Case Report

Two unusual sites of cystic lymphangioma in a child: A report of imaging profile with surgical and histopathologic findings

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Abstract Cystic lymphangioma (CL) is a benign lymphatic malformation mostly seen in the head and neck of neonates and infants. Abdominal CL is an unusual entity which may present in omentum, mesentery, abdominal wall, or solid organs. The authors present an unusual case with two separate abdominal cystic lymphangiomas.

Key Words: Abdominal pain, cystic lymphangioma, pediatric surgery

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INTRODUCTION

Cystic lymphangiomas (CLs) are benign congenital malformations of the lymphatic system, mostly present at birth or during infancy. Their usual sites are head and neck, axillary regions, and extremities. They remain harmless until they become complicated by hemorrhage, rupture, or infection, or impose pressure effect on vital structures.[1] The authors present an unusual case with two separate abdominal CLs.

CASE REPORT

A 9-year-old male was admitted to our emergency department (Emam Hossein Pediatric Hospital, Isfahan) with fever and abdominal pain since a day before admission. The pain was described as a continuous, non-referral pain which was localized in the right upper quadrant (RUQ) and associated with bulging at the site of pain. No other complaint or related history was noted. Physical examination revealed low-grade fever with a tender fluctuated mass (approximately 80 × 40 mm in size) in the right subcostal area with no skin manifestation.

Laboratory tests revealed leukocytosis 17,300/μl (90% neutrophils) and normal liver tests. Abdominal ultrasonography showed a multicystic lesion with several thick septa, occupying the right subphrenic area. It also revealed scattered hyperechoic lesions in the peripheral zones of spleen which had small cystic areas in their matrix. Further evaluation with double-contrast CT scan revealed a multicystic mass that was 103 × 42 mm in size, with thick and enhanceable septa in the right subphrenic region, anterior to the left and middle segments of the liver with compressive effect on the hepatic parenchyma, and also scattered hypodense lesions in the spleen [Figure 1]. The main differential diagnoses were multiloculated abscess, complicated CL, and exophytic hydatid cyst.

The patient was administered conservative treatment including analgesic and proper empirical antibiotics. After a week of hospitalization, No significant improvement was observed.

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He underwent an elective laparotomy. Surgical findings were a cystic mass which contained multiple hemorrhagic cysts adjacent to the posterior fascia of the anterior abdominal wall in RUQ, in addition to some small cysts in the spleen [Figures 2 and 3]. The cystic mass of abdominal wall was resected as far as possible and splenectomy was performed. Histopathologic study of the specimen showed large, thin-walled lymphatic channels in loose connective tissue [Figure 4]. All findings were compatible with a diagnosis of CL. The patient has been followed up for 2 months post-surgery, with no complication or recurrence.

DISCUSSION

CL, which has been also known as *hygroma*, probably arises from developmental failure of the lymphatic system to communicate with the venous system.[2] Classic manifestation of CL is a soft tissue mass in the neck or axillary regions of neonates or infants. Infrequently, it may be seen in thoracic, abdominal, or pelvic cavities, with various clinical and imaging features which cause CL to become a diagnostic dilemma.[1]

Abdominal CL is an unusual entity which may present in omentum, mesentery, retroperitoneum, abdominal wall, or solid organs. It remains asymptomatic until it presents with abdominal pain, a palpable mass, or gastrointestinal symptoms. Complications such as infection, hemorrhage, or rupture confuse its clinical and imaging features and cause to expand the list of differential diagnoses.[3]

In this paper, a child was reported with sudden onset of localized abdominal pain and bulging caused by rapid growth of a hemorrhagic CL of the abdominal wall.

Abdominal imaging studies discovered another lesion in the spleen. Imaging features of the splenic lesion were a helpful clue for consideration of CL. Splenic CL is also uncommon and tends to be located in the periphery, especially in the subcapsular region.

