

Case Report

Giant cystic retroperitoneal lymphangioma in an adult

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Received: January 17, 2013

Accepted: January 22, 2013

Published Online: January 28, 2013

DOI: 10.5455/jihp.20130122045401

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Keywords: Retroperitoneum, Cyst,
Giant, Lymphangioma

Abstract

Lymphangioma is an uncommon congenital malformation of the lymphatic system affecting usually the younger age group. These tumours frequently affect the neck and axillary region. Retroperitoneal lymphangiomas are rare and cause a clinical and pathological diagnostic challenge. Histopathological examination of the tumour is imperative for an accurate diagnosis. We report a case of a giant cystic retroperitoneal lymphangioma in an adult presenting as a mass in the left hypochondrium. Radiology revealed a large cystic mass in the retroperitoneum displacing the large bowel and compressing the left ureter. The mass was excised surgically and there is no evidence of recurrence on follow up.

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INTRODUCTION

Lymphangiomas are rare benign neoplasms characterized by the presence of proliferating lymphatic vessels. The most common sites involved are the neck, axilla and mediastinum accounting for 95% of the cases. Retroperitoneal lymphangiomas are rare lesions with incidence of approximately 1% [1, 2]. Lymphangiomas usually manifest within two years of age, however, retroperitoneal and intra-abdominal lymphangiomas become symptomatic in adulthood due to its slow progressing nature [3]. We report a rare case of giant retroperitoneal lymphangioma in a 51 year old male who presented with a short history of mass in the hypochondrium.

CASE REPORT

A 51 year old male presented with a 5 day history of painless, slowly progressive mass in the left hypochondrium. There was no history of fever, jaundice, weight loss, diarrhea or constipation. His general and systemic examination and laboratory investigations were within normal limits. Computed tomography of the abdomen revealed a large well

defined cystic lesion arising from the retroperitoneum at T10 –T11 level, anterior to the aorta, in the anterior pararenal space displacing the bowel loops and mesenteric vessels (**Figure 1**). Posteriorly, the lesion was seen causing compressing the left ureter and pelvicalyceal system resulting in moderate hydronephrosis. The lesion did not show any solid enhancing component, obvious calcification, hemorrhage or fat component. The features were suggestive of a benign cystic lesion of the retroperitoneum. Exploratory laparotomy with complete excision of the cyst was performed and the specimen was sent for histopathological examination (**Figure 2**). Grossly, the specimen consisted of a single cystic mass weighing 450 gram and measuring 12x6cm. Cut section showed a multiseptate cyst filled with serous fluid (**Figure 3**). On microscopy, large ectatic vascular channels lined by attenuated cells were seen with disorganized smooth muscle bundles and focal lymphoid aggregates in the wall (**Figure 4**). The features were consistent with giant cystic retroperitoneal lymphangioma. Follow up after six months, the patient is asymptomatic.

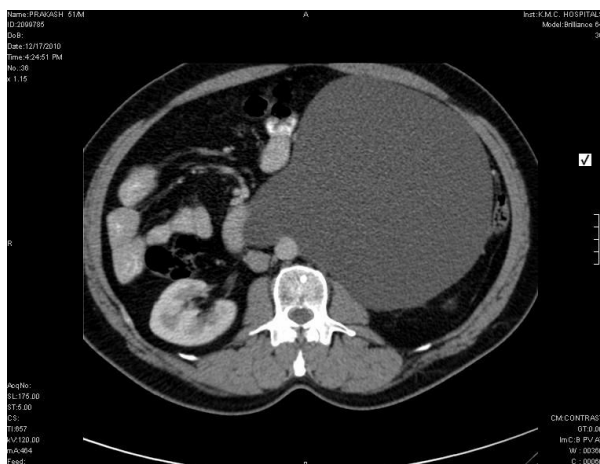


Figure 1. CT abdomen showing a large well defined retroperitoneal mass.

