

Case Report

Rare case of retroperitoneal cystic lymphangioma in an adult

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ABSTRACT

Cystic lymphangioma is benign tumor originating from the lymphatic vessels. Most commonly occurs in childhood and in the head or neck regions, retroperitoneal localization and presentations in adulthood are rare. Making a pre-operative diagnosis is challenging, and only possible subsequent to the histological examination. En bloc resection is the recommended treatment for cystic lymphangioma, and recurrence occurs due to an incomplete excision. We here present a case of 40 year female presenting with progressive palpable lump associated with pain in right quadrant of abdomen, which on computed tomography revealed a large retroperitoneal cystic lesion probably lymphangioma or cystic mesothelioma.

Keywords: Lymphangioma, Cyst, Retroperitoneal, Adult

INTRODUCTION

Retroperitoneal lymphangiomas occurs due to malformations of retroperitoneal lymphatic systems are extremely rare condition affecting adult patients.¹ Lymphangiomas being benign congenital malformation of lymphatic system are usually diagnosed in children and young adults and mostly located in head, neck and axilla where the loose connective tissue surrounding the structures allow lymphatic channels to expand and assume large proportions.²⁻⁴ Less than 5% of all lymphangiomas occur intra-abdominally and of them <1% occur in the retroperitoneum.⁵ When present intra-abdominally they generally present as diffuse lymphangiomatosis involving multiple organs such as liver, kidney, spleen, GI tract mesentery, mediastinum, bones and soft tissue.^{1,7} Hence solitary lymphangioma of retroperitoneum in an adult is a very rare tumor and very few cases have been documented. With this report we would like to share our experience in managing a female

adult with a giant cystic lymphangioma of the retroperitoneum complicated with kyphosis.

CASE REPORT

40 year old female, known case of kyphosis of dorso-lumbar spine presented with complain of progressively increasing lump in the right side of abdomen for the past one and half year associated with dull aching pain over right quadrant. The patient had history of caesarean 14 years back. On examination there was a palpable soft mass in the right hypochondrium reaching up to umbilicus and hypogastrium not moving with respiration. Infraumbilical midline scar of previous LSCS was noted.

Ultrasonography of abdomen revealed large multiloculated cystic lesion in the sub-hepatic region extending upto the pelvis. Contrast enhanced CT showed a large smooth thin walled multiloculated cystic lesion in sub-hepatic space extending upto right iliac fossa and hypogastrium region (Figures 1-3). It measured 19.5cm×12.4cm×12.8cm. There was multiple thin

enhancing septa. No evidence of solid components or calcifications were seen. Superiorly it indented the liver and pushed the bowel loops anteromedially.



Figure 1: Contrast-enhanced CT axial scan shows infrahepatic retroperitoneal lobulated-septated cystic mass.



Figure 2: Contrast-enhanced CT axial scan shows retroperitoneal lobulated large cystic lesion reaching upto right iliac fossa region.

We performed exploratory laparotomy and en bloc resection of retroperitoneal lymphangioma (Figure 4). There was a large multiloculated cystic mass extending from infrahepatic region to right iliac fossa. It was attached to right perirenal fascia, infrahepatic, subhepatic region and ascending colon. Around 700 ml of pale straw colored fluid was aspirated and en bloc resection was done.

Gross pathology revealed a well encapsulated mass measuring 19cm×11cm×17cm. Outer surface was lobulated and cut surface predominantly cystic (98%

multiple cysts ranging in size from 1-3cm). On microscopic examination multiple sections showed variably sized spaces lined by flattened cells enclosing pale eosinophilic lymph. The interstitial tissue showed paucicellular fibrocollagenous stroma with minimal lymphomononuclear inflammatory cell infiltrate and a few capillaries with occasional smooth muscle fibre.



Figure 3: Contrast-enhanced CT sagittal scan shows retroperitoneal lobulated-septated cystic mass extent from infrahepatic to hypogastric region.



Figure 4: En bloc resection specimen of cystic lymphangioma.

Following resection patient had an uneventful recovery. She was discharged on post op day 8 and is on regular follow up and currently is asymptomatic.

DISCUSSION

Lymphangiomas are generally considered to represent congenital malformation of lymphatic systems. Majority occurring relatively early in life. They develop as a result of lymphatic stasis due to congenital blockade or hypoplasia of regional lymphatic drainage.⁸ Isolated retroperitoneal lymphangiomas are extremely rare tumor,

more so when diagnosed in an adult patient. Despite controversy surrounding the hamartomatous, neoplastic or lymphangiectatic origin of this entity, general consensus points towards a benign etiology without any proven malignant potential.

Retroperitoneal lymphangiomas are mostly diagnosed incidentally or presenting with non specific symptoms such as vague abdominal pain or mass in affected site. The pain may be due to compression of surrounding structure. Sudden acute pain with peritoneal signs is not an unusual presentation and heralds cystic fluid leakage from the tumor.⁹

Typical USG or CT feature of lymphangioma is a cystic/multicystic intraabdominal mass with internal septations. However it may be difficult to come to an accurate preoperative diagnosis due to nonspecific nature of these findings. These findings may hold true for a myriad of other cystic conditions including but not limited to cyst adenocarcinoma, cystic teratoma, cystic mesothelioma, pseudomyxoma retroperitonii etc. although not done in our case MRI may offer better insight as this tumor is characterised by multilobulated heterogenous mass with low signal intensity in T1 and high signal intensity in T2 weighted images.¹⁰

Accurate diagnosis sadly can be made only by pathological examination. They are classified into three subtypes according to congenital dilated lymphatic channel: capillary (supermicrocystic), cavernous (microcystic) or cystic (macroscopic). Histologically they comprise lymphatic spaces lined by attenuated endothelium and filled with proteinaceous fluid (lymph) and may be unilocular or multilocular. Cystic lymphangiomas have the unique feature of having flat endothelial lining in place of cuboidal epithelium, lymphoid aggregates in the cyst wall and presence of smooth muscle fibres. Retroperitoneal lymphangiomas are usually of cavernous or cystic type.

Immunohistochemistry may show positive staining with CD31 or factor VIII related antigen but negative staining with cytokeratin. We did not perform IHC in our patient.

Preoperative diagnosis is difficult due to the rarity of disease and other more common cystic lesions must be kept in mind while arriving at a diagnosis. Diagnostic FNAC is not indicated due to risk of bleeding from wall and septa and the chance of secondary seeding in case of hydatidosis.

Surgical excision with en bloc resection is must to avoid recurrence. Other treatment modalities are not well

established and en bloc resection should be attempted wherever possible.

Owing to the high risk of recurrence patient must be followed up closely post-surgery. Clinical follow up and radiological investigations such as ultrasonography and CT scan are to be done at repeated intervals.

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