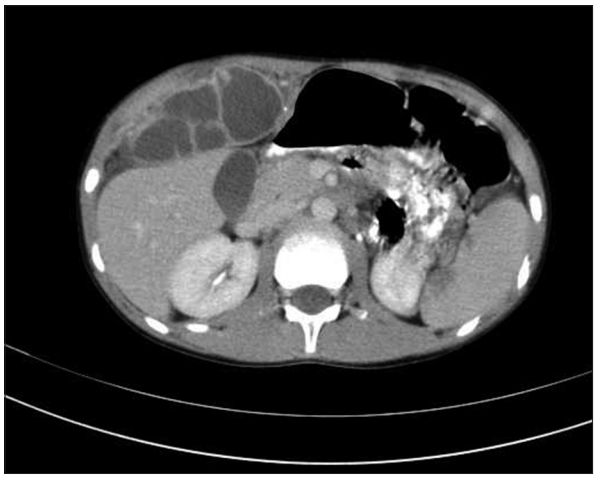

Figure 1: Double contrast axial CT scan showing a multicystic mass with thick and enhanceable septa in the right subphrenic region and hypodense lesions in the spleen

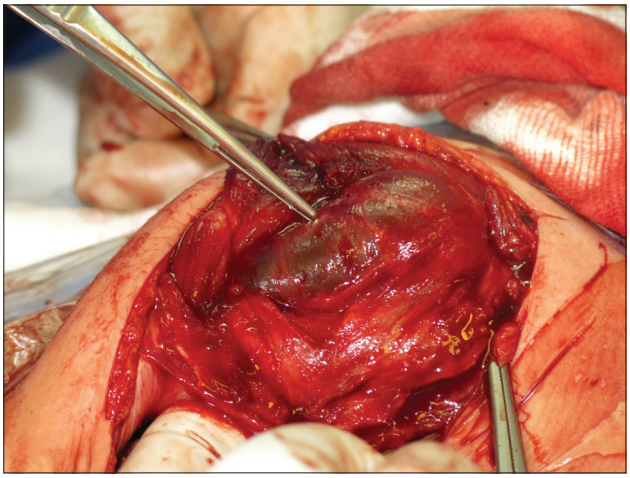

Figure 3: Intraoperative image of CL of the abdominal wall

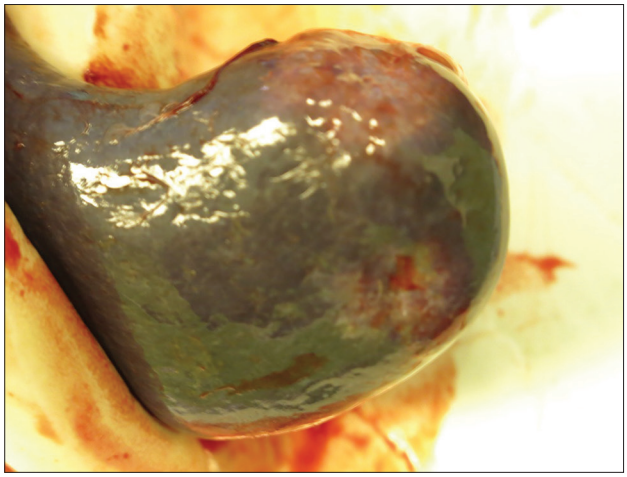

Figure 2: Intraoperative image of spleen with peripheral heterogenous echo texture

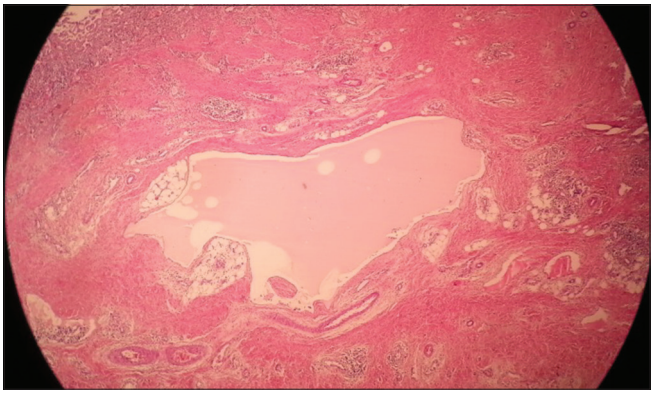

Figure 4: Histopathologic study of cystic mass of the abdominal wall showing large, thin-walled lymphatic channels in loose connective tissue

Splenic CL may sometimes be accompanied by CL in other organs.[4]

Before surgical intervention, multiloculated abscess was one of the primary diagnoses because of epidemiologic considerations and presence of fever. However, lack of response to widespread antibiotics weakened this diagnosis.

Surgical resection is inevitable in the cases of complicated abdominal CL. However, in asymptomatic patients with innocent CL, medical treatment using sclerosing agents may be considered.

In this patient, splenectomy was performed as the splenic CL was scattered and was in the risk of recurrence.

In conclusion, complicated abdominal CL should be put in the differential diagnoses of acute abdominal pain in children. Imaging modalities may be helpful for timely diagnosis and proper surgical decision.

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