

Case report

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Giant cystic Lymphangioma of Pancreas –A Rarity

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Abstract

Lymphangiomas are rare benign neoplasms common in head and neck region. Intraabdominal lymphangiomas are rare and pancreatic cystic lymphangiomas (PCLs) account for only 1% of such neoplasms. To the best of our knowledge only about 75 cases have been reported including this case and the average size was 20cms. Though benign, PCLs can cause severe abdominal discomfort and may even rupture. Here we report a case of PCL in a 21 year old male who presented with abdominal pain and epigastric mass. Clinicoradiological evaluation revealed a multicystic septate mass in association with the pancreas about 40x30cms. With the preoperative differential diagnosis of pseudocyst pancreas/cystic neoplasm we proceeded with exploratory laparotomy. A distal pancreatectomy and splenectomy was done since the lesion was arising from the body of the pancreas. Histological examination revealed a multicystic lesion lined by endothelial cells with lymphocytes in the wall and the diagnosis of PCL was made.

Key words Cystic, Distal pancreatectomy, Lymphangioma, Pancreas

Introduction

PCLs are rare neoplasms and usually present as a palpable mass or vague abdominal pain. They constitute only 0.2% of pancreatic neoplasms and show a female predilection (29:16). Our case was of a 21 year old male and the size was 40x30cm. Though lymphangiomas can attain any size review of literature shows that most were below 40cm.

Case report

A 21yr old male admitted with fullness of abdomen early satiety and decreased appetite of 2 weeks duration. He had jaundice 6 months back but no medical records are available. History of alcoholism was present but stopped 6

months back. There was no associated history of vomiting, loss of weight, TB or any other illness.

General examination was normal. Abdominal examination showed distended abdomen with flanks full and dilated veins over the anterior abdominal wall. Abdomen was non tender, tense with fluid thrill, and without any shifting dullness or any definitive mass or pulsations. Hernial orifices, external genitalia and supraclavicular areas were normal. Distal pulsations were normally palpable. Bowel sounds were normally present. All other systemic examinations were normal. All the routine investigations, liver function tests, renal function tests, random blood sugar, serum amylase and lipase were normal.

Radiological Evaluation

Chest X-ray was normal. USG abdomen revealed a large complex cystic lesion suggestive of pseudocyst of the pancreas, CECT abdomen

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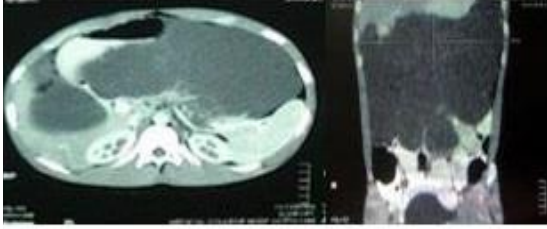


Figure 1: Suggests large intraperitoneal cystic lesion possibly Pseudo cyst pancreas

(Figure1) suggested a large intra peritoneal cystic lesion possibly a pancreatic pseudocyst.

After Clinico-radiological discussion, with the differential diagnosis of pseudocyst pancreas/cystic neoplasm we decided to do exploratory laparotomy for the patient.

Intra operative findings

The picture below shows the appearance of the lesion on midline laparotomy (Figure 2). Multiloculated cystic lesion of 40x30 cm size in the Supra colic Compartment (Figure 3), arising from the body of pancreas and encasing middle colic artery ,vein and Superior Mesenteric Vein.

Stomach was pushed Antero superiorly with a definite plane between stomach & the cyst. Rest of the viscera was normal. We had to decompress the larger cyst to do excision of cyst with distal pancreatectomy and splenectomy (Figure 4).



Figure 2: Shows the appearance of lesion on midline laparotomy



Figure 3: suggests the bulge in the transverse Meso colon which is pushed down due to the giant cystic mass

Pathological Findings

Gross examination revealed a partially collapsed cystic mass with pancreatic tissue and spleen. Cut section showed a multicystic septate mass with most of the locules filled with sero sanguinous fluid.

Microscopy revealed multiple cystic spaces lined by flattened endothelial cells, lumen showing eosinophilic secretions. Intervening stroma showed lymphocytic collections smooth muscles, fibrocollagenous tissue; cholesterol cleft formation and giant cell reaction. Pancreatic acini were seen in the periphery and also in between the cystic spaces (Figure 5). Immunohistochemistry revealed CD34 positivity for the lining cells (Figure 6).

Discussion

Lymphangiomas are benign tumors developing from congenital malformations of lymphatic vessels. The etiology in adults is thought to be lymphatic obstruction secondary to trauma or inflammation [1].

