Large cystic lymphangioma of the pancreas: a case report

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ABSTRACT

Lymphangioma of the pancreas is rare and presents as a large cystic mass in the retroperitoneum. The pancreatic origin can be confirmed by careful evaluation of cross sectional imaging. Preoperative differentiation from other pancreatic cystic neoplasms is difficult but possible. Large symptomatic lesions warrant surgery. The diagnosis is confirmed by typical features on histopathology and immunohistochemistry. Presented here is a case report of a pancreatic lymphangioma, discussed in the context of available literature.

KEYWORDS

Pancreatic lymphangioma – Differential diagnosis – Computed tomography – Histopathology – Immunohistochemistry

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Introduction

Lymphangioma of the pancreas is extremely rare and presents as a cystic neoplasm. There are no typical features and the diagnosis is often made on histopathology. Symptomatic patients need surgery and complete excision is curative.

Case history

A 54-year-old woman presented with a history of noticing vague pain and fullness on the left side of the abdomen for two weeks. The history was otherwise unremarkable but for the fact that she had nine pet dogs at home. On examination, there was a large mass extending from the left costal margin up to the anterior superior iliac spine and crossing the mid-line. Ultrasound was suggestive of a cystic mass arising in proximity to the distal pancreas. There were no definitive features of a hydatid cyst. Hydatid serology (enzyme immunoassay of *Echinococcus* serum immunoglobulin G) was negative. Contrast-enhanced computed tomography showed a 21.6 × 19.4 × 12.8-cm multiseptated cystic mass arising from the tail of the pancreas (Fig 1) in close relation to the splenic hilum and extending up to the pelvic inlet. The mass was not infiltrating any other adjoining viscera.

After evaluation, the patient underwent distal pancreatectomy and splenectomy. Intraoperatively, a large multiloculated cystic mass was seen arising from the tail of pancreas, occupying the whole of the lesser sac and extending into the

infracolic compartment through the transverse mesocolon (Fig 2). The patient made an uneventful recovery and was discharged on postoperative day 5 and remained well on follow-up 6 months later.

The macroscopic cut section showed a multiloculated thin-walled cystic mass with foci of organised jelly-like material. Haematoxylin and eosin sections revealed a multilocular cystic lesion lined by flattened cells. Foci of lymphoid aggregates were noted in the mesenchymal stroma and in the cyst lumen, with proteinaceous fluid in some of them. Immunohistochemical staining for CD31 highlighted the flattened cells lining the cysts (Fig 3).

Discussion

Lymphangiomas are rare benign tumours, mostly encountered in the paediatric population in the neck and axillary regions. Cystic lymphangioma of the pancreas was first identified in 1913 by Koch as a form of benign cyst secondary to blocked regional lymphatic ducts.¹ It accounts for less than 1% of lymphangiomas and less than 70 cases were reported until 2008.² To date, around 100 cases have been reported in the English literature, the largest series being that of ten patients.³ It is reported more often in females and occurs commonly in the distal pancreas. Symptoms are usually attributed to the large size of the lesion.⁴ The average size is around 12 cm³ and the index lesion is among the few reported lesions larger than 20 cm.⁴ Lymphangiomas are

Figure 1 Coronal contrast computed tomography (CT) showing (A) a large cystic septated lesion closely abutting the inferior body and tail of the pancreas (bold arrow), with a small cyst within the distal pancreas (thin arrow) and (B) arterial branches of the splenic artery (thin arrows) supplying the septae of the lesion. (C) Sagittal contrast CT showing insinuation of the cystic components of the lesion into the inferior aspect of the pancreatic tail (bold arrow), indicating the lesion to be of pancreatic origin.

Figure 2 (A) Intraoperative photograph depicting a large cystic lesion arising in the lesser sac (arrowheads), seen bulging through the transverse mesocolon (lowest arrow head), extending into the infracolic compartment. The stomach (thin arrow) and transverse colon (bold arrow) are clearly seen. (B) Resected distal pancreatectomy plus splenectomy specimen demonstrating the large cystic tumour (arrow head) arising from the pancreas (bold arrow). Also seen is the spleen (thin arrow).

believed to arise from sequestrations of lymphatic tissue during embryological development. However, it has been suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery or radiation therapy may lead to the secondary formation of lymphangioma.⁵

It is difficult to diagnose a pancreatic lymphangioma on the basis of cross-sectional imaging; although septae are seen, there are no typical diagnostic features. Possible differential diagnoses include a simple cyst, macrocystic serous cystadenoma, mucinous cystic neoplasm, solid pseudopapillary neoplasm or a lymphovascular malformation. There were no clinical or radiological features of pancreatitis in the present case so pseudocyst was not considered. The history of pet dogs led us to also consider hydatid cyst as a

Figure 3 Characteristic histological features of a lymphangioma. (A) Haematoxylin and eosin (H&E) preparation at 4x magnification showing pancreatic parenchyma with cystic spaces (arrow) filled with proteinaceous fluid. (B) H&E preparation at 4x magnification showing interspersed lymphoid clusters (arrow) within the cystic mass. (C) H&E preparation at 10x magnification showing lymphoid cells within the proteinaceous fluid of cyst (arrow). (D) Immunostaining at 10x magnification showing CD31 positivity in flattened cells lining the cystic spaces (arrow).

possibility but there were no daughter cysts or scolices seen on ultrasound. In many instances, owing to the large size of the cyst, it may not be possible to discern the site of origin of the cyst preoperatively. There is a report where a large cyst was thought to be ovarian in origin and the intraoperative findings proved otherwise.² In this case, the cyst was deriving blood supply from the splenic branches near the tail of pancreas and we were sure of the pancreatic origin. In small asymptomatic cysts finding chylous fluid with elevated triglyceride levels, lymphocytes without atypia or epithelial cells on endoscopic ultrasound may suggest a lymphangioma, thus avoiding surgery.⁶ Endoscopic ultrasound was not performed in this patient as it would not have provided any extra information that would alter the decision to proceed with surgery. The type of surgery depends upon location of tumour. In this instance, involvement of the splenic hilum precluded spleen preservation. Sometimes adjacent organ resection (stomach, colon) may be required. Laparoscopic resection is emerging as a gold standard for distal pancreatectomy. Since the index lesion was large and preoperative diagnosis was not certain, we adopted an open approach to prevent spillage of cyst contents.

In most instances, diagnosis is confirmed only at pathology. Macroscopically, cystic lymphangiomas appear as soft, multiloculated masses that contain serous, serosanguinous or lymphatic fluid. Histologically, dilated lymphatic channels of varying sizes are seen, separated by thin septae which comprise occasional smooth muscle fascicles and collagenous connective tissue. The cystic spaces are lined with flattened or cuboidal endothelial cells. Aggregates of

lymphocytes are found in the lumina and adjacent stroma.⁵ Immunohistochemistry shows positivity for endothelial markers such as factor VIII-R antigen, CD31, CD34, and D2-40 in some instances.⁵ CD 31 was positive in the index case and the histopathology was quite confirmatory for lymphangioma.

Pancreatic lymphangiomas have good prognosis when completely resected. The lesion will invariably recur if treated by aspiration or partial resection. Dense inflammatory reaction surrounding the lymphangioma can sometimes make dissection planes difficult. In this patient, it was a challenge to separate the wall of the tumour from the transverse mesocolon (Fig 2). Every effort should be made to excise the tumour completely without spillage and not leave any portion of the wall of the tumour behind. With R0 resection, long-term survival is the rule. There is no role for adjuvant therapy.

Although rare, pancreatic lymphangioma should be considered in the differential diagnosis of cystic neoplasms of the pancreas.

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