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Case Report

Multicystic Mesenteric Lymphangioma – A Case Report

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Abstract

Mesenteric cystic lymphangiomas are rare benign lesion of lymphatic system occurring in children. They are common in males with a median age of presentation of 2 years. It can either be asymptomatic or present as acute abdomen when the mass produces intestinal volvulus. It is often confused with mesenteric cyst both clinically and radiologically. The true nature of lesion is revealed on histopathological examination. We report such a case in a 3 year old boy who presented with complaints of abdominal pain and distension, which on evaluation thought to be mesenteric cyst which was excised and turned out to be a cystic lymphangioma in histopathological examination. Establishing the lymphatic nature is also important owing to the development of targeted therapy for aggressive and recurrent lesions. Immunohistochemistry, hence, hold an important role in this characterization and is expected to rise in significance in the future.

Keywords: Lymphangioma, Mesentry.

INTRODUCTION

Lymphangioma in children are extremely rare pathologic entities characterized by unusual location. They are more frequent in boys with a median age of presentation of 2 years. They often represent a challenge for pediatrician and pathologists from being asymptomatic to with an acute abdomen presentation. We hereby describe case of a 3 year old boy who presented with complaints of abdominal pain and distension, associated with vomiting for 2 weeks and was diagnosed as mesenteric lymphangioma post excision.

CASE REPORT

A 3 year old male child presented with complaints of recurrent abdominal pain and abdominal distension associated with loose stools and fever of 2 weeks duration. There was no relevant past history. Developmental milestones were attained normally. There was no history of recent travel. He was treated at the local health care facility suspecting an infectious etiology, but didn't improve.

On examination, he was febrile with mild tachycardia. Other vitals were stable. Abdomen was distended, and a soft non tender mobile 10x10 cm mass was palpated in the right upper quadrant of abdomen.

Ultrasound showed a large well defined multiloculated heterogenously hyperechoic lesion extending from epigastrium to both iliac fossa with multiple areas of cystic change and solid component, possibly a lymphoma (Fig 2). CECT abdomen revealed the mass to be a large multiloculated thick walled and septated lesion measuring 10x9x7cm in right upper quadrant with minimal ascites, possibly infected mesenteric cyst (Fig 1).

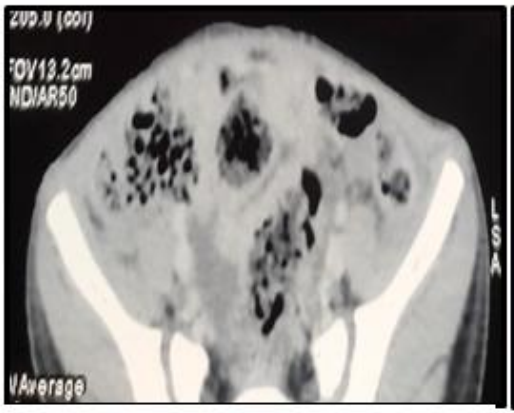

Fig-1: CECT Abdomen

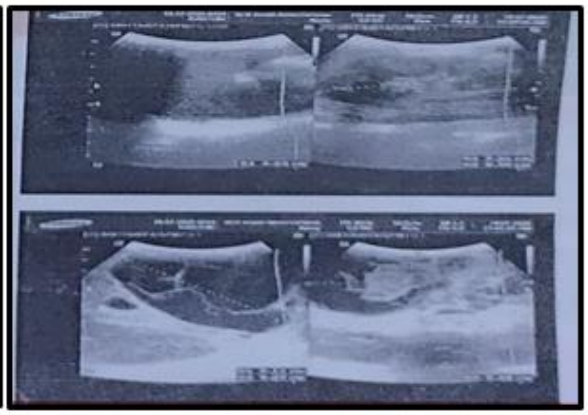

Fig-2: USG abdomen

The Surgeon proceeded with laparotomy and excised the cyst.

GROSS DESCRIPTION

A cystically enlarged mass weighing 200gm measuring 12x8x6 cm on cutting open showed mucoid material. On cut section it was a multiloculated cyst with smooth inner wall and foci showing aggregates of microcysts (Fig 3).

