Giant Retroperitoneal Lymphangioma: a Rare Entity

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Abstract Retroperitoneal lymphangiomas are rare benign tumors, and the exact incidence of which is not well known. They usually arise from the developmental abnormalities of retroperitoneal lymphatics or their traumatic rupture. Confusion with other cystic tumors of the retroperitoneum including those arising from the kidney and pancreas is common. They usually lack specific symptoms and are mostly detected accidentally, while very rarely, some may present with complications such as rupture, torsion, hemorrhage, or infection. We report a case of a 22-year-old female in whom preoperative diagnosis of retroperitoneal lymphangioma was made on the basis of radiological investigations. On exploration, a large lymphangioma arising retroperitoneally was identified and excised. Postoperative recovery was uneventful.

Keywords Lymphangioma · Retroperitoneum · Cystic abdominal mass · Surgical excision

Introduction

Retroperitoneal cysts are rare intra-abdominal cystic masses, and less than 1 % of lymphangiomas affect the small bowel mesentery, omentum, or retroperitoneum [1]. These intra-abdominal lymphangiomas are usually asymptomatic, or they

can present rarely with complications like infection, bleeding, torsion, or hemorrhage [2]. Preoperative diagnosis is usually made on the basis of USG, CT, or MRI abdomen which shows characteristic features [1]. Surgical excision is the preferred modality of treatment [1]. Here, we report a rare case of lymphangioma arising from the retroperitoneum. Complete surgical excision was done, and patient recovered well.

Case Report

A 22-year-old lady presented with history of central abdominal pain for 2 months. There was no history of bowel or bladder complaints. Per abdominal examination was normal. On ultrasound abdomen, a large thin-walled cystic lesion with fine septations was present in the retroperitoneum, occupying almost whole of the left abdomen and displacing bowel toward the right side. No solid component was seen. The pancreas, left kidney, and bilateral ovaries were seen separately and were normal. Contrast-enhanced computed tomography (CECT) abdomen revealed an approximately $22 \times 12 \times 12$ -cm ill-defined solitary cystic lesion in the retroperitoneum, occupying the left lumbar and iliac fossa region, crossing midline, and displacing bowel loops toward the right side and anteriorly (Fig. 1a). No calcifications and no invasion of surrounding structures were present, suggesting it to be a retroperitoneal lymphangioma. The patient subsequently underwent exploratory laparotomy which revealed a lobulated cystic mass arising from retroperitoneum which was causing a bulge in the mesentery and displacing the overlying small bowel loops to the opposite side (Fig. 2a). The cyst extended superiorly till the tail of pancreas and inferiorly till the distal sigmoid colon and was crossing the midline. Clear serous fluid was present within it (Fig. 2a). No invasion into the surrounding retroperitoneal structures was present.

Complete surgical excision was done, and postoperative recovery was uneventful. On histopathology, diagnosis of a cystic lymphangioma was confirmed.

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Fig. 1 a CECT abdomen showing a large retroperitoneal mass with displacement of bowel loops

Discussion

The differential diagnosis of a cystic tumor in the retroperitoneum includes both malignant and benign tumors. Malignant causes include germ cell tumors (teratoma), undifferentiated sarcoma, cystic metastases from the stomach or ovary, malignant mesenchymoma, biliary cystadenoma/carcinoma, and cystic mesothelioma [3]. Benign cystic lesions of the retroperitoneum include lymphangioma, microcystic pancreatic adenoma, and cysts of urothelial and foregut origin. Cysts of foregut origin include bronchogenic cysts, which contain cartilage or seromucinous respiratory glands; esophageal cysts, which are composed of well-developed layers of smooth muscle without cartilage; and simple foregut cysts [4].

Lymphangiomas are extremely rare benign neoplasms of the lymphatic system. The exact incidence is unknown due to their rarity. They can occur in any location where the lymphatics are normally found. Head and neck lymphangiomas (cystic hygromas) account for 75 % of reported cases followed by axilla (20 %) [4]. Retroperitoneal lymphangiomas account for approximately 1 % of all lymphangiomas. Hauser et al.

Fig. 2 a Intraoperative image showing a cystic retroperitoneal lesion. **b** Intraoperative image of retroperitoneal cyst showing clear serous fluid

have identified merely 185 cases in a review of the international literature [5].

The exact etiology of lymphangioma remains unclear. Congenital malformation of lymphatic vessels leading to sequestration of lymphatic tissue appears to be the primary etiology [5]. The various etiologies implicated in its pathogenesis include genetic predisposition, mechanical pressure and retention, traumatic factors, degeneration of lymph nodes, and disorders of endothelial lymphatic vascular secretion or permeability. However, the frequent development within areas where primitive lymph sacs occur suggests that lymphangiomas are malformations arising from sequestrations of lymphatic tissue that fail to communicate normally with the lymphatic system [6]. This results in the formation of uni- or multicystic tumors whose cavity is covered with a layer of endothelium and filled with chylous or serous material.

There are three types of lymphangioma commonly identified: capillary, cavernous, and cystic. Cystic type is the one commonly found intra-abdominally or retroperitoneally [5]. These consist of cystic areas covered by a single endothelial layer of foam cells and have a wall consisting of lymphatic spaces, lymphatic tissue, and smooth muscle cells [6]. The presence of lymphatic vessels in their wall distinguishes it from simple mesenteric cyst.

The clinical presentations of lymphangiomas are varied and depend largely upon their size and site. These range from largely asymptomatic state to acute-onset severe abdominal pain [3].

Abdominal ultrasound is a specific initial diagnostic test for intra-abdominal lymphangioma, which shows a well-defined mono- or polycystic lesion with internal septations [4]. Abdominal CT or MRI further characterizes the mass with respect to its size, location, involvement of other organs, and nature of its fluid [4]. Preoperative tissue diagnosis is generally not advised.

Treatment options range from injecting sclerosants to surgery. Surgery is the treatment of choice with en masse removal having excellent results [7]. Dissemination in the retroperitoneum is very rare, but a potentially life-threatening complication [8]. Injection of sclerosants such as alcohol and bleomycin into lymphangiomas has been described in the literature in nonsurgical candidates. However, induration of the cyst and infection often complicate these procedures.

Conclusion

Retroperitoneal cystic lymphangioma is an uncommon entity, and it should be included in the differential diagnosis of cystic abdominal lesions. The usual presentation is an asymptomatic

mass, but rarely, complications can be seen. The optimal treatment today is complete surgical excision.

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