Case Reports

Mesenteric cystic lymphangioma

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ABSTRACT

Abdominal cystic lymphangioma is a very rare congenital tumor of lymphatic origin. It usually appears in the pediatric age and frequently presents with non-specific symptoms and deceptive signs causing, at times, diagnostic dilemmas. Ultrasonography and computer tomography imaging are considered the diagnostic modalities of choice. Two cases of mesenteric cystic lymphangioma, one presenting as perforated appendicitis and the other as recurrent gastritis, are reported. Infection in the first and volvulus in the second case is behind the mode of presentation. The diagnostic approach and treatment are described, with emphasis on the operative tactic applied for upper jejunal resection. A high index of suspicion, accuracy and repeated physical examination and, most important, the liberal use of ultrasonography in all cases of unclear abdominal illness may contribute considerably to a correct diagnosis and decreased morbidity.

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Abdominal cystic lymphangioma (cystic hygroma) is a very rare congenital tumor of the lymphatic system. It is often classified with other cystic lesions under the descriptive topographical term "mesenteric cysts". However, cystic lymphangioma differs in pathological anatomy, histology, incidence and clinical behavior. It often appears in the first decade, usually presents with acute abdominal symptoms, and recurs following incomplete excision with a tendency to invasive growth.[1,2] It is, therefore, eminent to consider cystic lymphangioma a distinct clinical entity. This may lead to obtaining greater insight into the condition and more meaningful treatment. We report 2 cases of mesenteric cystic lymphangioma: One presenting as perforated appendicitis, the other as recurrent gastritis, and describe our diagnostic approach and treatment.

Case Report. ***Patient One.*** A 5-year-old Saudi boy was admitted through the emergency room with generalized progressive abdominal pain and intermittent vomiting of 29 hours duration. The boy was completely well before. There was no history of pain, admission to hospital or any other illness except sore throat 10 days prior to current presentation. On examination, the patient was ill looking and in distress. His body temperature was 38°C; pulse 150 beats/minute and blood pressure 100/70 mm Hg. The abdomen was tender with generalized guarding and point of maximal tenderness in the right iliac fossa. Rebound tenderness was positive. A complete blood count (CBC) revealed leucocytosis of 19900 with 90% neutrophils. Renal function tests and blood sugar was normal. The condition was diagnosed as diffuse peritonitis, most likely resulting from perforated appendicitis. On exploration, mesenteric cysts of different size were found intimately attached to the intestine, nearly 8 cm distal to the duodenojejunal junction. The largest cyst was roughly 7 cm in diameter, hypervascularized, under tension and covered with fibrin, while the remaining neighboring cysts were without inflammatory signs. There was 270° volvulus with subacute obstruction. A 15 cm long jejunal segment including all cysts was excised (**Figure 1**). The resection line was 2 cm below the ligament of Treitz, to ensure good blood supply, dictated by the anatomy of the

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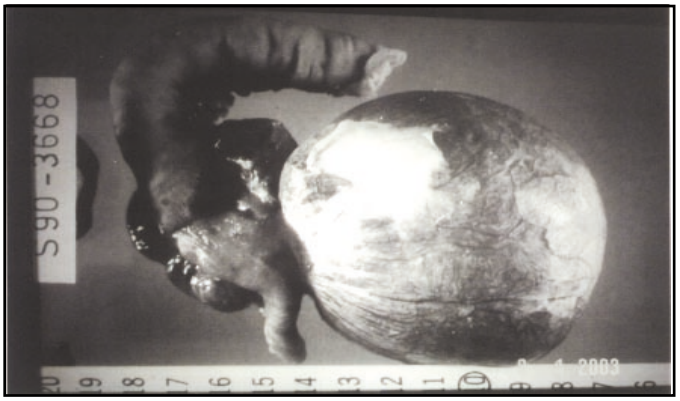

Figure 1 - Infected mesenteric cystic lymphangioma presenting as perforated appendicitis. Note increased vascularity, cyst tension and fibrin exudates.

