

Mesenteric Cystic Lymphangioma of Mesocolic Origin in a Three-Month-Old Infant: A Rare Differential Diagnosis of Abdominal Distension

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ABSTRACT

Cystic lymphangioma is a rare benign tumour occurring during childhood. This tumour, caused by lymphatic system malformations, commonly occurs in head and neck regions. Herein, we report the case of a three-month-old male infant, diagnosed with a large cystic mass in the abdominal cavity and groin. The infant presented with low-grade fever and significant abdominal distension. Abdominal CT scan (with a contrast agent) revealed a large cystic mass in the abdominal cavity. During surgery, the mass was found to originate from the mesentery and was inferiorly connected to the sigmoid colon. Also, a small cystic mass was detected in the left scrotal region. The mass was removed along with a portion of the sigmoid colon and the cyst in the groin. Later on, histopathological examination of the mass confirmed the diagnosis of mesenteric cystic lymphangioma.

Keywords: Mesenteric cystic lymphangioma, Infant, Pakistan

Introduction

Mesenteric cystic lymphangiomas are rare benign tumours in children. Although the etiology of these tumours is still undetermined, abnormal development of lymphatic system is assumed to be the main origin. Clinically, mesenteric cystic lymphangiomas may be confused with mesenteric cysts.

Mesenteric cystic lymphangiomas may remain asymptomatic, although they may result in various symptoms such as abdominal distention, abdominal pain, intestinal obstruction, and even rupture in case of enlargement. Herein, we present a case of cystic lymphangioma of the mesentery and groin in a three-month-old, male infant.

Case report

The present case was a three-month-old infant, suffering from abdominal distension and swelling of the groin for two weeks. Moreover, he presented with low-grade fever for five days. Medical and surgical history of the patient was unremarkable. He was born full term via lower-

segment caesarean section and antenatal records were insignificant. He was breastfed and vaccinated and reached the normal, age-appropriate developmental milestones.

On examination, the infant was febrile and dehydrated, showing a normal anthropometric status. Abdominal examination revealed a grossly distended abdomen, diffuse tenderness, dullness to percussion and normal bowel sounds. The patient also presented with left-sided scrotal swelling. Other systemic examinations were unremarkable.

Based on the assessments, complete blood count showed increased total leukocyte count (TLC) (TLC= $20.9 \times 10^9/l$) with neutrophilia and C-reactive protein (CRP) level higher than 12 mg/dL. Abdominal X-ray results were unremarkable, with the exception of gas-filled, distended and displaced gut loops. Ultrasonography of the abdomen/pelvis showed diffuse ascites and a cystic mass on the left side of the abdomen, extending to the scrotal region. Plain CT scan showed massive ascites, as well.

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Abdominal tap was applied to analyse the ascitic fluid. Analysis of ascitic fluid revealed increased cell count (6000/cmm), with 98% neutrophil predominance, high albumin level and markedly increased lactate dehydrogenase, whereas Gram staining and Ziehl-Neelsen stain were negative. The thyroid profile was normal and carcinoembryonic antigen was negative. Abdominal tuberculosis was also ruled out by evaluations.

The patient was managed with broad-spectrum antibiotics and supportive paraphernalia. CT scan with a contrast agent showed a large multi-septated cystic mass, extending from the ileum into the left iliac fossa and the scrotal region. Surgical intervention was planned and during laparotomy, examination of the mesentery revealed a giant multi-loculated cystic mass (17×16×14 cm³), attached inferiorly to the sigmoid colon.

The cysts were filled with chylous fluid. For treatment, the cysts, the mesentery containing the cysts and a small portion of the sigmoid colon were excised. The cysts in the left scrotal region were removed, as well. A specimen was sent for histopathology examination, confirming the diagnosis of mesenteric cystic lymphangioma.

Postoperative course was uneventful and the patient was followed-up regularly. Based on the one-year follow-up, the infant was asymptomatic and showed good progress. No radiographic evidence of tumour recurrence has been reported, so far.

Discussion

Intra-abdominal cystic lymphangiomas are rare congenital benign tumours. These tumours account for approximately 5% of benign tumours, found in infants and children. They may arise from any abdominal lymph structure and can be detected in the omentum, the mesentery, the mesocolon or retroperitoneum.

The exact underlying cause of these tumours is yet unknown. However, the most widely accepted hypothesis is the inability of lymphatic channels in the mesentery or retroperitoneum to communicate with the main vessels, resulting in the sequestration and formation of cysts (1, 2). However, this hypothesis fails to explain the invasive nature of many cystic lymphangiomas (2).

Overall, 50% of cases involve the head and neck regions and only 10% of these tumours occur in the internal organs (3). Within the gastrointestinal tract, small bowel mesentery is the most commonly involved site, followed by

retroperitoneal regions (2). The mean age of almost 90% of detected cases is two years (4). Also, a male predominance has been observed, with a male-to-female ratio of 5:2; the mean age of presentation is 2.2 years (3).

In terms of gross appearance, lymphangiomas can be divided into three major types including capillary, cavernous (mainly cutaneous) and cystic (mainly internal) lymphangiomas. Clinical presentation varies depending on the size and location of the tumour. In a previous study on a series of 22 patients, one-half of the cases had an asymptomatic abdominal mass, while the remaining subjects typically presented with chronic abdominal pain, with or without distension (5).

Lymphangiomas less commonly present as an "acute abdomen", characterized by acute abdominal pain, distension, vomiting, fever and peritonitis (3, 6). Ascites may be misdiagnosed when the cysts are large (5).

Radiological investigations are normally used to diagnose these tumours. Although plain abdominal radiographs typically fail to show these lesions, they may identify bowel displacement or obstruction⁶. In the present case, abdominal X-ray results were unremarkable, and the patient showed non-specific signs. In such cases, ultrasonography is the diagnostic method of choice. However, one limitation of ultrasonography is that the relatively large size of lesions may interfere with the evaluation of borders and sites of origin (2). To further evaluate these tumours, CT scan with or without a contrast agent can be used, as well.

Although abdominal lymphangioma is considered a benign tumour, it may become locally invasive. Complete surgical resection is the treatment of choice, especially when the mass is enlarged in size or leads to acute symptoms (3). On the other hand, simple aspiration is associated with a high recurrence rate (5). Complete en bloc resections usually result in excellent outcomes and prognoses in patients (7, 8). Among patients with incomplete resection, the recurrence rate has been estimated at approximately 10% (8). In our case, complete resection was carried out.

Weeda et al. reported two cases of mesenteric cystic lymphangioma, located near the duodenojejunal junction (9). Similarly, Minakshi et al. reported a case of mesenteric cystic lymphangioma in a two-month-old infant, involving the ileocolic junction (10). In children, ultrasonography is the preferred imaging modality for postoperative surveillance. However, no published guidelines are available to determine the

frequency of ultrasonography at which surveillance ultrasounds should be performed.

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