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Cystic lymphangioma of the pancreas

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Abstract

Lymphangiomas are benign cystic tumours of lymphatic origin. Most lymphangiomas occur in the neck and axillary region, and <1% occur in the mesentery or retroperitoneum. Lymphangiomas arising from the pancreas are extremely rare, with fewer than 70 published cases. Histologically, they are polycystic, with the cysts separated by thin septa and lined with endothelial cells. Though congenital, it can affect all age groups, and occurs more frequently in children and females. The authors report the case of a 53-year-old man, in whom a polycystic mass, 28 mm x 25 mm in size, was incidentally discovered by computed tomography in the tail of pancreas, during preoperative examination for a large asymptomatic epigastrocele. At surgery, a well circumscribed polycystic tumour was completely excised, with preservation of the pancreatic duct and the spleen. Histology confirmed a microcystic lymphangioma of the pancreas. Immunohistochemistry showed cystic endothelial cells reactivity to factor VIII-RA (++), CD31 (+++) and CD34 (±). The postoperative recovery was uneventful and the patient remained symptom free for two years. Although extremely rare, it is always a challenge to differentiate lymphangioma of the pancreas from other possible cystic – like neoplasms and should be taken into consideration.

Key words:
Cystic, Lymphangioma, Pancreas

INTRODUCTION

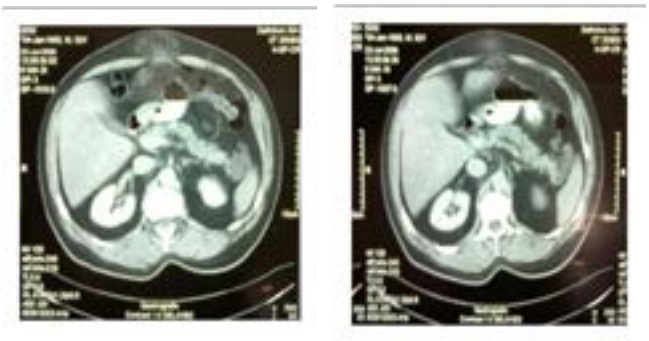
Lymphangiomas are rare benign cystic tumors that probably occur as a result of congenital malformations of the lymphatics leading to the obstruction of local lymph flow and the development of lymphangiectasia [1]. Gui et al described these sequestered lymphatic channels as a developmental abnormality rather than a true neoplasm [2].

Lymphangioma of the pancreas is extremely rare accounting for less than 1% of these tumors, and with fewer than 70 previously reported cases [3-7]. We present the case of an adult man with cystic lymphangioma of the pancreas and review the literature.

CASE REPORT

A 53-year-old man presented with a large asymptomatic epigastric hernia. He had no previous illness episodes. During preoperative examinations a computed tomography (CT) revealed a well-circumscribed polycystic lesion in the tail of the pancreas, about 3 cm in size, compressing the adjacent spleen, with thin septa within the lesion, and without dilatation of the pancreatic duct or wall calcifications (Fig. 1). Laboratory tests (carcinoembryonic antigen CEA, CA19-9, and serum amylase) were within normal limits and hydatid serology was negative. Preoperative diagnosis based on imaging investigations could not be made with certainty and a fine needle biopsy of the lesion was deemed of high-risk due to the location of the lesion and the possibility of malignant spread.

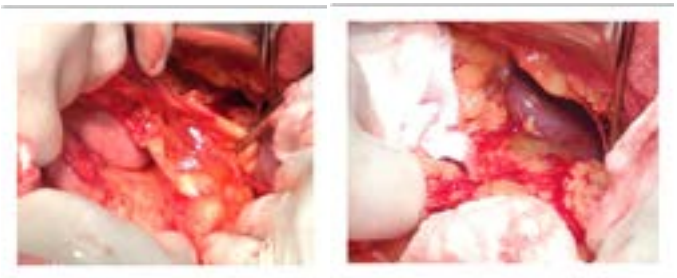
Figure 1. CT scan showing a well-circumscribed polycystic lesion in the tail of the pancreas, about 3 cm in size, compressing the adjacent spleen, with thin septa within the lesion.



