Giant Cystic Pancreatic Lymphangioma with Mediastinal Extension: A Case Report and Review of the Literature

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Abstract

Lymphangioma is an infrequent benign tumour that is formed usually from a congenital malformation of the lymphatic ducts causing lymphangiectasis. The abdominal location represents 1-5%, with dominance in the mesentery and retroperitoneum, however, the pancreatic location is very rare having described less than 100 cases published worldwide. The clinical symptoms of the lymphangiomas are non-specific and depend of the tumour's size and location. Here we present a clinical case of a pancreatic lymphangioma with mediastinal extension who was treated with a complete resection and this case shows that the diagnosis of cystic pancreatic lymphangioma must be taken as a differential diagnosis of pancreatic cystic lesions.

Keywords: Lymphangioma, Pancreas, Cyst

Abbreviations: CT: Computed Tomography; MRI: Magnetic Resonance Imaging; FNAB: Fine Needle Aspiration Biopsy; PET-CT: Positron Emission Tomography-Computed Tomography

Introduction

The lymphangiomas are benign cystic tumours which are usually the result of a congenital malformation of the lymphatic ducts causing local flow obstruction developing lymphangiectasia [1]. Histologically, these tumours have a protein and eosinophilic exudates within a few cysts separated by thin walls of endothelium [2].

The lymphangiomas are more frequent in children with varying symptoms according the tumour's location. Pancreatic presentation is very rare with about 70 cases published [1,3].

Case Report

A 56-year-old male was admitted to emergency service with fever over 12 hours and pain located in the epigastrium and left hypochondrium. The patient was not receiving any specific medication and his medical history did not suggest any major disease. Upon arrival, the patient presented good condition overall, stable hemodynamically with fever of 38.4°C. Physical examination revealed huge mass located in the epigastrium-left hypochondrium without tenderness.

Laboratory findings were leucocytosis of 16550 (93.3% neutrophils) and a C-reactive protein of 19.75 with rest of the parameters within normal limits. The CT scan
(Figure 1) reported a large multicystic mass adjacent to the body and tail of pancreas that extended up to left hypochondrium and epigastrium. The mass pushed the abdominal organs to the right side of the mediastinum, presenting another peripheral, pediculated and calcified appearance with a dense central region.

The study with MRI (Figure 2) described a multicystic mass of 29cm in its oblique diameter larger by 17cm in transverse diameter, with its epicenter in body-tail of pancreas and pancreatic atrophy signs were observed. Cysts extend superiorly to the right paraesophageal region and caudally to the left iliac fossa. Finally, it presented a porto-mesenteric axis increased (4 cm) diameter with a splenic vein elongated and many varicose veins. Diagnostic possibility was a mucinous tumour so it is continued with a PET-CT study observed a mild heterogeneous metabolic increase and the realization of a FNAB without malignancy.

A surgical procedure was contemplated by an approach via bilateral subcostal laparotomy for the realization of a total pancreato-duodenectomy with splenectomy getting a complete resection, in block, of the tumour. It was performed a complete liberation of the mass without affectation of the noble structures in the mediastinal extension.

The tumour measured 25 x 16 x 6 cm (Figure 3), it had a macronodular structure with a smooth surface and reddish colour, and the cysts contained a serous yellow-brown liquid. Microscopic description notes a coating of cysts with endothelial cells presenting immunohistochemical markers of CD31 and D2-40 positive and negative for CKAE1/AE3, CK7 and CD34. There were no malignant elements. The definitive histopathological diagnosis was pancreatic cystic lymphangioma.

The patient presented a satisfactory evolution and was discharged without complications after 15 days. Postoperative controls disease recurrence is not evident.

**Figure 1:** Abdominal CT scan shows a hypo-dense, capsulated and cystic lesion.

**Figure 2:** MRI images. It showed mediastinal extension in right paraesophageal region.

**Figure 3:** Pancreatic cystic lymphangioma.
Discussion

Cystic lymphangioma of the pancreas was originally described as a form of benign cystic lesion secondary to regional blockade of lymph ducts in 1913 by Koch [2].

