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Pancreatic lymphangioma

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CASE REPORT

Pancreatic Lymphangioma

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Abstract

Lymphangioma develops as a consequence of lymphatic malformation and blockage of lymphatic flow. Pancreatic lymphangioma is a rare benign tumour which can grow reasonably large prior to manifestation of symptoms. On imaging, it appears as a complex multiseptated cystic mass. However, this appearance is not diagnostic and overlaps with cystic pancreatic neoplasm. We present a case of pancreatic lymphangioma incidentally discovered in an elderly lady who was managed conservatively since surgery could not be performed as the patient was high risk for surgery. Imaging findings along with follow-up of the case and review of literature is presented.

Key Words: *Pancreatic lymphangioma. CT scan. Cystic tumours of the pancreas.*

INTRODUCTION

Lymphangiomas are frequently seen in children, mostly in the region of neck and axilla (95%).[1] Other sites of involvement reported in literature are lung, mediastinum, liver, spleen, colon, omentum, pancreas, genital organs etc. with about 1% occurring in mesentery or retroperitoneum and pancreas being an extremely rare site.[2]

On abdominal imaging it typically appears as a complex multiseptated cystic pancreatic mass. Less than 100 cases of pancreatic lymphangioma are reported in literature uptill now. We present a case of pancreatic lymphangioma incidentally discovered in an elderly lady. Imaging findings along with follow-up of the case and review of literature is presented.

CASE REPORT

An 84 years old frail looking female weighing 40 kg, presented with loss of appetite and weight loss. On examination, there was a right upper quadrant mass that was firm, mobile and non-tender on palpation. Ultrasonography was performed which showed a heterogeneous mass in the region of pancreas showing multiple cysts of varying sizes.

For further evaluation of this lesion, CT scan was carried out that showed a large 11.8 x 7.5 cm micro cystic mass with enhancing septae arising from head of pancreas and extending inferiorly (Figure 1). No soft tissue component or calcification was noted. The mass was closely abutting the duodenum, antrum of the stomach; segment-VI of liver, right kidney and gallbladder. It was causing scalloping of portal vein and prominence of pancreatic duct. There was no abdominal lymphadenopathy or mesenteric infiltration. The imaging diagnosis based on CT scan findings was micro-cystic mass of pancreatic origin. CEA and CA 19-9 were carried out and were within normal limits.

Core biopsy was carried out to establish the histopathological diagnosis which revealed multiple fragments of fibrocartilaginous tissue exhibiting cystic lesion composed of interconnecting channels lined by cuboidal to flattened epithelium with rounded nuclei and CD 31 positive. There was no evidence of malignancy. The final diagnosis was thus benign multicystic lesion with features favouring lymphangioma. Surgical excision was not done and the patient was managed conservatively due to the close proximity of the lesion to the adjacent structures and as the patient was old and frail and high risk for surgery and anaesthesia.

Nine months later, she again presented with abdominal discomfort and regurgitation, early satiety and retching and a repeat CT was performed to evaluate for any changes. However, on follow-up CT scan, the findings were essentially unchanged (Figure 1; right), therefore, her conservative management was continued till date.

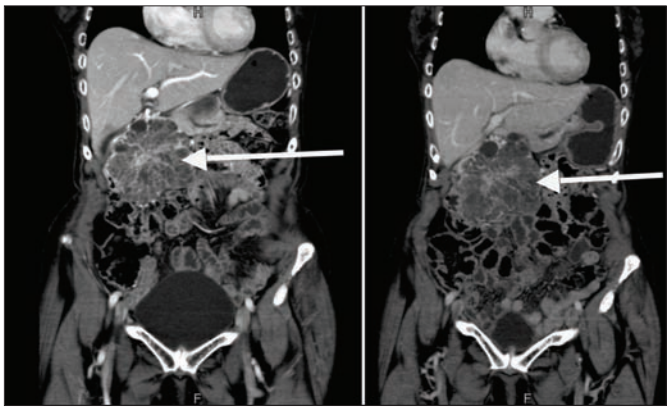

Figure 1: CT scan on left done in June 2009 showing multicystic mass in the region of pancreatic head; right: CT scan dated March 2010, not showing any change in size and appearance.

