Retroperitoneal Mass-lymphangiomyoma

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Abstract We report a case of retroperitoneal mass, in a postmenopausal lady, about a size 10–8 cm in left side of abdomen. This turned out pathologically lymphangiomyoma. Retrospectively when we searched in literature we found that, commonly lymphangiomyoma seen in premenopausal women and is associated with ascites or renal angiomyolipoma or plural effusion. But in our case CxR -PA was normal and abdominal USG was not showing renal or ovarian involvement. It was a rare case and presentation was rare than the literature.

Keywords Lymphangiomyomatosis · LAM · Angiomyolipoma · Lymphangiomyoma

Introduction

Lymphangiomyoma is a rare idiopathic disorder, seen in almost exclusively in females, in her premenopausal age group, due to benign proliferation of smooth muscle cells in the lung or in the lymphatics of retro peritoneum, kidney and rarely uterus. Here we present a case of lymphangiomyoma in the retro peritoneum in postmenopausal lady.

Case Report

65 yr old, postmenopausal lady came in surgery opd with history of pedal oedema, pain in left leg since one month, dry cough since two months and history of vomiting 2–3

times during these 2 months. On examination her vitals were normal. There was mild pallor and she had nonpitting pedal oedema over left leg which was tender and her nails were brittle. On per abdominal examination there was single, large 10*8 cm lump in left lumbar, inguinal and periumbilical area which was soft to cystic in consistency and appearing clinically in retroperitoneal area. It was fixed to posterior aspect and was not ballotable and not moving with respiration. Her routine lab investigations and complete blood count was normal. Her abdominal USG was showing 14*12 cm cyst (500 CC) with thick walls, internal echoes, some setae seen in left flank below left renal pole and medially at places it was touching aortic athermatous walls and tail of pancreas with no evidence of ascites. Her CT scan abdomen with contrast and IVU was consistent with USG findings and was showing good function of left kidney with mild hydronephrosis. Her Doppler of both limbs and CXR PA was normal. Considering this as retroperitoneal cyst patient was subjected to laparotomy and she was explored with left paramedian incision. Cyst was in retro peritoneum, reaching up to abdominal aorta and displacing lower pole left kidney and upper ureter, upwards and laterally. Ureter was dissected out and in toto excision of cyst was done and incision was closed after keeping drain in retro peritoneum. The specimen was sent for histopathology. Her postoperative period was uneventful and she was discharged on 8th post op day.

Histopathology

Her histopathology report was showing mass composed of ramifying network of lymphatic spaces lined by a single layer of endothelium, surrounded by fascicles and bundles of smooth muscle cells and occasional lymphoid follicles

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and congested vascular spaces are noted amongst the smooth muscle bundles. It was a multiloculated cyst suggestive of lymphangiomyoma.

DISCUSSION

Lymphangiomyoma is a presentation of extra pulmonary form of lymphangiomyoma either in retro peritoneum (16%) or kidney or abdominal lymph nodes. Patients with lymphangiomyomas (LAM) may present initially as retroperitoneal mass and one or two year later may develop pulmonary lymphangiomyomatosis [6]. LAM is a rare disorder of unknown origin that almost exclusively affects women of childbearing age. It is characterized by proliferation of abnormal smooth-muscle cells (lymphangiomyomatosis cells) in the pulmonary interstitium and along the thoracic and abdominal lymphatics. Lymphangiomyomatosis may be associated with tuberous sclerosis complex [8]. The most common manifestations of lymphangiomyomatosis are pulmonary symptoms including progressive dyspnoea, recurrent pneumothoraces, and chylous effusions [4]. Extra pulmonary lymphangiomyomatosis as the initial presentation of the disease is highly unusual. The most common forms of extra pulmonary lymphangiomyomatosis include renal angiomyolipoma (54% of cases), enlarged abdominal lymph nodes (39%), and lymphangiomyoma (16%). Less commonly, ascites (10%) and hepatic angiomyolipoma (4%) may be present [1, 2]. If the diagnosis of extra pulmonary lymphangiomyomatosis precedes that of pulmonary lymphangiomyomatosis, the patient usually develops chest symptoms within 1–2 years [3]. Radiologic diagnosis is difficult when LAM involves only the retro peritoneum or when it involves the retro peritoneum before it involves the lungs. Cystic lymphangiomas are similar to lymphangiomyomas in patients with LAM; however, the former consists of endothelium-lined lymphatic spaces, without the muscle or LAM cells in the walls that are diagnostic of lymphangiomyoma. When enlarged lymph nodes are seen in conjunction with lymphangiomyomas, a neoplastic process—such as lymphoma, metastasis, or primary cystic retroperitoneal tumour—may be misdiagnosed. Liposarcomas, leiomyosarcomas, and fibrosarcomas with cystic degeneration can be identified because they usually have solid components. [7] Old hematomas, abscesses, urinomas, and lymphoceles must also be considered in the differential diagnosis of a lymphangiomyoma. A fat-fluid level in a retroperitoneal cystic mass is a recognized feature of a mature teratoma [5].

We describe a patient with extra pulmonary lymphangiomyoma presenting as the retroperitoneal mass without lung involvement in postmenopausal female. She needs close follow up for pulmonary, renal, lymph node involvement.

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