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Oncology

Retroperitoneal cystic lymphangioma-a case report

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ABSTRACT

Retroperitoneal lymphangiomas (RL) are rare benign cystic tumors of the lymphatic system, which account for 1% of all lymphangiomas, and till now less than 200 cases have been reported. We are presenting a 35 years old woman, who was admitted with symptoms of recurrent left lumbar pain. The preliminary diagnosis from CT scan was retroperitoneal cyst. The patient went under explorative laparotomy and whole cyst mass was removed from the surrounding structures using sharp and blunt dissection.

Introduction

Retroperitoneal lymphangiomas (RL) are rare benign cystic tumors of the lymphatic system, which accounts for 1% of all lymphangiomas, and till now less than 200 cases have been reported. Due to their rarity the preoperative diagnosis if often difficult. Although they can be asymptomatic for a long time, the most frequent clinical symptoms of RL are abdominal or back pain, fever, fatigue, weight loss, and hematuria. Their appearance may be uniseptal or multiseptal capillary, cystic or cavernous tumor mass.

Case presentation

We are presenting a 35 years old woman, who was admitted in Urology Department with symptoms of recurrent left lumbar pain, irradiating towards inguinal area for two days. The patient also complains from nausea and vomiting. She had past episodes of intermittent hematuria. Upon physical examination no abdominal mass was palpated and the bowel sounds were diminished in all quadrants. All laboratory results were in normal ranges.

The contrast computed tomography (CT) scan of abdomen and pelvis defined a large 18.5 cm \times 14.2 cm multiple cystic, confluent masses with retroperitoneal location, which includes the retrocrural and anterior perirenal area (Fig. 1). A bloated stomach was found due to the compression of the duodenum and also reduced size of the left kidney as a result of chronic left renal artery compression. The attenuation coefficient of the mass was between +10 and 15 HU. The preliminary

diagnosis was retroperitoneal cyst.

The patient went under explorative laparotomy and whole cyst mass was removed from the surrounding structures using sharp and blunt dissection. On histological examination was found that the mass contains variable-sized cystic spaces lined by flattened endothelium consistent with lymphatic vessels, which on immunostaining were positive for CD31 and D2-40 and were negative to spectrum keratin and calretinin (Fig. 2). Therefore, the diagnosis of a retroperitoneal cystic lymphangioma was histologically confirmed.

Discussion

Lymphangioma was first described by Koch in 1913. With regards to the etiology of lymphangiomas there is a theory suggests that they develop from a congenital malformation of lymphatic vessels, leading to blockage of lymphatic flow and lymphangiectasia. Three histological types of lymphangiomas are present: cystic, capillary, and cavernous. Retroperitoneal lymphangioma is most commonly of cystic type, like in our presented case.

The clinical signs and symptoms of retroperitoneal lymphangiomas are variable and the diagnosis often can be challenging. At the beginning they are asymptomatic and the first symptoms are abdominal distention, mild abdominal pain, abdominal asymmetry due to enlarging mass. A rare clinical manifestations such as anemia and back pain have also been reported. ^{2,3} Our patient was presented with symptoms of recurrent left lumbar pain and also past symptoms of hematuria.

The treatment of retroperitoneal cystic lymphangioma is surgical

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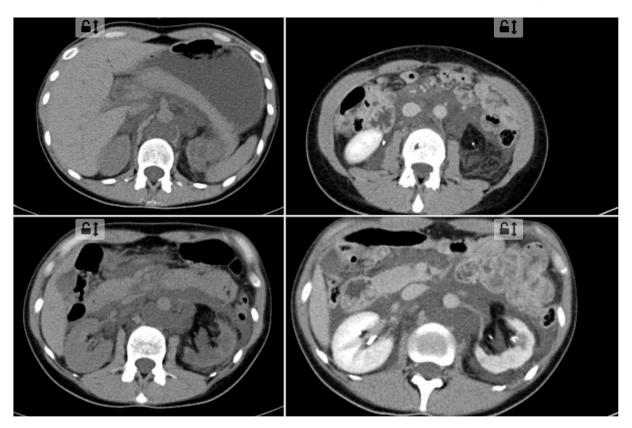
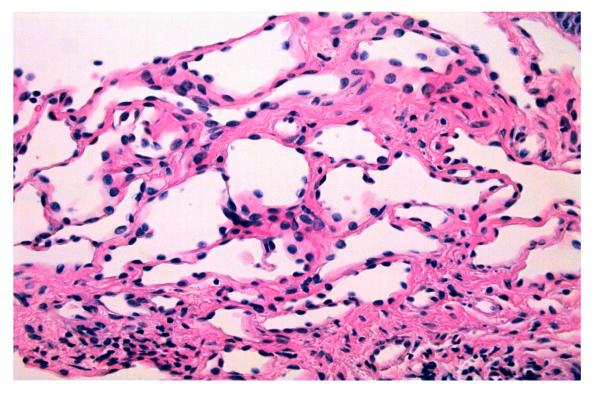


Fig. 1. CT scan of left retroperitoneal lymphangioma.



 $\textbf{Fig. 2.} \ \ \textbf{Histopathology image of retroperitoneal lymphangioma}.$

excision, which can be performed via either laparotomy or laparoscopy. The nonoperative treatment such as aspiration of contents and injection of sclerosant agents is another modality that has been demonstrated to be effective. The total surgical excision is the treatment of choice to

avoid recurrence, progressive growth, infection, rupture, and bleeding. 4,5 Surgical excision was done in our case too.

Conclusions

Lymphangiomas are rarely develop in the retroperitoneum and they are usually asymptomatic. When the retroperitoneal lymphangiomas reach large sizes, symptoms like back pain and hematuria can occur and the method of treatment is total surgical excision.

Declaration of competing interest

The authors declare that they have no competing interests.

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