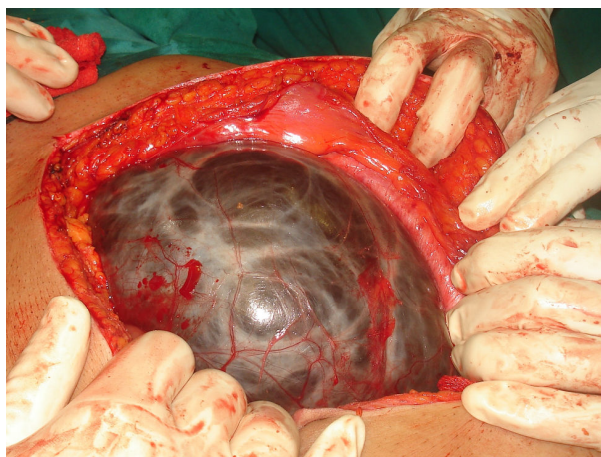


Figure 2. Intraoperative photograph showing a retroperitoneal cystic mass.

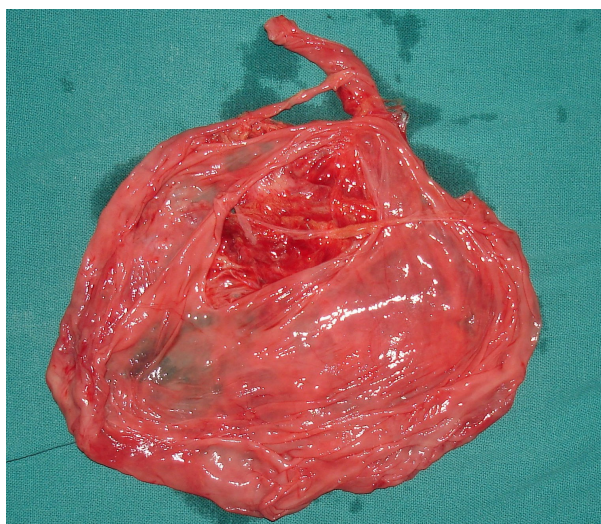


Figure 3. Multiseptate cystic mass.



Figure 4. Microphotograph shows large ectatic vascular channels lined by attenuated cells surrounded by stromal lymphocytic aggregates (H&E, x200).

DISCUSSION

Lymphangiomas are regarded as malformations that arise from sequestration of lymphatic tissue that fail to communicate normally with the lymphatic system. It is postulated that lymphangioma is an early developmental sequestration of lymphatic vessels that do not establish connections with the draining vessels during the intrauterine life. Fewer than 5% occur in the abdominal cavity of which the retroperitoneum is the least common site [1, 4].

The clinical presentation of lymphangioma depends on the anatomic location and tumor size [5]. Large tumors may present as asymptomatic masses, most often detected incidentally by radiology or surgery for other causes [6]. These tumors may cause symptoms due to compression of adjacent structures leading to a diagnostic consideration of sarcomas. Abdominal lymphangiomas most often present as acute abdomen mimicking intestinal obstruction or appendicitis [6, 7].

Radiology has a limited role in the preoperative diagnosis of lymphangiomas. Ultrasonography and CT reveal multiseptate or multilocular cystic masses. A urogram may demonstrate displaced kidney or ureters [3, 8].

The final diagnosis of lymphangiomas is based on histopathological examination of the tumor. It comprises dilated and interconnected vascular channels lined by flattened endothelial cells with intervening connective tissue septae containing smooth muscle fascicles, lymphocytes and adipose tissue [6]. Histologically, lymphangiomas are classified as capillary, cavernous and cystic lymphangiomas. Retroperitoneal lymphangiomas are commonly cystic [4].

Differential diagnosis of cystic retroperitoneal lymphangiomas include benign lesions such as cysts of urothelial and foregut origin, microcystic pancreatic adenoma, retroperitoneal hematoma, abscesses, duplication cysts, ovarian cysts and pancreatic pseudocysts, malignant tumors such as cystic mesothelioma, teratoma, undifferentiated sarcoma, cystic metastases (especially from ovarian or gastric primaries) and malignant mesenchymoma. Histopathological features in conjunction with lymphatic immunohistochemistry markers like lymphatic vessel endothelial receptor-1, vascular endothelial growth factor-3, prox-1, and monoclonal antibody D2-40 aids in accurate diagnosis [7, 9].

The treatment of choice for retroperitoneal lymphangiomas is complete surgical resection, however, aspiration and injection of sclerosing agents have also been recommended. Due to the high incidence of recurrence, a regular follow up is suggested [4, 5].

In conclusion, retroperitoneal lymphangiomas are rare benign neoplasms which may pose a diagnostic dilemma for surgeons and pathologists. However, vigilant clinical, radiological and pathological examination establishes the diagnosis.

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