Multiple factors are attributed to the genesis of a cystic lymphangioma: these include the presence of malformation of the lymphatic system, due to a proliferation of aberrant lymphatic tissue, without communication with the normal system, which is formed during prenatal development. In adults, other less common causes are the presence of blocking of the lymphatic drainage by abdominal trauma, surgery, infections etc. So the final result is that a secondary lymphangiectasis occurs [1]. Lymphosarcoma or adenocarcinoma malignant degeneration is extremely uncommon.

The cystic lymphangiomas can occur in any location where the normal lymphatic tissue is found. The most common locations are head and neck (75%). The intra-abdominal location is very uncommon (1-5%), being most common in the lactating males in the mesenteric region, the mesocolic, greater omentum and retroperitoneal location, appearing almost at a 90% before age two of life [3,4]. However, it has been described that the pancreatic location predominates in adults and women [3]. Inside the pancreas, the most frequent location is in pancreatic tail, followed by the head, the body and the location of the entire pancreas.

The clinical symptoms of the cystic lymphangiomas found are non-specific and variable depending on location and the size of the mass: may be asymptomatic or can be present chronic or acute abdominal pain, palpable mass or fever, mainly. Presenting as acute abdomen is observed in cyst's break, infections, bleeding or bowel obstruction. This presentation with acute abdominal pain due to breakage of one cyst is most frequent in children, chronic development is common in adults [2,3,5]. Specifically, the clinical symptoms of pancreatic lymphangiomas include abdominal pain and the presence of the mass that it can be confirmed by physical examination [6]; however, pancreatitis, weight loss, and laboratory abnormalities are not usual manifestations of pancreatic lymphangiomas [7].

The diagnosis can be: (1) an ultrasound of the abdomen where the pancreatic cysts will be observed. (2) CT scan and MRI show a well-circumscribed, encapsulated, water isodense, polycystic tumour, which help guide diagnosis, also determine the localization and preoperative size of the lesion, the relationship between this and adjacent structures and potential complications of surgery. (3) FNAB showing a serous exudate with a high content of eosinophils. However, these tests are nonspecific so it is currently very difficult to reach a preoperative diagnosis of pancreatic lymphangioma. Diagnostic confirmation of pancreatic cystic lymphangioma is in pathological analysis [1,8].

The differential diagnosis is between different possibilities such as pancreatic pseudocyst, mucinous cystadenoma, serous cystadenoma, cystic carcinoma of the Wirsung's duct and congenital neoplasms [9].

The surgical procedure should be a complete removal of the tumour and the affected organs that surround it because, despite being considered a benign entity, it has a high capacity of recurrence with increasing of the tumour, infiltration of neighbouring organs and increase of the risk of lymphatic spills [10]. Like incomplete excision being the only reason for recurrent diseases the
goal is obtaining a complete resection of the tumour with surgical margin resection free [5].

The pathological study, macroscopically it defines as a nodular lesion with reddish surface and a smooth membrane. Within the lesion, it presents a serous eosinophilic exudate. On microscopic examination, it would reveal liner flat endothelial cells in cysts along lymphatic ducts with smooth muscle in the wall of the wall and pancreatic tissue [11]. An immunohistochemical study can be realized: endothelial cells present positive staining response for markers factor VIII-R Ag, CD 31, D 2-40 and negativity for CD 34 which are markers for lymphatics ducts [1,2,12]. Since the lymphangiomas cells do not undergo differentiation, other types of stains such as mucicarmina, alcian blue or PAS staining are not useful, so the use of immunohistochemistry is only justified to reach an exact diagnosis [1].

**Conclusion**

We present a very rare case of giant cystic pancreatic tumour with mediastinum extension and corresponding to a cystic lymphangioma so this type of tumour should be taken into consideration as a differential diagnosis of pancreatic cystic lesions.

**Declarations**

The authors declare no conflict of interest. The case report was approved by ethics committee of our institution.

**Contribution to the work**

Miguel Torres did the writing this paper and reviewed related literatures.

Miguel Torres, Francisco Sánchez-Bueno and Gloria Torres performed the surgery.

Jesús de la Peña made the histologic study.

Matilde Fuster made the radiologic study.

Miguel Torres and Gloria Torres collected and took intraoperative and radiological images.

Pascual Parrilla was the superior director who was referred to when managing the clinical case.

**References**


