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A rare cause of intussusception in children: Mucinous cystadenoma of the appendix

Çocuklarda invajinasyonun nadir bir nedeni: Apendiks müsinöz kistadenomu

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Summary

Although intussusception is a common cause of intestinal obstruction in children, intussusception of mucinous cystadenoma (MCA) of the appendix characterized by distension of the lumen due to accumulation of the mucoid substance is rarely reported. Diagnosis of MCA is usually made perioperatively. In these cases, as a general approach, an appendectomy is performed for cystadenoma, and right hemi-colectomy is recommended for cystadenocarcinoma. However, the type of surgical treatment is related to the dimensions and histology of the lesions. A 12-year-old boy was admitted to our clinic with symptoms of a mobile and painful mass in the right upper quadrant of the abdomen. Abdominal ultrasonography showed intussusception as well as a large, cystic and solid structure extending from cecum to transverse colon. Reduction of intussusception and right hemi-colectomy were performed subsequent to laparotomy. Pathologic diagnosis was reported as MCA. Although MCA is seen rarely, it always should be kept in mind in differential diagnosis of intussusception and intra abdominal mass, especially in children with abdominal pain and distension.

Key Words: Child, mucinous cystadenoma, intussusception, appendix, mucocele.

Özet

İnvajinasyon, çocuklarda bağırsak obstrüksiyonun yaygın bir nedeni olsa da, apendiksteki mukoid madde birikimi ile karakterize müsinöz kistadenoma bağlı invajinasyon (MCA) nadiren bildirilmektedir. MCA tanısı genellikle perioperatif yapılır. Bu tip olgularda, kistadenom için apendektomi ve kistadenokarsinom için sağ hemi-kolektomi, genel bir yaklaşım olarak önerilmektedir. Ancak cerrahi tedavinin şekli, lezyonun boyutu ve histoloji ile ilgilidir. Oniki yaşında erkek olgumuz karnın sağ üst bölgesinde ağrılı kitle şikayeti ile kliniğimize başvurdu. Batın ultrasonografisinde invajinasyon ile birlikte çekumdan transvers kolona kadar uzanan geniş kistik bir yapı saptandı. Laparotomiyi takiben invajinasyon redüksiyonu ve sağ hemi-kolektomi uygulandı. Patolojik tanı MCA olarak rapor edildi. MCA nadiren görülmesine rağmen özellikle karın ağrısı ve şişkinlik ile başvuran çocuklarda, invajinasyon ve intraabdominal kitle ayırıcı tanısında akılda tutulmalıdır.

Anahtar Sözcükler: *Çocuk, müsinöz kistadenom, invajinasyon, apendiks, mukosel.*

Introduction

Mucinous cystadenoma (MCA) is a rarely seen pathology of the appendix, characterized by a cystic dilatation of lumen with stasis of mucus. The incidence of this pathology changes between 0.2% and 0.3% in appendicectomy specimens (1).

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This tumor is mostly reported in elderly patients and extremely rare in children (2). Patients may present with various clinical signs and symptoms such as abdominal mass or cyst rupture. Rupture of the MCA may result in the clinical condition of pseudomyxoma peritonei. A correct diagnosis may help to avoid iatrogenic rupture during surgery for intussusception in children. In this report, a case of intussusception by a giant MCA of the appendix is discussed with relevant literature.

Case Report

A 12-year-old boy admitted to our clinic with symptoms of a painful abdominal mass and abdominal distension. In a detailed medical history, it was understood that he had intermittent right abdominal pain, nausea and anorexia in the previous week. His blood pressure was 120/70 mmHg with regular pulse of 88/min. He had a body temperature of 36.8 °C. On physical examination, although the abdomen was flat and soft, a mobile and painful mass was palpated in the right upper quadrant of the abdomen. Routine hematologic and biochemical analysis were within normal ranges. Abdominal plain graphy was normal. A Computed tomography (CT) scan of the abdomen showed a hypodense mass which was 9x8x9 cm in diameter. In the mass, solid and cystic structures extending from cecum to transverse colon were detected (Figure-1). Abdominal ultrasonography revealed that the intussusception extended from the cecum to the transverse colon. The other abdominal structures were completely normal. With these findings diagnostic laparoscopic intervention was planned. During the operation, firstly intussusception was reducted. A mass of about 10x5 cm dimension was determined in the cecum (Figure-2). The patient underwent a right hemi-colectomy and end to end anastomosis. The tumor weight was 245 grams. In the pathologic evaluation of the tumor, extensive mucin in the cystic cavity lined by tall columnar epithelium was seen. There was no evidence of malignancy. According to these findings the histopathological diagnosis was mucinous cystadenoma (Figure-3). The patient was discharged 5 days postoperatively without any complications. On follow up visits recurrence was not detected.

