Self-assessment corner

A rare abdominal cystic mass with unusual presentation

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A 10-year-old girl was admitted with the chief complaints of intermittent abdominal pain for 10 days and an episode of bloody stool passage two days prior to the admission. Physical examination showed tenderness over the right abdomen. Laboratory data on blood and urine were unremarkable. Radiography of the abdomen showed ileus. Ultrasonography of the right abdomen (figure 1), computed tomography (CT) (figure 2), and barium enema (figure 3) were subsequently performed.

Figure 1 Sonogram of the right abdomen

Figure 2 Contrast enhanced CT of the abdomen

Figure 3 Barium study of the colon

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Questions

- 1 Describe the imaging findings.
- 2 What is the most probable diagnosis?
- 3 What is the differential diagnosis of a cystic mass in the right abdomen?
- 4 How do the features of the condition in adults or older children differ from those in infants or younger children?

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Answers

QUESTION 1

Sonograms (figure 1) revealed a cystic mass with multiple septa (black arrows) as well as a target-like solid mass with concentric layering (white arrows). CT scans (figure 2) revealed a cystic mass (white arrows) in the right upper abdomen and a target-like mass (black arrows) with fatty component in the right lower abdomen. Radiographs (figure 3) of the barium enema showed a mass in the proximal ascending colon (arrows) which migrated to the distal ascending colon (arrowheads) on post-evacuated film.

QUESTION 2

The most probable diagnosis is cystic lymphangioma or enteric duplication cyst of the ascending colon with intussusception.

QUESTION 3

The differential diagnosis of cystic mass in right abdomen should include choledochal cyst, pancreatic and nonpancreatic pseudocyst, dermoid cyst, ovarian cyst, enteric duplication, mucocele of appendix, mesothelial cyst and cystic lymphangioma.¹⁻³

QUESTION 4

In cases of intussusception, underlying pathology is present in 5% of children under two years of age, in 25% of older children, and in 92% of adults.^{4,5}

The classic presentations of intussusception including sudden, episodic abdominal pain, vomiting, currant-jelly stool and sausage-like palpable mass, are found more frequently in infants and young children with idiopathic intussusception. Intussusception in older children or adults, as shown in this case, has a less consistent presentation, offer a greater diagnostic challenge, and is more likely to have an underlying cause. Bloody stool and palpable mass are the most characteristic presentations to suggest a diagnosis of intussusception.⁶

The common lead or predisposing factors in children include Meckel's diverticulum, gastroenteritis, lymphoma, Henoch-Schönlein purpura, postoperative causes, polyps, duplication, idiopathic thrombocytopenic purpura, cystic fibrosis, and mesenteric adenitis.⁴ In adults, the predisposing factors are predominantly tumours (63%) and postoperative causes, and half of the tumours are malignant.⁵ Adult intussusception has a demonstrable cause in 92% of cases. Therefore, laparotomy is strongly indicated when intussusception occurs in

adults or when predisposing factors are delineated by imaging studies in older children.

Discussion

Approximately 90–95% of all intussusceptions occur in children.⁵ Lymphangiomas are believed to be congenital and more than 50% of cases are apparent at birth; 90% appear by the age of two years.¹ They may grow slowly to manifest compression symptoms. There are three major types of lymphangioma. The lesion in this report was apparently a cystic hygroma. The other two types are simple lymphangioma and cavernous lymphangioma.

About 95% of lymphangiomas occur in the head, neck and axilla; the other 5% are located elsewhere in the abdomen, mediastinum, parotid glands, scrotum, bone and the extremities.⁶ Lymphangiomas in the abdomen occur mostly in the mesentery, omentum or mesocolon, and very rarely in the liver, spleen and kidney.¹ Cystic lymphangiomas of the gastrointestinal tract are extremely rare.

In a review of 79 cases of colonic lymphangioma, the age ranged from one to 78 years, the lesions ranged in size from 0.5 to more than 10 cm in diameter, distributed evenly from cecum to rectum.⁷ Of 79 cases, one (1.3%) was a child, and three (4%) developed intussusception. To our knowledge, only five cases of intestinal lymphangioma which presented as intussusception have been reported.^{2,7,8}

Various imaging techniques, including barium studies, ultrasound and CT can delineate intussusception. The septa in a cystic mass are usually more accurately delineated by ultrasound than by other techniques, as was shown in our patient.¹ As the cystic mass with septa in this report was contiguous to the intussusception, the cyst was presumed to be cystic hygroma.

The sonographic findings of intussusception, including mass with target sign, pseudo-kidney sign, hay-fork sign and multiple concentric layering, were all found in our patient. The CT findings of intussusception were characterised by the presence of thickened bowel wall as well as soft tissue and fat attenuation within the bowel lumen, producing a layering or stratifying effect.

Final diagnosis

Cystic lymphangioma of the ascending colon presenting as intussusception.

Keywords: intussusception; lymphangioma; ultrasound

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