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DISCUSSION

Lymphangioma are benign slow growing tumours and the majority occurs in the neck, axilla and mediastinum in the paediatric population. The age ranged from 2 to 81 years with mean age of 28.9 years.[3] The age at presentation of our patient was 84 years which is more than the age reported in literature.

Pancreatic lymphangioma is a very rare accounting for less than 1%.[4] It originates from extra-lobular connective tissue, as a result congenital malformation of the lymphatics of the dorsal duodenum. The entity is common in girls and women with female: male ratio being 1.8:1. This disease entity lacks specific clinical symptoms and can grow reasonably large prior to manifestation of symptoms. The clinical presentation is inconsistent, unclear and depends on the location, size and mass effect of the lesion. Patients may be symptomatic presenting with abdominal pain, nausea, vomiting and palpable abdominal mass etc. or may be asymptomatic. The presently reported patient presented with non-specific symptoms and the tumour was detected incidentally as is also reported in literature. Complications like anaemia, infection, haemorrhage, torsion, volvulus, rupture, intestinal or ureteral obstruction presenting as acute abdomen have also been described.

There are no associated laboratory findings specific to this abnormality. Abdominal imaging like ultrasound and CT scan assist in the diagnosis and evaluation of such lesions, however, imaging by itself is not 100% specific to differentiate lymphangiomas from other cystic pancreatic lesions like pseudocyst, cystadenomas, congenital cysts and ductal carcinomas.[5] On ultrasound lymphangiomas usually appear as complex cystic masses. This is due to the presence of internal septa or internal echoes. Infrequently lymphangiomas exhibit calcification which is a characteristic of pancreatic cyst adenoma. Computed tomography provides greater details as compared to ultrasound in delineating pancreatic pathology even though differentiating lesions is still difficult. On CT scanning, pancreatic lymphangioma characteristically appear as a well circumscribed, thin walled, low density, homogenous cystic mass, which may be uni or multi locular with thin enhancing endocystic septae.[6] The appearance of mass on CT scan in this case was similar to the features described in literature. MRI can usually further confirm many of the findings on ultrasound and CT by showing a mass that is hypo-intense on T1 and hyper-intense on T2; in addition, MRI is better than CT in exclusion of communication between cyst and pancreatic duct.[7]

Differential diagnosis of cystic lymphangioma of pancreas from other cystic lesions is critical since it modifies the available therapeutic options. Pseudocyst on ultrasound appears as an anechoic usually unilocular cyst without septations and solid component but there may be an associated pancreatic duct dilatation.[8] On CT scan, it is characterized by lack of septa, loculations, solid component and cyst wall calcification. Serous cyst adenoma resembles cystic lymphangioma showing multiple cysts with very thin septae but central stellate scar and sunburst calcification favours cyst adenoma. Mucinous cyst adenoma, on the other hand, are generally poorly circumscribed large solid cum-cystic masses composed of varying sized cysts with septae, enhancing tumour growth with frequent peripherally located amorphous calcification. In this patient, the tumour was well circumscribed without calcification or enhancing solid component.

The final diagnosis is histopathological and supported immonohistochemically, with the endothelial cells showing reactivity to factor VIII-R antigen, CD-31 positivity and CD-34 negativity. These markers are sensitive, specific and reliable for the identification of lymphatic and capillary endothelium.[9] The therapeutic treatment option is complete surgical excision,[10] since incomplete excision may result in recurrence. From this case, and literature review, we can conclude lymphangioma of pancreas is a rare benign tumour and should be included in the differential diagnosis of cystic pancreatic neoplasm. Although complete excision of tumour is the treatment of choice, there are cases in literature that do not report any treatment similar to this case, when no significant progression of the disease process was noted on radiological follow-up.

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