appendicitis in pregnancy

Pregnancy may cause acute appendicitis and

therefore appendicitis becomes a common disease in the emergency department [17]. Because there is no radiation, ultrasound is considered the first choice of screening and diagnosis of appendicitis in pregnant women. Diagnostic parameters such as sensitivity and specificity ranged from 67% to 100% and 83.3% to 96%, respectively [18]. Although ultrasound plays an important role in the diagnosis of appendicitis in the

first trimester and the second trimester, the diagnosis in the third trimester is not the case. Due to anatomical changes, the results are unsatisfactory [17]. By gradually enlarging uterus, the appendix moves from the right lower abdomen to the upper right abdomen. As a result, migration pain may shift to the upper right quadrant. However, local tenderness caused by inflamed peritoneal irritation is no longer evident.

As mentioned above, graded compression ultrasound is of great value in the identification of acute appendicitis. However, this technique is difficult to perform in pregnant patients who are pregnant in second and third trimester in the supine position. Because the enlarged uterus does not allow enough compression. The examiner should scan the patient in the left or posterior oblique position instead of the supine position [19]. Typically, the appendix presents an incompressible blind-ended tubular structure in the right abdomen with a maximum diameter of more than 6 mm. Characteristics of appendicitis, such as fat stranding, fluid collection or extraluminal air, constitute evidences of appendicitis [20–22].

Pediatric appendicitis

Common signs and symptoms of appendicitis include fever, right lower quadrant abdominal pain, rebound tenderness, nausea, vomiting or diarrhea [23]. Less than 50% of children with appendicitis may have typical signs and symptoms [24,25], which are more difficult for young children to recognize. Ultrasonography is an important imaging tool for the diagnosis of appendicitis in children without severe disease. Grade compression techniques are also suitable for pediatric examinations. But the examiner may be in a dilemma because the child may not be able to tolerate the discomfort caused by the oppression. Additional techniques such as manual compression may be helpful [26]. For obese children, it is difficult to see normal appendix. Hörmann et al. [27] found that the sensitivity of overweight children's appendix visualization was 21%. The use of low frequency transducers enhances the sensitivity of obese children.

Ultrasound findings in children with appendicitis are similar to those in adults. The maximum outer diameter is an important indicator of appendicitis in children. Studies have shown that the accuracy of ultrasonography is 85% when ultrasound examiner used 6 mm as the minimum outer diameter in children with abnormal appendix [28].

However, the maximum outer diameter of the normal appendix can be increased to 14 mm in children with cystic fibrosis [29,30]. Inflamed appendix is always enlarged and incompressible. In addition, secondary

mesenteric fat, fluid and other secondary ultrasound signs are collected as auxiliary markers in the diagnosis process [28].

Appendiceal neoplasms

Carcinoid tumors

Carcinoid tumors are neuroendocrine tumors derived from Kulchitsky or enterochromaffin cells. Carcinoid tumors secrete serotonin, which can cause typical carcinoid symptoms such as flushing and diarrhea [31]. The incidence of carcinoids in the appendix ranges from 33% to 50% of all appendix tumors [32,33]. The incidence of carcinoid cancer in women is twice than that of men [34]. Many patients have clinical features suggesting acute appendicitis. Most appendix carcinoids tumors are accidentally discovered during surgery [31]. Carcinoid tumors may be malignant. Metastasis from the typical carcinoid tumors of the appendix is very rare [35].

On ultrasound, carcinoid tumors tend to be a small oval mass. Carcinoid tumors may block the appendix tip and have ultrasound features that mimic appendicitis [36]. When the tumor reaches a sufficient size or demonstrates calcification, the tumor itself may be visualized during scanning [37].

Mucinous appendiceal neoplasms

Appendicular mucinous tumors refer to a range of tumors, from simple mucinous cysts to complex pseudomyxoma peritonei [38]. The classification of such tumors is complex and controversial. According to the WHO 2010 classification of mucinous tumors, there are three main categories: 1) Mucinous adenoma, 2) low-grade appendicular mucinous neoplasm (LAMN) and 3) appendiceal adenocarcinoma [39].

On ultrasound, mucinous tumors present with dilated cystic lesions with or without mural calcification [15]. The thin-walled onion skin sign at the appendix is the specific appearance of the appendix mucocele [40]. Pseudomyxoma peritonei is usually the secondary appearance of a perforated mucinous appendicular tumor. Encapsulated lesions were found in the peritoneum. Most studies have reported a summary of typical ultrasonographic findings of pseudomyxoma peritonei, including scalloping of the liver margin and ascitic separation [41]. However, Tsai [42] showed that pseudomyxoma peritonei could present as cystic masses with hypoechoic walls and an anechoic interior within the abdominal cavity. Another study showed calcification in the pelvis and upper abdominal cavity [43].

Lymphoma

Although extra-hospital non-Hodgkin's lymphoma is

usually located in the gastrointestinal tract, appendicular lymphoma is rare. The proportion of lymphoma found in all appendectomy specimens was reported to be 0.0015% [44]. The most common clinical feature is acute tenderness in the right abdomen, which would possibly mimic appendicitis. On ultrasound, diffused thickened murals appear as hypoechoic. Appendicular lymphomas often present with enlargement of the appendix and keep its vermiform appearance [45].

Endometriosis

Endometriosis is a disease caused by migration and implantation of endometrial cells on other organs outside the uterine cavity, e.g., the ovaries, fallopian tubes and external uterus. Endometriosis can also be found in the gastrointestinal tract. The incidence of endometriosis affecting the appendix accounts for approximately 3% of all cases [46]. It is believed that this disease does not occur in isolation but is usually accompanied by ovarian involvement [47]. Endometriosis can mimic other appendicular disease on ultrasound, but a non-vascular echogenic mass can make it stand out from the other kinds of diseases [15].

Conclusion

Although appendicitis is the most common appendiceal pathology, there are also other appendicular diseases. The corresponding clinical signs and symptoms are usually atypical, but can mimic acute appendicitis or other pathologies. Ultrasound is superior to other imaging modalities because it does not require patient preparation and is non-invasive, cost-effective, safe and repeatable. Sonographers should be aware of these diseases and associated ultrasound findings in order to make a definitive diagnosis.

Conflicts of Interest

The authors report no conflict of interest in this work.

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