CYSTIC LYMPHANGIOMA OF JEJUNAL MESENTERY MIMICKING ACUTE APPENDICITIS: CASE REPORT

Marko Mesić[1], Irenej Cigit[1], Mirko Žganjer[1], Ante Čizmić[1], Božidar Župančić[1] and Zrinko Madžar[2]

[1]Department of Pediatric Surgery, Zagreb University Children's Hospital; [2]Clinical Department of Surgery, Sestre milosrdnice University Hospital Center, Zagreb, Croatia

SUMMARY – Cystic lymphangiomas of the small bowel mesentery are rare manifestations of intra-abdominal tumors. Usually, they are discovered incidentally during examination for an unrelated abdominal illness. We present a case of a 4-year-old boy who was admitted to our hospital because of the right lower quadrant acute abdominal pain suspect of acute appendicitis. At laparotomy, a giant, cystic, encapsulated and lipomatous mesenterial mass was found, 15x15x10 cm in size, infiltrating the jejunum. The tumor was located 70 cm from Treitz's ligament. Extirpation of tumor mass with intestinal resection of the involved loops was necessary. Pathologic examination confirmed the diagnosis of mesenteric cystic lymphangioma. Although they are rare, cystic mesenteric lymphangiomas should be considered as a possible cause of acute abdomen and treated with surgical resection. Prognosis after surgical removal is excellent.

Key words: *Cystic lymphangioma; Mesenteric lymphangioma; Acute appendicitis; Intra-abdominal tumor*

Introduction

Abdominal cystic lymphangiomas are rare congenital benign malformations of the lymphatic system with uncertain etiology, predominantly occurring in children. They are more frequent in boys (M/F ratio, 5:2) with a mean age at presentation of 2 years[1,2]. Usual locations are small-bowel mesentery, followed by the omentum, mesocolon and retroperitoneum[3]. There is a variety of clinical manifestations, ranging from asymptomatic abdominal masses to symptoms of an acute abdomen. Therefore, they may be discovered incidentally during examination for an unrelated illness, as in our case of a 4-year-old boy who was admitted because of acute abdominal pain.

Correspondence to: *Marko Mesić, MD*, Department of Pediatric Surgery, Zagreb University Children's Hospital, Klaićeva 16, HR-10000 Zagreb, Croatia
E-mail: mrkmesic@yahoo.com

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Case Report

A 4-year-old boy was admitted to our hospital because of the right lower quadrant acute abdominal pain suspect of acute appendicitis. According to the parents' report, he was suffering from abdominal pain for the last 24 hours. Physical examination revealed slightly distended abdomen with palpatory dominant pain in the right lower abdominal quadrant. Laboratory findings showed mild leukocytosis (12.23x10^9/L). There was no vomiting and the patient was subfebrile (37.5 °C).

Eventually, surgery was indicated because of worsening of abdominal pain. During preoperative preparation, an unusual palpable mass was noticed in the umbilical region, so medial laparotomy was performed instead of usual Sprengel's incision.

On laparotomy, a giant, cystic, encapsulated and lipomatous mesenterial mass was found, 15x15x10 cm in size, infiltrating the jejunum (Fig. 1). The tumor was located 70 cm from Treitz's ligament. Tumor mass

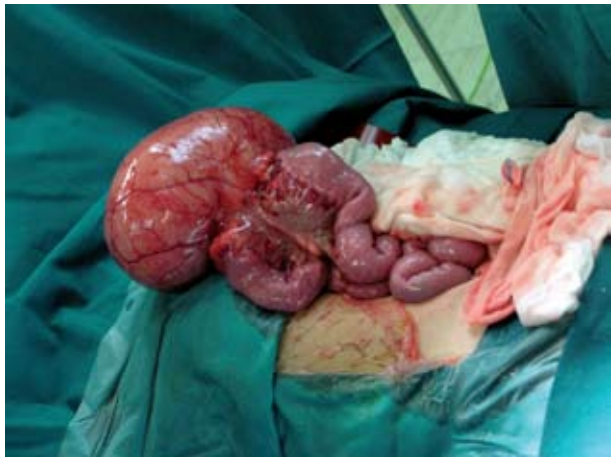

Fig.1. Giant mesenteric cystic lymphangioma, 15x15x10 cm.

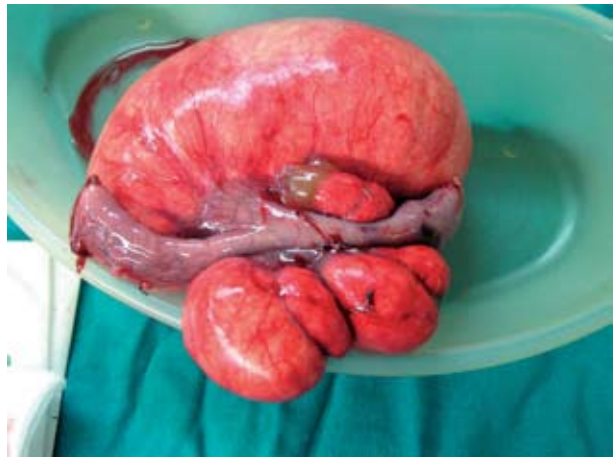

Fig. 2. Excised tumor mass along with 15 cm of the jejunum.

was excised along with 15 cm of jejunum (Fig. 2). The bowel continuity was restored with an end-to-end anastomosis. Also, appendectomy was performed.

