Giant appendix or an appendiceal mucocele? 
Case report of an 11-year-old child

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Abstract: We present an 11-year-old male child with an enormous appendix that was regarded as an appendiceal mucocele. The disorder is very rare and usually appears in middle aged patients. It is a clinical diagnosis. It could cause a variety of symptoms, especially, acute appendicitis and unidentified lesion in the right iliac fossa. According to the reasons, it could be just a curiosity without any relevancy or the sign of a malignant lesion with bad prognostic factors. The histopathological findings prove the origin.

Keywords: appendix, appendicitis, appendiceal mucocele

Introduction

The appendix extends from the caecum. Its length can range from 2 to 20 cm. Its usual diameter is about 7–8 mm [1, 2]. Its well-known disorder is acute appendicitis that could cause severe medical conditions [3, 4] and was described by Reginald Fitz in 1886 [5]. It occurs most frequently between the ages of 5 and 40 [6].

Appendiceal mucocele is a rare entity, the prevalence is about 0.2 to 0.3% of all appendectomies [7–14]. It was described by Rokitansky in 1842 [15]. This is a macroscopic clinical diagnosis. It is delineated as the dilatation of the appendix because of abnormal intraluminal accumulation of the mucus that results from the chronic obstruction of the appendiceal neck. It usually appears in middle aged patients. The prognosis of appendiceal mucocele depends on the histopathological findings. If the cause of the obstruction is bilestone, foreign body, or feces, it is just a clinical diagnosis and the disorder should be considered as benign. On the other hand, mucinous tumors (adenoma or adenocarcinoma) could also obstruct the lumen of the appendix, which condition is the pseudomyxoma peritonei. The disorder is lethal without treatment, because the enormous amount of gelatinous ascites results in the dysfunction of the abdominal organs. The prognosis with complex treatment could be favorable [7–17].

Case Report

An 11-year-old boy was admitted to our Department of Pediatrics because of typical clinical diagnosis of acute appendicitis with onset of symptoms one day earlier. The blood tests verified an increased serum level of white blood cells (23.9 g/L), and the other measured parameters were within normal ranges. The ultrasonography of the abdomen showed a tubular inflamed lesion in the right iliac fossa (Fig. 1).

Open surgery was performed under general anesthesia. During the exploration of the right lower abdomen, we found a lot of intraabdominal clear mucinous fluid and the lesion, that was similar to an inflamed small intestine. There were no signs of peritonitis. After raising the suspicious mass out of the abdominal cavity, we made sure of an appendiceal mucocele (Fig. 2). The length of the organ was 10 cm, and its diameter was between 1.3 and 2.5 cm. It was inflamed and suspicious of perforation. The intestines were untouched. Appendectomy was performed.
The child became asymptomatic briefly. The serum level of white blood cells fell to 11.1 g/L 2 days after the operation. On the third postoperative day, the boy was discharged and healed without complications.

The histopathological examination showed the acute phlegmono-gangrenous inflammation of the appendix without any exact sign or cause of the abnormal intraluminal accumulation of the mucus; however, the mucosa
Appendiceal mucocele of an 11-year-old child

Discussion

Appendiceal mucocele is a very rare condition and typical for the middle aged patients. It is a clinical diagnosis. The distension of the appendix is caused by the obliteration of the appendiceal neck. The reasons could be benign, but severe conditions caused by mucinous tumors may occur with low incidence. The prognosis of pseudomyxoma peritonei depends on the dignity. Primarily, surgery is necessary to avoid the compression of the intraabdominal organs. If the histopathological findings show mucinous adenocarcinoma, adjuvant chemotherapy has to be administered [16–21].

Appendiceal mucocele usually causes a variety of symptoms. Acute appendicitis and a mass in the right iliac fossa appear the most frequently. The diagnosis is sometimes difficult. The ultrasonography and computed tomography can verify the lesion. During the surgery, the clinical diagnosis can be established. The histopathological findings prove the origin [7–11].

In this case report, an 11-year-old child who underwent surgery because of the clinical diagnosis of acute appendicitis is presented. The radiologist should suspect that the tubular lesion is not the common appearance of this disorder. The mass in the right iliac fossa was not palpable because of the abdominal pain. There were no suspicious signs of an appendiceal mucocele before surgery. The disorder is very rare, especially in children, so the lesion found during the operation was not expected. The histopathological examination showed the acute phlegmono-gangrenous inflammation of the organ without the fact of mucus accumulation in the lumen and any exact histological cause of the obstruction of the appendix; however, the mucosa was necrotic and some segments seemed to be hyperlastic with mucinous retention, so the assessment could be difficult. This finding may correlate to the suspected perforation of the appendix that could explain the enormous amount of intraabdominal clear mucinous fluid without peritonitis and the lack of mucus accumulation in the lumen of the appendix. First and last, the pathologist did not regard the lesion on the dot as an appendiceal mucocele; however, it was the clinical diagnosis [8–12].

The case report indicates that the surgeon may discover an unidentified mass during surgery because of acute appendicitis in children, that can be an appendiceal mucocele; however, it is a really uncommon situation. There are only two publications available about appendiceal mucocele in children in which the cause of the obstruction was a benign mucinous tumor [22, 23].

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Fig. 3. Histological image of the appendiceal mucocele. Haematoxylin–eosin stain, magnification: ×60
accuracy of the data analysis. IBB: data collection and analysis, writing; MN: data collection; AK: data collection; OB: data collection and analysis; AR: data collection; LV: data collection.

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