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An unusual site of retroperitoneal lymphangioma

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Abstract

Background: Lymphangiomas are isolated malformation of lymphatic vessels. 65% of cystic lymphangiomas occur at birth, with 90% occurring in patient aged <2years, commonly found in the neck, axilla and rarely seen in abdomen.

Case Presentation: A 14year old female child presented with complains of colicky abdominal pain in the past 5 days, localized to the peri-umbilical region. On examination patient had tenderness in the umbilical region. Ultrasonogram of abdomen probably an infected mesenteric cyst. CECT abdomen done d/d – enteric duplication cyst, mesenteric cyst, choledochal cyst. Patient underwent a laparotomy and complete cyst excision. Patient had an uneventful post-operative recovery and was discharged on day 5. Histopathology of the swelling was suggestive of cystic lymphangioma.

Conclusion: Abdominal Lymphangiomas is a rare condition which is usually asymptomatic and incidentally diagnosed. We are reporting this case because of its rare location that is retroperitoneal paraduodenal, closely mimicking duplication cyst and with varied differential diagnosis, which has different management options.

Keywords: Cystic lymphangioma, duodenal lymphangioma, abdominal mass

Introduction

Lymphangiomas are isolated malformation of lymphatic vessels that have been disconnected from the normal lymphatic system ^[1, 2]. 65% of cystic lymphangiomas occur at birth, with 90% occurring in patient aged <2years, commonly found in the neck, axilla and rarely seen in abdomen ^[1, 3]. Abdominal lymphangiomas are often asymptomatic. The content of the cyst may be infected; therefore, the cyst must be resected without spillage of the cyst fluid into the abdominal cavity.

Case report

A 14year old female child presented with complains of colicky abdominal pain in the past 5 days, localized to the peri-umbilical region. On examination patient had tenderness in the umbilical region in a otherwise soft abdomen. No mass was palpable. Complete hemogram, liver function test and renal function tests were within normal limits. Ultrasonogram of abdomen showed a 9*5cm cystic swelling with heterogenous content in the right hypochondrium, probably an infected mesenteric cyst. CECT abdomen done d/d-enteric duplication cyst, mesenteric cyst, choledochal cyst.

Patient underwent a laparotomy and complete cyst excision. Intra operatively the medial wall of the cyst was tightly adherent to the lateral wall of 2nd part of duodenum (Figure 2). Cyst was removed along with a part of serosa of the duodenum (Figure 3). Patient had an uneventful post-operative recovery and was discharged on day 5.

