Case Report

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Retroperitoneal lympangioma in an adult - a case report

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Abstract:

Lymphangiomas are rare, benign, hamartomatous congenital malformations of the lymphatic system that may involve any part of the body. Retroperitoneal lymphangiomas account for less than 1% of cases which may present as a palpable abdominal mass and can cause diagnostic dilemma with other retroperitoneal cystic tumors, including those arising from the liver, kidney and pancreas. Due to rarity and difficulties in diagnosing preoperatively case has been reported.

Key words: Lymphangioma, Retroperitoneal cystic tumors, Retroperitoneal lymphangioma

Introduction:

Lymphangiomas are a congenital malformation of the lymphatic system, with less than 1% arising in the retroperitoneal space [1]. The first case of retroperitoneal cystic lymphangioma was reported by Gaudier and Gorse in 1913 [2]. These may be capillary, cystic or cavernous, with a uniseptate or multiseptate appearance [3]. True retroperitoneal lymphangioma fulfilling the criteria for pathological diagnosis are relatively rare and are usually found incidentally during surgery, autopsy or lymphography. An interesting and rare case of a retroperitoneal cystic lymphangioma in a 45-year-old female patient is described here.

Case report

A 45-year-old woman came with complains of abdominal discomfort and a mass in the right side of the abdomen of 3 months duration with no history of vomiting, loss of appetite, bowel or bladder disturbances. Physical examination revealed 14×10cm non-tender, freely-mobile; firm mass over the right flank. On head raise test, the mass became less prominent and on knee-elbow position it disappeared which confirmed its retroperitoneal location. There was no organomegaly or lymphoedema of the extremities. The external genitalia were normal. Per rectal and per vaginal examination were unremarkable.

Routine laboratory investigations were within normal limits. Radiography of the kidney/ureter/bladder area showed a huge mass occupying the right flank of abdomen. Ultrasonography of the abdomen revealed that the mass was cystic in nature with multiple septa and an irregular margin. A computed tomography (CT) scan of abdomen and pelvis showed a large 14.4 cm × 14.6 cm × 9.2 cm multilocular cystic mass with enhancing septations. The attenuation coefficient of the mass was in the +10 to 15 hu range. The mass was well-circumscribed in its entirety, and there was no evidence of an invasive component.

Surgical exploration was done by lower paramedian incision, a huge multilocular cystic mass, inferior to the right kidney was found, confined in the retroperitoneal space, slightly pushing the right ureter medially. The mass was dissected out carefully without injuring the surrounding structures and the great vessels (Figure 1).

Gross examination revealed a mass of shiny, smooth surface of 12×10×2cm^3, and sectioning revealed multiple cysts filled with cloudy fluid (Figure 2). On histology, the mass was composed of variable-sized cystic spaces lined by flattened endothelium. The larger spaces had fascicles of smooth muscle. Small lymphoid aggregates and reactive lymph nodes were present. The stroma showed acute and chronic inflammation, oedema and fibrosis (Figure 3). Thus, the diagnosis of a

retroperitoneal multilocular-cystic lymphangioma with inflammation was histologically confirmed.

Figures:

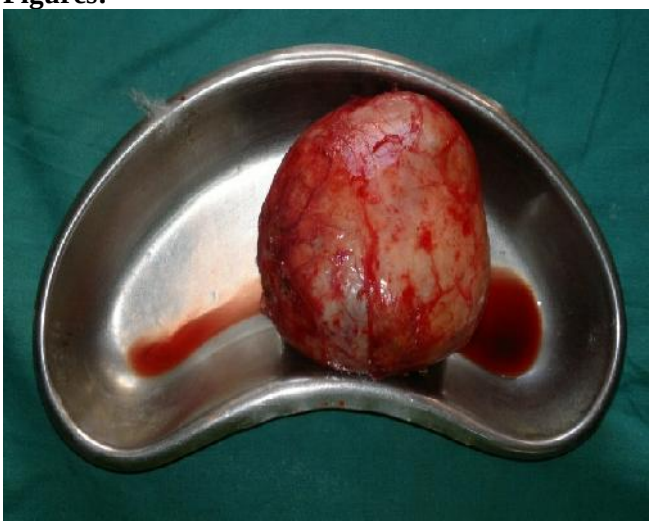

Figure 1: Gross specimen of retroperitoneal lymphangioma.

