

## CASE NOTE

# Huge cystic lymphangioma of the pancreas

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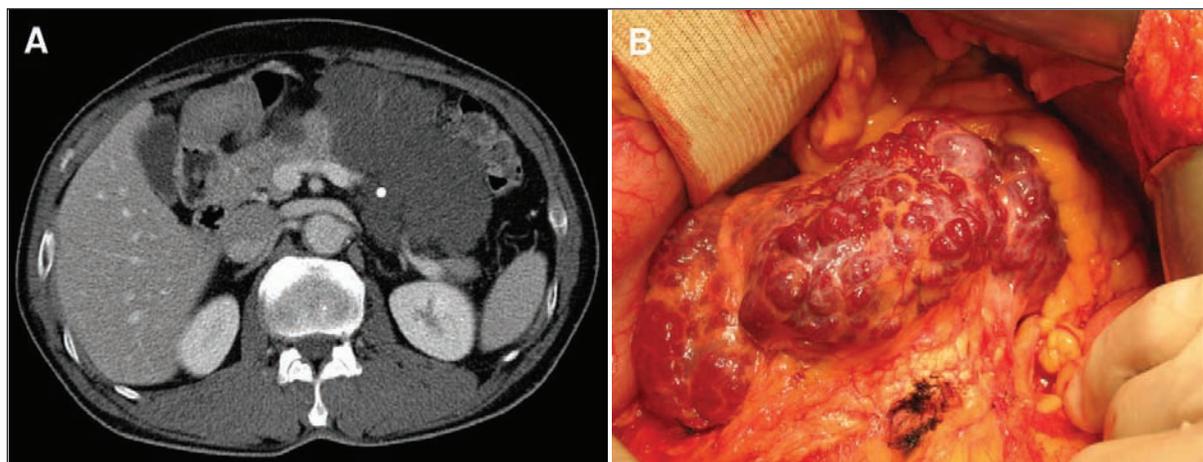
**C**ystic lesions in the pancreas include pseudocysts, simple cysts, serous cystadenomas, mucinous cystic neoplasms and intraductal papillary mucinous neoplasms. Another rare cystic lesion is pancreatic lymphangioma. Lymphangiomas are rare benign tumours that are most often seen in the neck and axilla (95%). The abdominal cavity is a rare site of origin, and most reported cases developed in the retroperitoneum or mesentery.<sup>1</sup> We describe the case of a patient with a pancreatic cystic lymphangioma that mimicked a cystic pancreatic tumour.

**CASE REPORT**

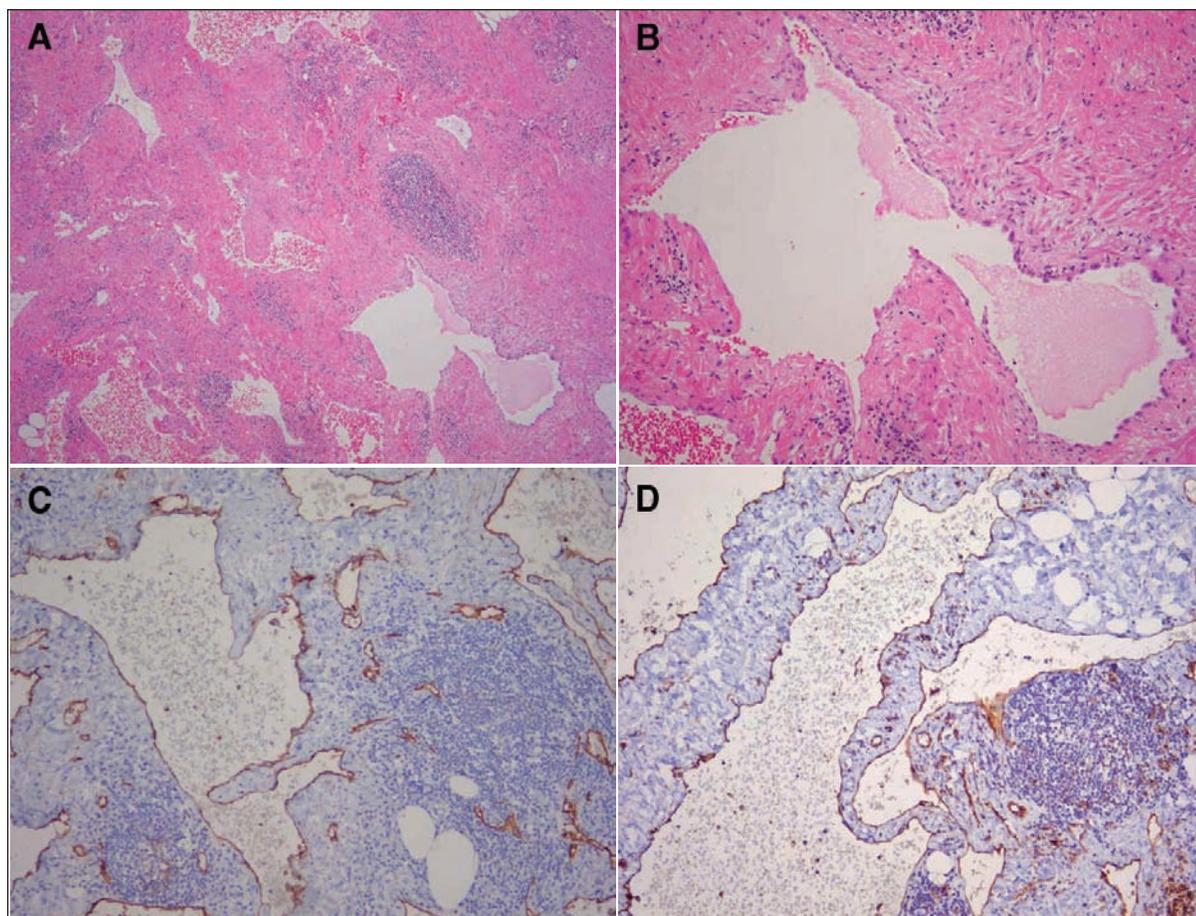
A 66-year-old man presenting with a palpable mass in the left upper abdomen was referred to our hospital. His medical and family history were unremarkable. He had no history of previous abdominal surgery.