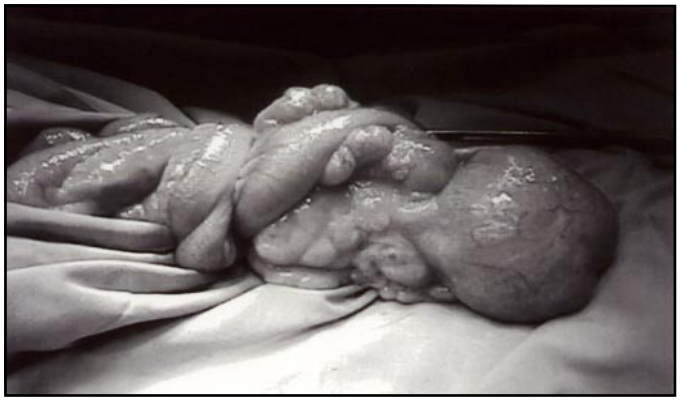

Figure 2 - Mid-ileac mesenteric cystic lymphangioma was found in the pelvis exercising traction on the mesentery, causing a 720° volvulus, and presenting as recurrent gastritis. Note the multitude of twists.

straight jejunal arteries. The continuity of intestine was reestablished by an end-to-end anastomosis using interrupted 3-0 vicryl sutures. Splinting the anastomosis with a nasogastric tube for gravity drainage has minimized the hazard of anastomotic leak within the high-pressure zone of the duodenojejunal flexure. The patient reported immediate relief of symptoms on the first postoperative day, and CBC became normal on the second. The histopathology revealed acutely inflamed cystic lymphangioma. One week later, he was discharged in good condition.

Patient 2. An 11-year-old Saudi girl was admitted to the pediatric ward because of abdominal pain and recurrent vomiting diagnosed as gastritis. The first attack occurred 7 months prior to current presentation, and was followed several times by similar episodes. Each time, the diagnosis of gastritis was made and antacids and antispasmodics were prescribed. At the last visit, the pain was unusually severe, agonizing, localized to the epigastrium, and boring to the back; the antacids and antispasmodics brought little relief. The patient was admitted for investigation. On examination, she was ill looking and in distress. Her body temperature was 37.7°C; pulse 105 beats/minute and blood pressure 125/65 mm Hg. The abdomen was tender in the epigastrium, but not distended or rigid. Complete blood count, renal and liver function tests, amylase, lipase and urine analysis were normal. Chest and plain abdominal radiographs were unremarkable. Ultrasonography of the upper abdomen revealed signs of chronic cholecystitis with query sludge in the gallbladder. The barium meal raised suspicion of tuberculosis or Crohn's disease in the distal jejunal and proximal ileac loops. Endoscopic retrograde cholangiopancreatography was normal. On repeated rectal examination, a cystic lesion was suspected outside the rectum. Ultrasonography (U/S) of the lower abdomen confirmed a cyst posterior to the urinary bladder, measuring 6.5 x 3.5 cm. Abdominal computed tomography (CT) corroborated the pelvic cyst, but the organ of origin remained indefinite. On exploration, multiple mesenteric cysts, the largest of approximately 8 cm diameter were found nearly 100 cm proximal to the ileocecal valve. The mesentery was extremely stretched and twisted, causing a 720° volvulus (**Figure 2**). The gallbladder was dusky with thickened wall and increased vascularity. The appendix and terminal ileum were normal. The volvulus was twisted back and all the cysts, including the closely involved intestine of 20 cm length, were resected. The continuity of the intestine was reestablished by an end-to-end anastomosis using 3-0 vicryl-interrupted sutures. Cholecystectomy and appendectomy were completed. The histopathology revealed multiple multilocular cystic lymphangioma, chronic cholecystitis and normal appendix. The patient reported improvement on the first postoperative day. The course of recovery was uneventful, and she was discharged on the 7th postoperative day in good health. She has been followed up for 3 years without evidence of recurrence.

