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Cystic lymphangioma of the Mesentery - An Unusual Case

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ABSTRACT *Cystic lymphangiomas are uncommon lymphatic anomalies of congenital origin, which usually are seen in the head and neck. We report a case of mesenteric lymphangioma presenting with abdominal pain and distension. The lesion was surgically excised and evaluated histopathologically revealing a thin-walled cystic structure lined by attenuated endothelial cell lining confirmed by immunohistochemical staining with CD34. Surgical excision was curative with no long term complications. Cystic lymphangiomas usually present as incidental findings but they can also present with abdominal distention, hemorrhage, infection, and rupture. The present case highlights the importance of considering cystic lymphangioma in the differential diagnosis of abdominal mass especially in children.*

Cystic Lymphangioma is an uncommon benign lesion which usually presents before the age of three years and is preferentially located in the head & neck region.[1] It also occasionally present in other organs like liver, spleen and bones and rarely at intraperitoneal and retroperitoneal sites.[1, 2] It is thought to be a lymphatic anomaly of congenital origin.[2] Here we present a case of cystic lymphangioma of the mesentery (CLM) with immunohistochemical study.

A 9-year-old female child presented with vomiting, abdominal pain and distension since 10 days. On examination there was predominantly right sided abdominal distension with tenderness. Computed tomography of the abdomen was done and it revealed dilation of the proximal bowel loops and a large hypodense non-enhancing lesion, extending from below the right hepatic lobe to the pelvis superior to the urinary bladder, with a thin wall and incomplete internal septae [Figure 1]. The possibility of mesenteric cyst with subacute intestinal obstruction was suggested. Exploratory laparotomy was done. Intraoperatively a large cyst attached to the proximal ileum and compressing the adjacent bowel loops was noted. Resection of the cyst along with part of the ileum was performed and tissue was submitted for histopathological examination.

Gross examination revealed a smooth, thin walled, translucent cyst (13 x 10 x 4 cm) attached to mesentery and serosa of the small bowel [Figure 2A]. The cyst showed thin, incomplete internal septae and contained a clear yellow serous fluid. Microscopically, sections from cyst wall revealed smooth muscle bundles overlined by attenuated endothelial cells on the luminal aspect. Immunohistochemical studies showed immunoreactivity of the endothelial lining for CD34 (clone QBEnd/10, Novacastra) while smooth muscle wall was immunoreactive for smooth muscle actin (SMA) (clone 1A4, Dako) [Figure 2B]. Based on these findings, a diagnosis of mesenteric cystic lymphangioma was offered. Post-operative recovery was uneventful and the patient was discharged on 15th postoperative day.

Most cases of lymphangioma present in the cervical or axillary region. Less than 3% cases of lymphangiomas occur in the abdomen which includes sites like omentum, mesocolon or retroperitoneum.[2] Only a few cases are reported in the mesentery.

The pathogenesis of mesenteric lymphangiomas is ill understood, however they are believed to be developmental in origin. Their association with volulus & lymphadenitis with lymphatic obstruction has also been documented.[2]

It must be noted that despite their benign nature CLMs may cause significant morbidity & mortality due to their large size and possibility of infection.[1] The clinical presentation is variable and symptoms depend on the size, location and complications like torsion, hemorrhage, infection and rupture.[3] CLMs may be discovered as incidental findings or may present as chronic progressive abdominal distension and pain. Acute presentation may also be seen in the form of acute abdominal pain, distension, vomiting & fever due to infection.[3] Complete surgical excision is the treatment of choice & the prognosis is very good.[1] Our experience with this case highlights the importance of CLM as a differential diagnosis in a child presenting with cystic lesion in the abdomen.

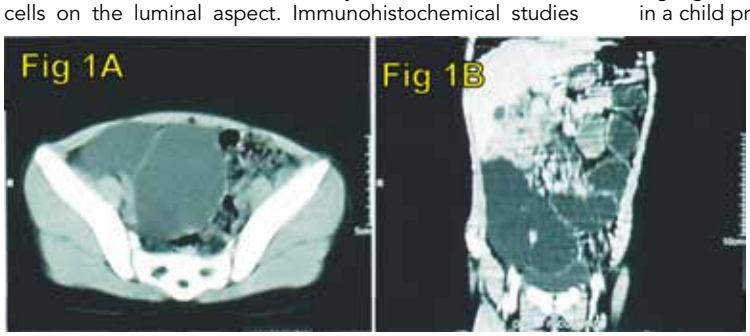

Figure 1: Computed tomography images in (A) Transverse & (B) Saggital plain showing a large hypodense non-enhancing cystic lesion extending from inferior aspect of right hepatic lobe to pelvis. The cyst is thin walled with few incomplete septae.

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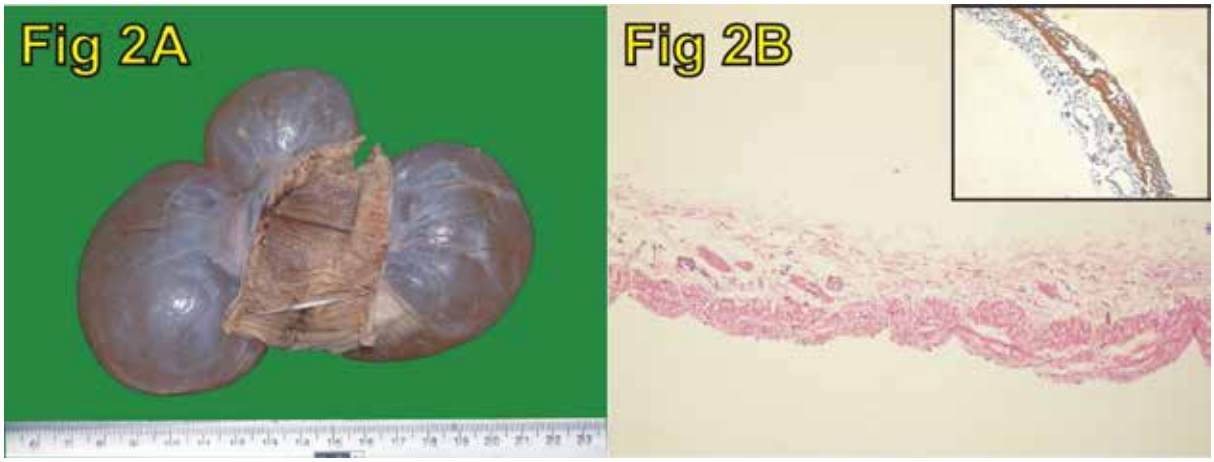

Figure 2: (A) Thin walled mesenteric cyst with attached segment of small intestine. (B) Microscopy – Cyst wall composed of bundles and fascicles of smooth muscle. Endothelial lining is attenuated (Hematoxylin-eosin stain x100). Inset shows smooth muscle fascicles showing immunoreactivity for smooth muscle actin(SMA).

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