On physical examination, we palpated a 15-cm hard mass in the left upper quadrant. A computed tomography (CT) scan of his abdomen showed a large cystic lesion with multiple septation, measuring about 13.0 × 8.5 × 9.0 cm. The mass appeared to completely encase the body and tail of the pancreas (Fig. 1). From these radiographic findings, we diagnosed a cystic neoplasm of the pancreas.

Serologic screening for the tumour markers AFP, CA 19-9 and CEA revealed normal serum levels. At laparotomy, we identified a large multiloculated cystic mass originating from the body and tail of the pancreas (Fig. 1). The patient underwent distal pancreatectomy and splenectomy. Histological examination revealed that the tumour contained numerous dilated lymphatic spaces of varying sizes separated by collagenous septa (Fig. 2). The endothelial cells lining the surface of the cystic spaces were positive for CD31 and CD34



**Fig. 1.** (A) Abdominal computed tomography scan showing a large septated cystic tumour, 13.0 × 8.5 × 9.0 cm in size, with water-dense contents to the pancreatic body and tail. (B) At laparotomy a multiloculated cystic tumour was located in the body and tail of the pancreas.



**Fig. 2.** Hematoxylin and eosin staining showed (A) variably sized vascular endothelium-lined spaces with cyst walls containing various amounts of collagenous connective tissue, irregularly smooth muscle fascicles and mature lymphocytes (original magnification  $\times 40$ ). (B) The cystic spaces are lined by flattened endothelial cells (original magnification  $\times 100$ ). Immunohistochemistry showed that the tumour cells are (C) immunoreactive for CD31 and (D) CD34 (original magnification  $\times 100$ ).

(Fig. 2). The final pathologic diagnosis was cystic lymphangioma of the pancreas. The patient had an uneventful postoperative course, and there was no evidence of recurrence in the 20 months after his surgery.

## DISCUSSION

Pancreatic lymphangioma is an extremely rare tumour, described for the first time by Koch in 1913.<sup>2</sup> It is more commonly found in women and more commonly located in the body and tail of the pancreas.<sup>3</sup>

The etiology of lymphangiomas remains unclear; a well-established theory suggests that lymphangiomas arise from sequestrations of lymphatic tissue during embryologic development. However, it has been suggested that abdominal trauma, lymphatic obstruction, inflammatory process, surgery or radiation therapy may lead to the secondary formation of such a tumour.<sup>1</sup> Histologically, lymphangiomas are classified into 3 types: simple capillary, cavernous and cystic lymphangiomas.<sup>1,2</sup>

The formation of a cystic lymphangioma of the pancreas may be due to the abnormal development of the

lymphatic system in the dorsal mesoduodenum. This is supported by the fact that cystic lymphangiomas of the pancreas appear to originate from the extralobular connective tissue of the pancreas.<sup>3</sup>

The clinical picture of pancreatic lymphangiomas is usually symptomatic (92.2%). Abdominal pain and palpable mass seem to be the most common symptoms.<sup>2</sup>

Imaging investigations can suggest the diagnosis of cystic lesion and its pancreatic origin: an ultrasound will show a complex cystic mass owing to internal septa and a CT scan will show a hypodense mass with a thin wall and septa. Computed tomography is considered superior to ultrasonography in delineating pancreatic pathology even though differentiating lesions is still difficult.<sup>4</sup>

In our patient's case, the diagnosis of a pancreatic lymphangioma was supported immunohistochemically by positive staining of factor VIII-R antigen, CD31 and CD34, which are sensitive and specific markers for the identification of lymphatic and capillary endothelium.<sup>3</sup>

The optimal treatment is radical excision, since incomplete resection may lead to recurrence.<sup>3</sup> Although lymphangioma is a benign lesion, it often behaves in an

aggressively invasive manner and can grow to an enormous size. Therefore, resection of adjacent organs may be required to accomplish complete excision.

In pancreatic cystic lesions, the correct diagnosis can be established only via laparotomy and subsequent pathologic interpretation. Although intra-abdominal lymphangiomas are rare lesions, pancreatic lymphangioma should be included in the differential diagnosis of pancreatic cystic neoplasms.

**Competing interests:** None declared.

## References

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