Fig-3: Excised specimen showing multiloculated cyst

MICROSCOPY

Sections from mass showed multiple irregular vascular spaces lined by flattened epithelial cells with a collagenous stroma accompanied by lymphoid tissue.

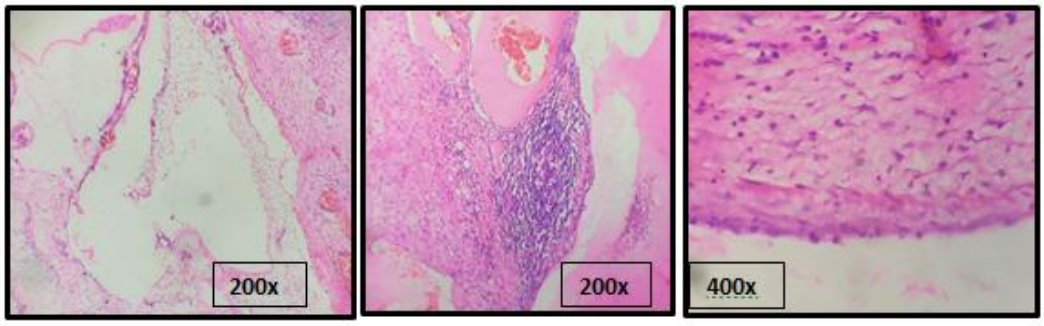

Fig-4: Microscopy of the cyst wall lining

The lining of the cyst wall was positive for the markers CD31 and CD 34 and negative for Calretinin.

DIAGNOSIS

Benign multicystic mesenteric lymphangioma.

DISCUSSION

Lymphangioma is a rare benign intra abdominal anomaly of uncertain etiology, predominantly occurring in children usually in neck, head and axillary regions. Lymphangioma of small

bowel mesentery is described in less than 1% of lymphangiomas [1].

They are thought to develop from an embryological failure of lymphatic system; lack of communication between small bowel lymphatic system and the main lymphatic system during fetal development. This results in the formation of blind cystic lymphatic spaces [2]. The differential diagnoses include multicystic mesothelioma, lymphocele, dermoid cyst, lipoma, teratoma and liposarcomas [3].

The clinical presentation is varied from being asymptomatic to producing abdominal distension, mass and intestinal obstruction. It presents as an acute abdomen when the mass induces rotation of small bowel resulting in volvulus with a consequent closed loop bowel obstruction [4]. The treatment is surgical excision.

A pathological classification was proposed which divided mesenteric cysts into Type 1 (pedicled) and Type 2 (sessile), which are limited to mesentry and can be excised completely with or without resection of the involved gut. Type 3 and 4 which are multicentric, extend to retroperitoneum and require complex operations [5].

The cysts are traditionally divided into 3 histologic types – capillary, cavernous and cystic [6]. Capillary type usually originates in skin and consist of uniform small thin walled lymphatic spaces. Cavernous type is composed of dilated lymphatic spaces of varying size and a lymphoid stroma. Cystic type shows dilated lymphatic spaces associated with collagen and smooth muscle in stroma and they lack connection to adjacent normal lymphatic spaces.

Immunohistochemical evaluation is a useful tool to identify lymphatic origin of the cyst and to differentiate it from mesothelioma which is a close differential diagnosis [7]. The D2-40 stain certifies the lymphatic nature of lining cell while they are positive for Calretinin in mesothelioma. It is differentiated from lymphangiomyoma where HMB 45 show positivity in smooth muscles around lymphatic vessels in the latter. A new approach of infantile lymphangiomas following surgery is regarding use of specific lymphatic markers Panel including D2-40, Prox 1, VEGFR 3, PDGFR, Ki 67 which may improve the characterization of such lesions regarding their prognosis, recurrence rate and targeted therapy implementation especially for those with aggressive behavior [7].

CONCLUSION

We hereby described a case of multicystic lymphangioma in a 3 year old boy who presented with features of subacute intestinal obstruction. Evaluation revealed an intra abdominal mass which radiologically suggested an infected mesenteric cyst and excision was done. Histopathology and IHC showed it to be a mesenteric lymphangioma. Being a rare entity and often confused clinically and radiologically, the histopathological study is crucial in its diagnosis. And role of immunohistochemistry is also gaining importance with new targeted therapies for recurrent cases.

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