Discussion. Abdominal cysts have recently been classified into cysts of lymphatic, mesothelial, enteric, or urogenital origin, beside cystic teratoma and pseudocysts.[3,4] Cystic lymphangioma (cystic hygroma) of the abdomen is a very rare lesion of lymphatic origin with an incidence of 2 per 28,000 pediatric admissions to our hospital. This is, inexplicably, nearly twice as high as the incidence of 4 per 100,000 pediatric admission reported.[5] The etiology remains unknown, although many theories have been proposed to explain the pathogenesis.[6]

Cystic lymphangioma is commonly described in the head and neck regions, followed by the trunk and extremities. All other sites including the abdomen comprise less than 5% of cases.[7] In contrast, to "mesenteric cyst", cystic lymphangioma is usually symptomatic (88-100%), reflecting its more aggressive nature.[1,3] The most common clinical presentations are

abdominal pain, increased abdominal girth, and palpable abdominal mass, tenderness on palpation, vomiting and nausea. Also, vague symptoms such as constipation, anorexia, fatigue, diarrhea and gastrointestinal bleeding have been reported. The severity of symptoms is dependent on size, location and possible complications of the lesion such as rupture, torsion, infection or hemorrhage.[5,8] This is reflected well in both our cases (**Figures 1 & 2**). This lesion, as we have seen, often gives rise to non-specific symptoms and deceptive signs, which frequently cause diagnostic dilemmas, and exposes the patient to unnecessary investigations. The agonizing pain with radiation to the back, in the second case, is most likely due to traction in the mesentery and torsion of the small intestine.

The accurate diagnosis has rarely been made ahead of the operation in the era before U/S.[8] Plain radiographs, barium studies and intravenous pyelography may demonstrate displacement of the alimentary, urinary tract, or both and other non-specific radiological signs, but the so yielded information is of little value for making a precise diagnosis. Ultrasonography and CT are deemed the diagnostic tools of choice.[5] Both give information on the cystic nature of the lesion, its location, size, thickness of the wall, and possible site of origin, although U/S demonstrates the internal nature of the cyst more precisely than CT.[3] In selected cases, magnetic resonance imaging may provide more specific information on location and affected organ.[3] However, substantial overlap in the imaging findings of the different abdominal cystic lesions may result in confusing cystic lymphangioma with ovarian cysts, intestinal duplication, and necrotic tumors of smooth muscle such as leiomyoma and leiomyosarcoma, cystic mesothelioma and cystic teratoma, lymphoma and retroperitoneal cysts.[3,5,8,9] In our first case the mode of presentation left no doubt about the diagnosis of perforated appendicitis and none of the said diagnostic tools has been considered. Acute appendicitis, intussusception and ovarian cysts were the most frequent erroneous diagnosis reported.[5,8]

At laparotomy, cystic lymphangioma can be suspected by its gross appearance. It is often intimately attached to the intestinal wall, multiple and multilocular.[1,3] The treatment modality of choice is complete excision. This is rarely achieved by enucleation. Complete excision commonly requires the resection of the closely attached intestine, as observed in both our cases. Incomplete excision bears the hazard of recurrence with a tendency for invasive growth.[1,2] Therefore, resection of rather dispensable organs such as spleen and tail of pancreas has been advocated to completely remove a lymphangioma.[10] However, the excision is at times by necessity incomplete, because the tumor is insinuated between and around vital structures. In these cases, external marsupialization is suggested. The lining endothelium is then destroyed by sclerosants such as tincture of iodine, or 10% glucose solution to minimize recurrence.[11] Simple drainage procedures are regarded as historical.[5] They are frequently complicated by infection and recurrence.

In conclusion, cystic lymphangioma is a very rare cystic tumor of lymphatic origin. Lack of knowledge or unawareness of the condition may result in unnecessary, partially invasive investigations and inappropriate treatment with increased morbidity. A high index of suspicion, accurate physical examination and the liberal use of U/S in all cases of unclear abdominal illness may contribute considerably to a correct diagnosis and decreased morbidity. Long term follow up, and early detection of recurrence would, furthermore improve the ultimate outcome.

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