Retroperitoneal cystic Lymphangioma in adulthood – A case report

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Abstract

Background: Cystic lymphangioma is a benign lymphatic system malformation, rarely diagnosed in adulthood. Head and neck are commonly affected and abdominal location is rare. Its presentation ranges from asymptomatic to severe symptoms (bleeding, rupture, infection, volvulus). The diagnosis is often made after surgery and confirmed by histopathological examination.

Methods: A case report of a 57-year healthy female patient presented with abdominal pain, postprandial fullness, sporadic vomit and imaging revealing a 9-centimeter retroperitoneal cyst is described.

Results: She was submitted to surgery and histology confirmed the diagnosis of cystic lymphangioma.

Conclusion: Retroperitoneal cystic lymphangioma is a very rare disease. Preoperative diagnosis is challenging and definitive treatment is surgical resection in symptomatic patients.

Background

Cystic lymphangioma is a benign malformation of the lymphatic system. Congenital lesions occur when primary lymphatic cysts fail to converge with the main lymphatic system.[1, 2] Acquired lesions are caused by obstruction between lymphatic and venous systems due to inflammation, trauma or degeneration.[3] Head and neck are more commonly affected (75%), followed by axillae (20%) and abdomen (5%).[4] Amongst abdominal cases, mesentery is the main location but the gastrointestinal tract, spleen, liver, kidneys and adrenals may also be affected.[3] Retroperitoneal location is even rarer (1%).[4] Most of cystic lymphangiomas (60%) are diagnosed in children less than 5 years of age.[3] To the best of our knowledge, only about 200 cases have been described in adults so far, but the real prevalence may be underestimated due to nonspecific clinical presentation and difficult recognition of the disease.[2, 4] Patients with slowly growing lesions, mainly in retroperitoneal location, may present an asymptomatic palpable mass in the abdomen.

Abdominal pain is the commonest symptom in bigger tumors, but abdominal distension and constipation can also occur. Uncommon complications that may cause acute abdomen situations are intracystic or gastrointestinal bleeding, infection, cystic rupture and volvulus.[3] This report aimed to present the case...
of a retroperitoneal cystic lymphangioma in adulthood and a brief review of the literature.

Case report

A 57-year healthy female patient presented to the clinic with a nonradiated, moderate and constant epigastric abdominal pain. She referred also postprandial fullness and sporadic vomit, but denied weight loss and gastrointestinal bleeding. Physical examination was innocent. Laboratory work-up (including blood count, liver and pancreatic enzymes and carcinoembryonic antigen) was unremarkable, as well as esophagogastroduodenoscopy. Abdominal ultrasonography revealed a 9-centimeter cystic lesion, located inferiorly to the left kidney and dislocating the aorta. Computed tomography (CT) excluded renal and aortic invasion and suggested a possible relation with distal duodenum (Figure 1). Patient was submitted to an uneventful open, anterior and total resection of the cystic lesion, which did not invade adjacent structures (Figure 2 and 3). Patient did well in the postoperative period and was discharged in day 4. Histology revealed a cystic lesion with thin and translucent wall with 9.5-centimeter of maximum diameter. Immunohistochemical study was compatible with cystic lymphangioma with endothelium staining for podoplanin (D2-40) but not for calretinin (Figure 4 and 5). Patient remains asymptomatic and with no evidence of recurrence during 42 months of follow-up.

Discussion

Preoperative diagnosis of retroperitoneal lymphangioma is difficult and it is frequently an incidental radiological finding. Ultrasound often shows a well limited, simple or multilocular cyst with thin septation and clear fluid or hyperechogenic content if bleeding or calcifications exist. CT allows a better assessment of the relation with adjacent organs. Cystic content is better characterized by Magnetic Resonance Imaging as well as perivascular extension of the lesion.[2, 3] This patient declined Magnetic Resonance because of “claustrophobia sensation” and diagnosis was not possible preoperatively, despite ultrasound and CT.

Differential diagnosis of abdominal cystic lymphangioma include lymphoma, cystic mesothelioma, teratoma, sarcoma, lymphangioma, adenoma, hematoma, abscess, duplication cyst, ovarian cyst, postoperative lymphocele, lymphadenopathy, ovarian or gastric cystic metastases. In spite of excellent imaging tools, diagnosis of cystic retroperitoneal lymphangioma is often made after surgery only and after confirmation by histopathological examination. Diagnostic criteria are dilated lymphatics lined with flat endothelium rich in lymphoid tissue with no atypical cells.[3, 4] Immunohistochemically, cystic lymphangioma endothelial cells express factor VIII-related antigen, CD31 and CD34, but negative staining with cytokeratin [1, 5], as

Figure 1A and 1B: CT scan and Intraoperative aspect of the lesion
Asymptomatic patients can be proposed to conservative treatment reserving surgery for symptomatic patients, considering the absence of final diagnosis. Percutaneous drainage or aspiration with or without injection of sclerosant agents present a high rate of recurrence and must be reserved for patients not fit for surgery.[2, 3] Surgery (open/laparoscopic, retroperitoneal/anterior) is the definitive treatment and it may be limited by tumor location and relation with adjacent structures. Lymphostasis must be meticulous to avoid complications as lymphocele or chylous ascites. Recurrence can achieve 17–40% depending on total or partial resection.[2, 3] In the present case, the presence of symptoms, probably due to extrinsic compression of duodenum, the lack of diagnosis, and the size of the lesion, a laparotomy was proposed, which allowed the complete resection of the lesion through an anterior approach.

**Conclusion**

Retroperitoneal cystic lymphangioma is a benign disease, very rare in adults, that must be considered as a differential diagnosis of intra-abdominal cystic lesions. Preoperative diagnosis is challenging and in symptomatic patients definitive treatment is surgical resection.

**Main novel aspects:** We present a rare disease, even rarer in an adult, and in an uncommon location

**References**