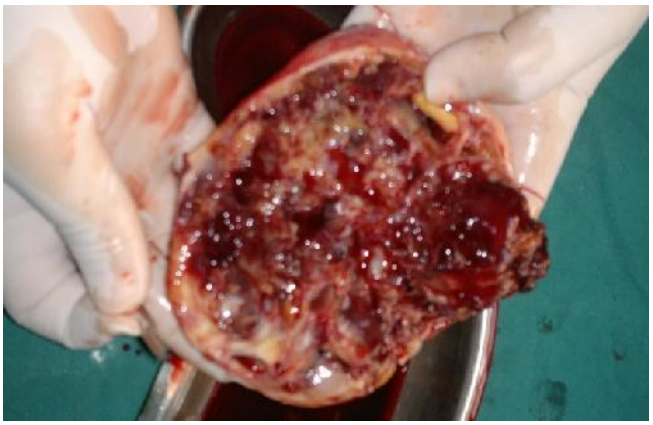

Figure 2: cut section showing multiple cysts filled with cloudy fluid.

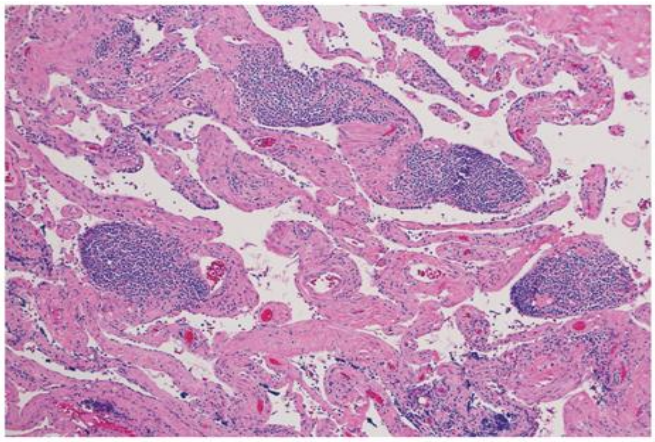

Figure 3: HPE showing cystic spaces lined by flattened endothelium with lymphoid aggregates.

Discussion

The lymphatic system is developed during the third or fourth fetal month from 2 paired and unpaired endothelial channels proliferate centrifugally from these sacs, which are located in the neck, mesenteric root, and bifurcation of the femoral and sciatic veins [4]. Early sequestration of lymphatic vessels that fail to establish connections with normal draining lymphatics gives rise to lymphangioma. Lymphangiomas are congenital and benign. As a result of budding of preexisting spaces and collection of fluid they enlarge, form unilocular or multilocular cystic masses and can encroach on vital structures.

Approximately 50% of lymphangiomas are present at birth, and almost 90% are diagnosed before the age of 2 years and most frequently affected sites are the head and neck (75%) followed by the axilla (20%). The remainder (approximately 5%) of the lymphangiomas is intra-abdominal arising from the mesentery, retroperitoneum or greater omentum. The retroperitoneum is the second-most common location for the abdominal lymphangiomas after mesentery of the small bowel which develops due to an abnormal connection between the iliac and retroperitoneal lymphatic sacs and the venous system, leading to lymphatic fluid stasis in the sacs. As no definite diagnostic tests are available, these present a diagnostic dilemma. These are usually of cavernous or cystic types [5], of which most reported cases have been of a cystic nature, as was this case.

Most retroperitoneal lymphangioma are asymptomatic, and are discovered incidently on routine imaging, surgery or in autopsy. Most common presentation is slowly growing mass, as seen in our case.