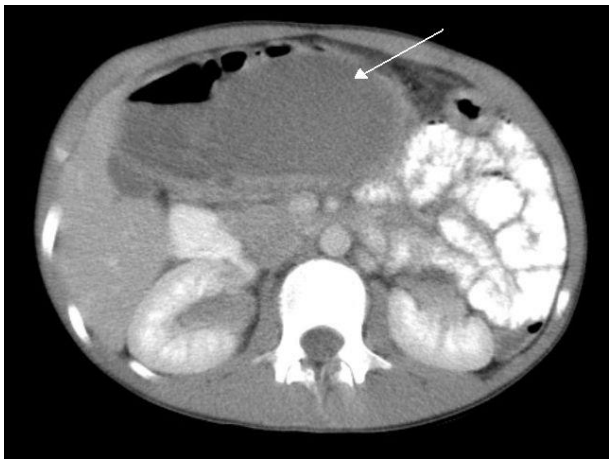

Figure-1. CT scan shows an oval, large and hypodense cystic mass (arrow) in the upper part of the cecum.

Discussion

The term of mucocele is often used as a general macroscopic descriptive term for dilatation of the appendiceal lumen by mucinous secretions (2-6). MCA is the most common form, accounting for 63% to 84% of mucocele cases (2,4,7). Mucinous cystadenoma is a tumor of the appendix associated with cystic dilatation (3,4). The incidence of this entity changes between 0.2 and 0.95% (5). It has been reported that MCA is seen more frequently in women and those over 50 years (1, 4, 6). On the contrary, MCA in children is rarely seen and frequently of ovarian origin (3).

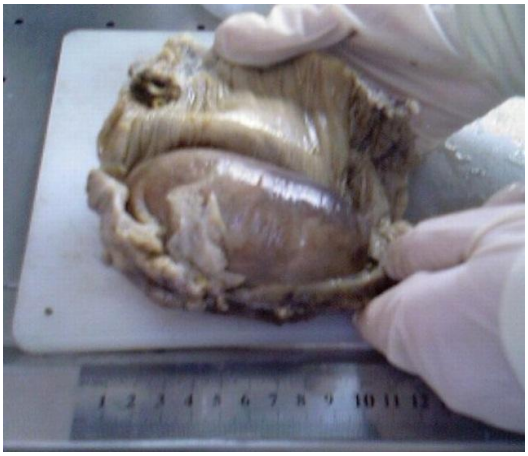

Figure-2. Macroscopic appearance of the mass.

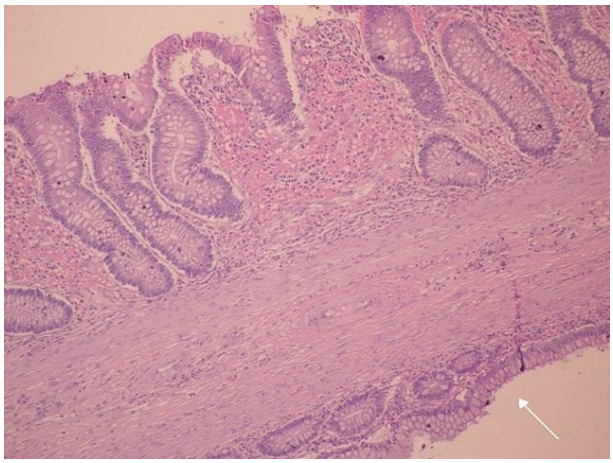

Figure-3. A histopathological view of the mass (Arrow: Mucinous cells layer) (Hematoxilen & Eosin, 25x).