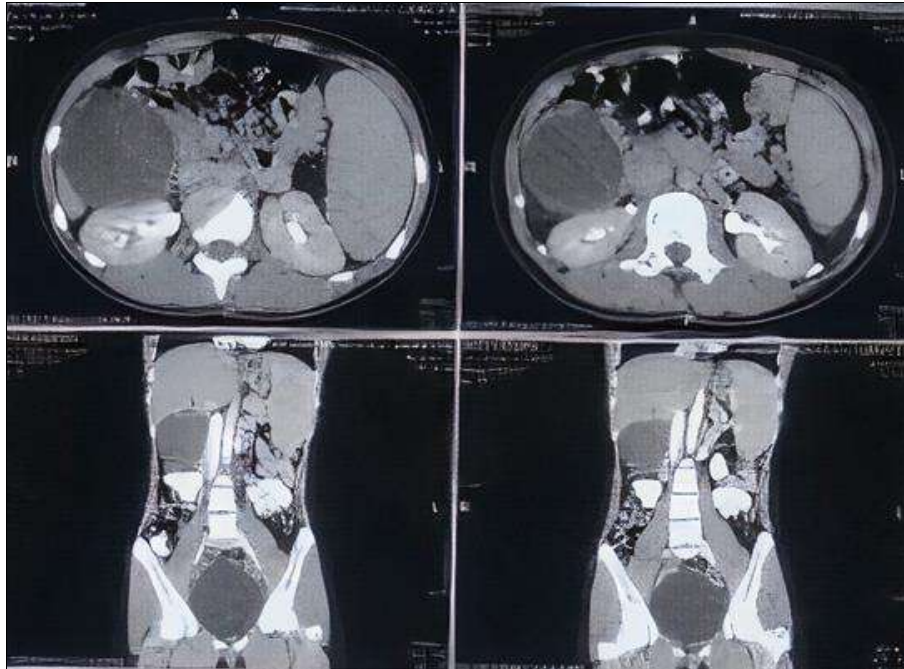


Fig 1: CECT abdomen

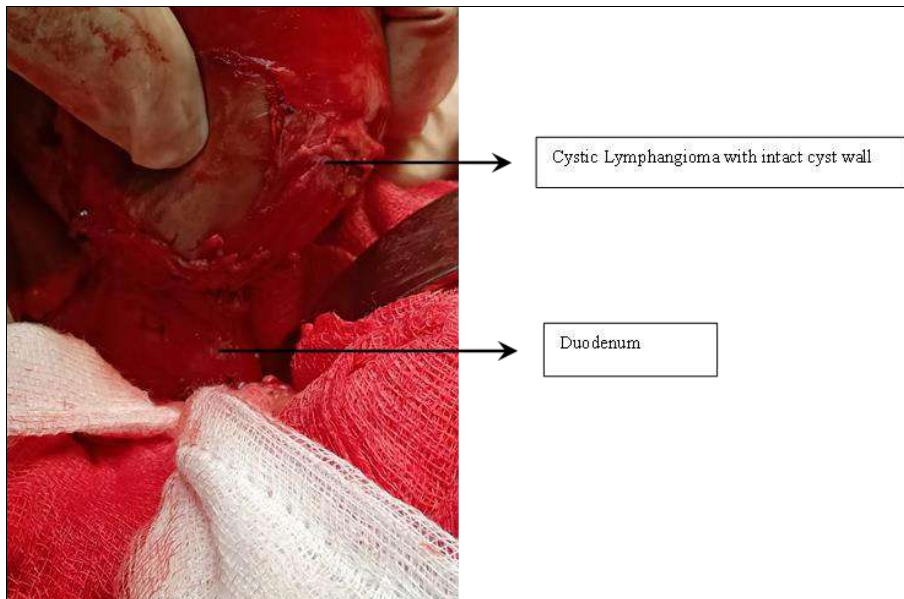


Fig 2: Intra operative: duodenum attached to the cyst wall.



Fig 3: Size of the intact cyst

Histopathology of the swelling was suggestive of thin walled cyst, devoid of lining epithelium. Cyst wall showed lymphoid follicles, interspersed dilated and congested sinus like spaces and hemosiderin laden macrophages (Figure 4 &

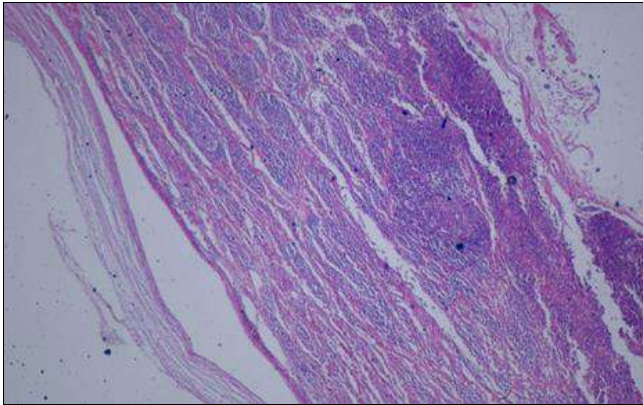


Fig 4: Cyst wall with lymphoid tissue

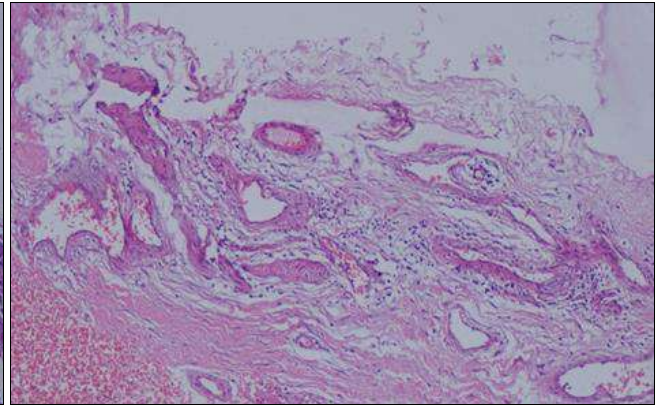


Fig 5: Cavernous vascular channels

Discussion

Peripheral lymphatic systems develop from primitive bags of the venous system. During embryogenesis, a default connection between lymphatic channels and venous system cause formation of lymphatic cyst [1,2].

Half of the cystic lymphangiomas would be present at birth, whereas 90% of cystic lymphangiomas would grow until the age of 2 years. The cystic lymphangioma represents 7% of abdominal cystic lesions in adult; it may be related to the gastro- intestinal tract, spleen, liver, kidney, adrenals, and pancreas (1). In gastro- intestinal tract it is most commonly seen in small bowel (85%), mesocolon (10%), and retroperitoneum (5%) [1,4].

Abdominal lymphangiomas are often asymptomatic. In symptomatic patient abdominal pain is the most common symptom. However, in case of large growth it can also present with palpable mass, intestinal obstruction, or a volvulus. Men and women are affected similarly in adulthood. Ultrasonography is useful initially. CT scan is more specific in the diagnosis and detailing of the morphology and character of the lesion. The presence of sediment at the bottom of the cyst is a sign highly suggestive of the diagnosis of cystic lymphangioma [5].

The differential diagnosis includes malignant tumors such as cystic mesothelioma (multilocular), teratoma (presence of fat and calcification), Mucinous cystadenoma (unilocular) and benign tumors such as lymphangioma, cysts of urothelial and foregut duplication cysts [1,6].

Histopathological examination is the final proof of diagnosis of cystic lymphangioma. Cyst wall has an attenuated endothelial lining, surrounding rich adipose tissue and scattered smooth muscle fibers, small lymphoid aggregates appears focally [1,5].

Asymptomatic patients should be followed by repeated imaging. The aspiration of the cyst with or without injection of sclerosing agent has long term affects, such as frequent recurrence. Open or laparoscopic surgery is an option for cystic lymphangioma especially in its abdominal location. Complete excision should be attempted, making sure not to affect other organs. When cystic lymphangioma is found in solid organs, partial or total excision of cyst with or without associated parenchymal resection maybe considered [2-4,7].

5). Histopathological diagnosis of cystic lymphangioma had been reported. Child was asymptomatic and had a normal sonogram at 3 months of follow-up.

Conclusion

This case was reported in view of its abdominal retroperitoneal paraduodenal location, which is rarely reported in literature, patient presented with vague clinical symptoms and posed a diagnostic dilemma. Radiological scan showed multiple differential diagnoses, closely mimicking duplication cyst and with varied differential diagnosis, which have different management options.

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