A laparotomy was ultimately performed. At laparotomy the lesion was found in the tail of the pancreas, near the spleen, and did not involve the main pancreatic duct (Fig. 2). The lesion was excised intact and the main pancreatic duct and the spleen were preserved. No other pathology was found within

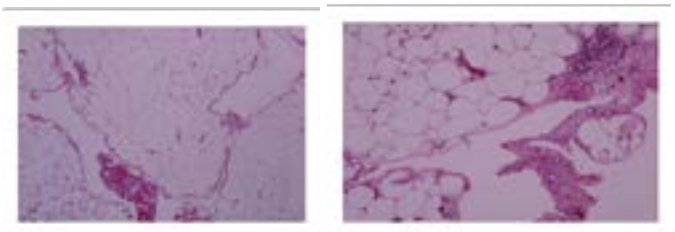
the abdomen, and the postoperative recovery was uncomplicated. The patient was asymptomatic 12 months postoperatively with no evidence of recurrence on subsequent abdominal imaging.

Figure 2. The polycystic tumour in the tail of the pancreas (A), near the spleen (B).



Histopathology revealed a polycystic structure, measuring 28 mm × 25 mm, lined by a flat endothelial epithelium, with small lymphatic spaces, abundant lymphoid tissue and smooth muscle present in the cystic wall. The cysts were separated by thin hypocellular septa (Fig. 3). Immunohistochemistry showed immunoreactivity to the factor VIII-R antigen (+++) and CD 31 positivity (+++), and CD 34 (±). Findings were consistent with a microcystic lymphangioma of the pancreas.

Figure 3. Pathology examination shows dilated lymphatics, lined by a flat endothelial epithelium, with small lymphatic spaces, abundant lymphoid tissue and smooth muscle present in the cystic wall [(HE stain, x190 (A) and x330 (B)].



DISCUSSION

Lymphangioma of the pancreas is rare, accounting for less than 1% of lymphangiomas [3,8]. It occurs more frequently in females and is often located in the distal pancreas [9,10]. The tumor size may vary between 3 and 20 cm in diameter (average 12 cm) [4,11]. The initial clinical symptoms are variable and may include abdominal pain, nausea, vomiting, and a palpable abdominal mass, although an acute abdomen has also been described [12]. In some cases, however, the cysts are asymptomatic and are discovered as an incidental finding, as in the present case. No specific or significant laboratory abnormalities have been reported. US typically shows a polycystic tumor usually with calcifications, which are typical for cystadenomas of the pancreas [13]. On CT, the tumor appears as a well-circumscribed, encapsulated, water-isodense, polycystic mass with thin septa, similar in appearance to cystadenomas, which occur far more frequently [4,14].

Differential diagnosis includes pancreatic pseudocysts, mucinous and serous cystadenomas, other congenital cysts and pancreatic ductal carcinoma with cystic degeneration [11]. The imaging characteristics of cystic lymphangioma may be useful in the differential diagnosis from other cystic tumors of the pancreas [4]. The final diagnosis is histological with the endothelial cells showing immunohistochemical reactivity to factor VIII/R antigen and CD 31 (+) positivity [5], as seen in our patient. Differential diagnosis using serum tumor markers may also be useful, because mucinous cystic neoplasms of the pancreas are immunoreactive for CEA and CA19-9 [8]. Most cases of non-functioning islet cell tumors with cystic degeneration have a thick wall and an irregular inner surface. Over 90% of cystic solid and papillary epithelial neoplasms are associated with dilatation of the pancreatic duct [2]. Patients with pseudocysts often have a history of acute or chronic pancreatitis [4].

A complete surgical excision is curative although not necessary because these tumors never cause problems as they do not have malignant behaviour and are not expected to increase in size. However, since preoperative diagnosis is not usually possible, most of the cases are directed to the operative theatre as possible cystadenomas and are excised as it happened in our case.

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