Although the etiology of the MCA is not clear, its pathogenesis is well known. Epithelial tumors of the appendix may be classified as a mucinous or nonmucinous. Most of these tumors which are seen in the appendix are mucin rich, demonstrating circumferential mucosal involvement (7). Cystadenoma and mucinous cystadenoma are frequently used terms for benign neoplastic mucoceles. The MCA is a mucocele that

implies a dilated appendiceal lumen caused by an abnormal accumulation of mucus. According to characteristics of the epithelium, four subgroups of mucoceles were identified (2, 4, 7). Type-1 mucocele is known as simple mucocele (2-21%) characterized by normal epithelium, resulting from obstruction of the appendiceal outflow and luminal dilatation up to 1 cm. Type 2 mucocele (5-25%) is defined as hyperplastic epithelium where luminal dilatation is between 2 to 6 cm. Type 3 is defined as the benign mucocele (63-84%). Epithelial villous adenomatous changes with low-grade epithelial dysplasia are characterized by marked distention of the lumen up to 6 cm in this form. Our patient belongs to this group. Type-4 is malignant mucinous cystadenocarcinoma (11-20%). The luminal distention is usually severe. There is always the risk of rupture, either spontaneous or accidental (2, 4).

Patients can be asymptomatic or may present with abdominal pain, an abdominal mass, rectal bleeding or intussusception. As in our case, a palpable mass can be found in 50% of the cases (3, 5, 8). The intestinal obstruction is a rarely reported complication and frequently caused by intussusception (2, 3). There was no intestinal obstruction in the present patient.

Due to the nonspecific nature of this disease, preoperative diagnosis is difficult. The lesion may be identified by radiological and endoscopic imaging methods. In this context, a colonoscopy may show intraluminal lesions (2, 5). CT scan of the abdomen is important in the diagnosis of the disease. As in our case the typical finding in cystadenomas of the appendix is a round, thin-walled and encapsulated cystic mass associated with the cecum (2, 7). In addition, the wall of cystic mass is variable in thickness and has not been shown to correlate with malignancy (5, 8). Ultrasonography often shows a cystic and encapsulated lesion, firmly attached to the cecum, with liquid content and internally variable echogenicity. However, actual diagnosis is usually made during histopathologic examination of the excised specimen. In our patient, CT findings were similar to those reported in the literature. However ultrasonography showed intussusception in the transverse colon and there was no cystic lesion.

It has been recommended that all mucoceles should be removed, particularly those with a diameter greater than 2 cm (7, 8, 10). Treatment consists of aggressive surgical debulking of all apparent mucinous tissue. Since the laparoscopic approach can lead to rupture and pseudomyxoma peritonei, the standard treatment of MCA is exploration and appendectomy (9). A right hemicolectomy is performed when the cecum is involved (2, 4, 5). This approach, compared with appendectomy alone, leads to a significant improvement in the survival rate and a lower recurrence in patients with cystadenocarcinoma (7). For this reason, the present patient underwent a right hemicolectomy. There is an association between appendiceal mucoceles and other tumors, particularly carcinoma of the colon (11% to 20%) and tumors of the ovary (2, 5, 10). In a study, a synchronous tumor for which surgery was considered was found in 29% of patients (10). Patients with a benign mucoceles have a better prognosis, with 5-year survival rates of 91% 100%, even in cases with extension of mucus into the extra-appendiceal spaces (2, 7). In malignant mucoceles, however, the 5-year survival rate markedly diminishes to 25%, due to complications of pseudomyxoma peritonei (2, 7). However, no recurrence has been reported of MCA in the literature.

As a result, it can be said that although MCA of the appendix is a rarely seen pathology in children it should be considered in differential diagnosis of intussusception, especially in cases with abdominal mass. This uncommon and potentially malign entity is usually surgically curable, if diagnosed before the rupture.

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