**CYSTIC LYMPHANGIOMA OF THE MESENTRY - A RARE ENTITY IN A YOUNG FEMALE**

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**ABSTRACT**
Lymphangioma is a rare benign condition characterized by proliferation of lymphatic spaces. It is usually found in the head and neck of affected children. Lymphangioma of small bowel mesentry is rare. It can cause fatal complications such as volvulus or involvement of mesenteric arteries requiring emergency surgery. We present a case of a 13-year-old female presented with acute abdomen, emergency exploratory laparotomy performed and an attempt was made to derotate the bowel and excision of the cyst along with resection of ileal intestinal segment was sent. Histopathological examination and Immunohistochemistry (IHC) confirmed the diagnosis of a Mesenteric Lymphangioma.

**Key Words:** Mesenteric lymphangioma, Intra Abdominal, Cystic mesothelioma, Lymphatic cyst, IHC

**INTRODUCTION**
Lymphangiomas are commonest tumors of infancy uncommon, congenital malformations of the lymphatic system and 75% occur in cervical region. 90% of all lymphangiomas manifest in first two years of life. However intraabdominal locations of lymphangiomas are very rare, usually slow growing and manifest in early adulthood (Lt Col & Col, 2007). We report a case of mesenteric lymphangioma in 13-year-old girl who presented with acute abdomen.

**CASE REPORT**
A 13 yr old female child presented with acute abdomen with episodes of vomitings and tenderness in right iliac fossa, clinically diagnosed as acute appendicitis. X-Ray abdomen was non contributory. In Ultrasonogram of abdomen right iliac fossa was obscured by bowel gases. Laboratory investigations revealed hematological parameters within normal range. Emergency laparotomy performed and surgical notes revealed - twisted small bowel segment with a cyst measuring 6*5cm in the mesentry of ileum and collapsed jejunum. Resected ileum with cyst was sent for histopathological examination.

**Gross**
We received small bowel segment of 6 cm in length and cut open and collapsed cyst of size 6*6cms. Contents were milky white in colour. Cut section of intestinal segment revealed normal appearing mucosa and attached mesentry with the cyst wall (Fig 1).

![Figure 1: Gross showing small bowel segment with cut opened multiloculated cyst in mesentry](image-url)
Microscopic examination showed ileal segment with mucosa and other layers normal. Serosal aspect showed multiple cystic spaces lined by flattened endothelial lining surrounding stroma showed smooth muscle cells and lymphoid tissue. A differential diagnosis of lymphatic cyst, cystic mesothelioma and lymphangiomyoma were considered. The sections were subjected to Immunohistochemistry with markers CD34(QBEnd/10) and CD31(JC/70A).
DISCUSSION
Cystic lymphangiomas are rare benign malformations of the lymphatic system, generally they are common in children with male preponderance (M>F) (Konen et al., 2002). Most commonly occur in neck(75%) and axilla (20%). Approximately 5% are intraabdominal arising from mesentery or retroperitoneum (2%). However they can occur in any location where lymphatics are encountered.

It is speculated that lymphangiomas develop due to sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system, or from abnormal budding of the lymphatic endothelium (Enzinger & Weis, 1995). Other potential causes are thought to include abdominal trauma, localized lymphatic degeneration, radiation and lymphatic obstruction.

Abdominal lymphangioma is unusually classified with other mesenteric and retroperitoneal cysts. There is several classifications of these abdominal cystic tumors, but among them the one based on their origin and histological features is the most commonly used. This classifies them into 6 groups: (a) cysts of lymphatic origin (lymphatic cysts and lymphangiomas),(b) cysts of mesothelial origin (benign or malignant mesothelial cysts), (c) dermoid cysts, (d) enteric or duplication cysts, (e) cysts of urogenital origin and (f) pseudocysts (infectious or traumatic) (Ahmid, 2004).

Intra abdominal lymphomas are two types- cystic lymphangioma and lymphocystic.Cysticlymphangioma mostly occurs in the first decade of life with a female predominance while lymphatic cysts in later life.the estimated incidence is about 1/20,000 children (Tandon et al., 2012). Histologically both have endothelial lining, but the wall in cystic lymphangioma contains smooth muscle tissue while that of lymphatic cyst does not. cysticlymphangiomas are more prone for recurrence and infiltration while lymphatic cysts tend to be localized.

Intra-abdominal lymphangiomas are of four types. Type I, the pedicled type, enlarge rapidly and cause torsion. Type II, the sessile type, is located within the mesentery of the bowel. Type III, the retroperitoneal type, involves the retroperitoneal structures, such as the mesenteric root, the aorta and the vena cava. The type IV multicentric type extensively involves the intra-abdominal and retroperitoneal organs (Losanoff et al., 2003).

Cystic lymphangioma should also be differentiated from cystic mesothelioma and Lymphangiomyomas.they have similar histological appearance. Immunohistochemically, lymphatic endothelium is positive for CD31,CD34 and D2- 40, Factor VIII related antigen, endothelial receptor- 1, vascular endothelial growth factor-3, prox-1 expression and negative for cytokeratins (Hornick et al., 2005). Calretinin and HMB-45 can differentiate lymphangiomas from multi-cystic mesothelioma and lymphangiomyoma respectively.

The treatment of abdominal lymphangioma is complete radical resection, as with an incomplete resection, one faces the danger of a recurrence with tendency for invasive growth.

CONCLUSION
Herein we report this case of Intraabdominal Mesentric Lymphangioma occurring in young female as it is rarely encountered and differential diagnosis of all the cysts of lymphatic origin should be included. Clinical history and IHC will help in final diagnosis. Cysticlymphangiomas should be included in differential dignosis of cystic abdominal masses presenting with acute abdomen. It is advisable to go for a complete surgical resection to avoid recurrence with follow up.

REFERENCES