Histopathologic examination described a baggy looking formation of 15 cm in diameter, developing from bowel wall, with smooth lumen and filled with chylous-milky looking fluid. Tumor wall was impregnated with dilated lymph vessels. Lymphatic endothelium was positive for D2-40 expression, suggesting the diagnosis of mesenteric cystic lymphangioma. The early postoperative course was uneventful.

At 6-month follow-up, the patient remained asymptomatic without recurrences.

Discussion

Cystic lymphangiomas are rare congenital benign malformations of the lymphatic system, accounting for 5%-6% of all benign tumors in infants and children[3,4]. Generally, they are common in children, 40% usually present by the age of 1 year and 80% by the age of 5 years. Head and neck are involved in 50%, and internal organs in only 10% of cases[5]. Sixty percent of these masses are present at birth. Abdominal cystic lymphangiomas are very uncommon. Diagnosis of mesenteric lymphangioma is eventually made histopathologically following surgical resection. Characteristic finding is endothelial lining with connective tissue, smooth muscle fibers, and dilated lymph vessels[5]. Intraluminal content can be serous, serosanguinous, or chylous[4,5]. Lymphatic endothelium is positive for D2-40 expression.

There are several theories about their development. Proliferation and dilatation of blind-ended lymphatic sacs during fetal development leads to the formation of blind cystic lymphatic spaces lined by endothelial layers and lack of communication between small bowel lymphatic tissue and the main lymphatic vessels[6]. The theory of a congenital etiology may be supported by the fact that most cases are presented in childhood. Other potential causes are thought to include abdominal trauma, localized lymphatic degeneration, and lymphatic obstruction[4,6].

Intra-abdominal lymphangiomas may be asymptomatic or lead to acute abdominal pain, which depends on mass size and location.

The possible complications are hemorrhage, torsion, or rupture of the lymphangioma. Pain can also be caused by compression of the surrounding structures or intestinal obstruction and volvulus.

Differential diagnosis includes other fluid-filled lesions such as pseudocysts, dermoid cysts, enteric duplications, lymphoceles, or neoplasms like mesotheliomas, pancreatic tumors, lipomas, teratomas, leiomyosarcomas, neurofibromas or liposarcomas[7,8].

Diagnostic imaging of the mesenteric cystic lymphangioma is based on abdominal ultrasound examination, computerized tomography (CT) scan, or magnetic resonance imaging (MRI)[9,10]. In our case, we did not perform ultrasound examination because we had

certain clinical and laboratory findings indicating that we were dealing with acute appendicitis.

Diagnostic imaging, ultrasound, CT and MRI confirm the presence of the mass, but it is not sufficient to make definitive diagnosis. However, Jain *et al*. report on using multislice spiral CT to confidently establish the diagnosis prior to surgery[11].

Treatment of mesenteric cystic lymphangioma is primarily by surgical resection using open surgery. However, laparoscopic resection has been reported[12]. Regarding the intimacy of lymphangioma and intestinal wall, it is usually necessary to perform bowel resection, as it was in our case.

In some patients, spontaneous regression has been reported encouraging a conservative approach[13]. Conservative treatment includes aspiration and injection of a sclerosing agent. This can be recommended for emergency decompression, but as definitive therapy they have a high recurrence rate. Alternative injection of alcohol[14], bleomycin[15], or OK432[16,17] into the tumor has also been reported in cases where radical surgery was not technically possible.

Although mesenteric cystic lymphangiomas are benign lesions, and in the majority of cases asymptomatic, they can lead to severe abdominal complications due to aggressive and invasive growth. Therefore, they should be considered on differential diagnosis of acute abdomen in childhood. Only radical excision can be optimal treatment and prevent recurrence.

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Sažetak

CISTIČNI LIMFANGIOM MEZENTERIJA JEJUNUMA KOJI OPONAŠA AKUTNI APENDICITIS: PRIKAZ SLUČAJA

M. Mesić, I. Cigit, M. Žganjer, A. Čizmić, B. Župančić i *Z. Madžar*

Cistični limfangiomi mezenterija tankog crijeva pripadaju rijetkim intraabdominalnim tumorima. Obično se nađu slučajno tijekom obrade zbog neodređene trbušne boli. Prikazuje se slučaj četverogodišnjeg dječaka hospitaliziranog zbog bolova u donjem desnom abdominalnom kvadrantu sumnjivih na akutnu upalu crvuljka. Nakon učinjene laparotomije našla se velika, cistična, inkapsulirana, lipomatozna mezenterijska masa veličine 15x15x10 cm koja je infiltrirala jejunum. Tumor je bio smješten oko 70 cm aboralno od Treitzova ligamenta. Učinjena je ekstirpacija tumora s infiltriranom jejunalnom vijugom. Patohistološki nalaz je potvrdio dijagnozu mezenterijskog cističnog limfangioma. Iako rijetki, cistični mezenterijski limfangiomi se mogu smatrati mogućim uzrokom akutne abdominalne boli i liječiti kirurškom resekcijom. Prognoza je nakon kirurškog odstranjenja odlična.

Ključne riječi: *Cistični limfangiom; Mezenterijski limfangiom; Akutni apendicitis; Intraabdominalni tumor*