Though common in head and neck region, pancreas is a rare site. To term it as Pancreatic Cystic Lymphangioma (PCL), the neoplasm should arise within the pancreatic tissue substance or should be attached to the pancreas by a pedicle or should be adherent to it [2].



Figure 4: Picture showing the resected specimen

Histologically, lymphangiomas are of three types.- cystic, cavernous and capillary. Most reported cases are of cystic type. Any age group is affected but the mean age group is 40 years, with a wide range from 2-81 years [3]. Females are more affected than males (29:16) [3]. Macroscopically they are soft cystic masses filled with serous or serosanguinous or lymphatic fluid [4]. Rarely phlebolith like calcification may be present.

Histology shows interconnected vascular channels of varying sizes separated by thin septae. The vascular spaces are lined by endothelial cells positive for CD31, CD34 or D240 (specific marker) [5] and the walls contain variable amounts of collagen, connective tissue, irregular smooth muscle fascicles, occasional adipocytes and lymphocytic collections. The smooth muscles found in the wall of a cyst in combination with lymphocytes both in the wall and vascular lumina are most suggestive of

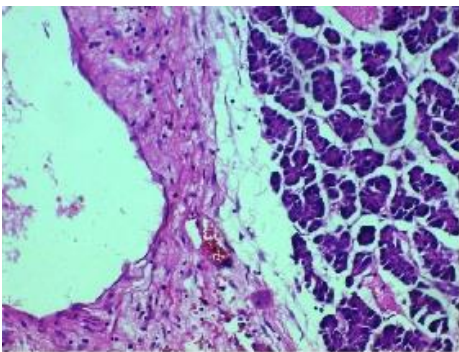


Figure 5: photomicrograph showing cystic spaces with endothelial lining and pancreatic acini in the periphery

diagnosis.

The common presentations are as palpable masses or abdominal pain. Smaller lesions are asymptomatic and are detected incidentally during imaging or surgery for unrelated diseases. Vomiting and nausea are infrequent symptoms and rarely PCLs can present as acute abdomen [6, 7, 8, 9]. Since first reported by Koch *et al.*, in 1913 [10] only less than 100 cases have been reported till date. Sonological studies can reveal a multi cystic septate mass. CECT abdomen can provide additional information about the capsule, septations of the cyst, its thickness and the characteristics of the fluid within [11]. However a definitive diagnosis cannot be made preoperatively by imaging alone. Some authors suggest the use of Endoscopic Ultrasound guided FNA study of cyst fluid for preoperative diagnosis [12, 13, 14]. But in our case it was not done because the clinical differentials considered were pseudocyst pancreas and other cystic neoplasms of pancreas like serous and mucinous cystadenomas. More over due to the huge size and symptoms an immediate surgery was planned. Huge PCLs can cause a significant discomfort with compression of adjacent organs. Ideally a complete surgical resection is mandatory to prevent recurrence (7%) [15]. In our case in order to obtain a complete resection, we did a distal pancreatectomy along with splenectomy. Follow-up USG abdomen and random blood sugar level after two years are within normal limits.

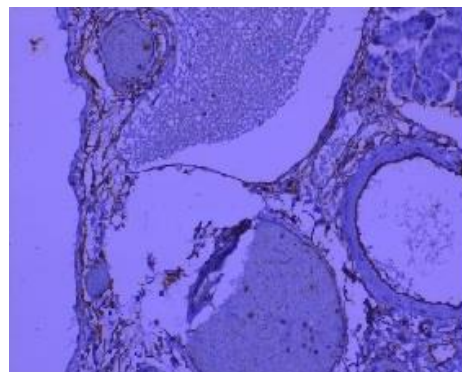


Figure 6: IHC showing CD34 positivity for the lining cells

Conclusion

PCLs are very rare benign cystic neoplasms of the abdomen. Although rare, this should be considered in the differential diagnosis of multicystic lesions of pancreas especially those presenting as abdominal masses. Surgical excision is the only known modality of treatment and is usually curative with excellent prognosis.

Authors' Contribution

CPJ: Conception, preparation of manuscript and final approval of the version to be published.

SEU: Conception and design of manuscript. Drafting the article and revising it critically for important intellectual content

LDP: Conception, design and preparation of manuscript

PU: Conception, design and preparation of manuscript

SV: Conception and design of manuscript

JSK: Design and preparation of manuscript

Conflict of Interests

The authors declare that there are no conflicts of interests.

Ethical Considerations

The written informed consent was obtained from the patient for publication of this case report.

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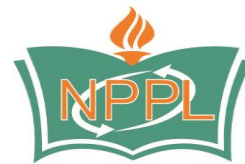
None

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