Pre-operative diagnosis of retroperitoneal lymphangioma, in general, is challenging and rare, prior to laparatomy or laparoscopy. Our primary radiological investigations did include the possibility of a retroperitoneal lymphangioma. The likelihood of pre-operative diagnosis is greatest when imaging is combined with biopsy. But guided biopsy of the lesion is not attempted, due to the threat of potential dissemination of malignancy if any.

Final diagnosis of lymphangioma is made by pathological examination of the specimen after surgical excision, and is based on well-established Criteria. These include a well-circumscribed, cystic lesion, with or without endothelial lining, a stroma composed of a meshwork of collagen and fibrous tissue, and a wall containing focal aggregates of lymphoid tissue [5]. Accordingly, Histology from the mass in this case showed variable-sized cystic spaces

lined by flattened endothelium. The larger spaces had fascicles of smooth muscle. Small lymphoid aggregates and reactive lymph nodes were present. The stroma showed acute and chronic inflammation, oedema and fibrosis.

Lymphatic vessel endothelial receptor- 1, vascular endothelial Growth factor-3, prox-1, and Monoclonal Antibody d2-40 are the markers for specific endothelial cells of lymphatic tissues that are used in immunochemical studies of lymphangioma. As the patient was not affordable immunchemical studies were not done.

As it is of benign nature, retroperitoneal lymphangioma warrants a conservative surgical approach such as aspiration; however, in most, surgery is required, due to the morbidity associated with the tumor size, infrequent spontaneous regression of the cyst, and for definitive diagnosis [6]. For cystic retroperitoneal lymphangioma, a simple total excision is preferred treatment to avoid complications like superinfection, progressive growth, rupture or bleeding. Historically described procedures such as cyst-enterostomy or marsupialization have now become obsolete. In this case simple total excision of the cyst was done, which seemed an appropriate treatment. Patient tolerated procedure well without any clinically significant postoperative course and was discharged on post operative day 6.

Recurrence after surgery is rare, but it increases following conservative treatment modalities like aspiration, drainage and sclerosis. Recurrence does occur if there is incomplete removal of the lesion. Dissemination in the retroperitoneum is a very rare, but potentially a fatal complication [7].

Conclusion

Retroperitoneal lymphangioma is an uncommon lesion in adults, and radiological investigation like ultrasound provides important preoperative diagnostic information for effective surgical approach and management. Authors emphasize the need of keeping in mind retroperitoneal lymphangioma as different possible diagnoses of a retroperitoneal cystic lesion. These benign tumours have an excellent prognosis and cure can be achieved with complete surgical excision.

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References:

1. Hauser H, Mischinger HJ, Beham A, Berger A, Cerwenka H, Razmara J, Fruhwirth H, Werkgartner G. Cystic retroperitoneal lymphangiomas in adults. Eur J Surg Oncol 1997; 23: 322-326.
2. Gaudier H, Gorse P. Lymphangiome kystique abdoménoscrotal retropéritonéal. Presse Méd 1913;21:458–89.
3. Bhavsar T, Saeed-Vafa D, Harbison S, Inniss S. Retroperitoneal cystic lymphangioma in an adult: a case report and review of the literature. World J Gastrointest Pathophysiol 2010; 1(5): 171-176.
4. Wilson SR, Bohrer S, Losada R, Price AP. Retroperitoneal lymphangioma: an unusual location and presentation. J Pediatric Surg. 2006;41:603–605.
5. Koshy A, Tandon RK, Kapur BM, Rao KV, Joshi K. Retroperitoneal lymphangioma. A case report with review of the literature. Am J Gastroenterol 1978; 69: 485-490.
6. Enzinger FM, Weis SW. Tumors of lymph vessels. In: soft tissue tumors. St. Louis: Mosby-Years Book, 1995: 679-700.
7. Nishio I, Mandell GL, Ramanathan S, Sumkin JH. Epidural labor analgesia for a patient with disseminated lymphangiomatosis. Anesth Analg 2003; 96: 1805-1808.