CASE REPORT

Epidermoid cyst within an intrapancreatic accessory spleen

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SUMMARY

This is a case of an epidermoid cyst in an intrapancreatic accessory spleen at the tail of the pancreas. Concurrent epidermoid cyst within the accessory spleen is an exceedingly rare entity. The patient initially presented with abdominal pain, nausea and vomiting. Imaging studies revealed a 3.6 cm cystic mass in the tail of the pancreas that was concerning for a mucinous cystic neoplasm (MCN). The patient underwent a robotic distal pancreatectomy for a suspected diagnosis of MCN, which following histologically examination was found to be an epidermoid cyst of an intrapancreatic accessory spleen. Further imaging characteristics are needed to characterise and differentiate these lesions from those of malignant potential. Robotic distal pancreatectomy is a safe alternative with many benefits including decreased length of stay and decreased blood loss.

BACKGROUND

Accessory spleens are morphologically and functionally similar to normal spleens and are incidentally found in 10%–30% of the population.¹ Approximately 60% are found at the splenic hilum and 33% at the lower pole of the spleen.² Other locations of accessory spleens, although more rare, include the fundus of the stomach,³ pancreatic tail,⁴ greater omentum,⁵ retroperitoneum^{6,7} and pelvis.⁸

Since first described in the literature in 1980 by Davidson *et al*,⁹ epidermoid splenic cysts remain extremely rare. On review of 42 327 autopsy records, Robbins *et al* found a splenic epidermoid cyst in 0.07% of the population.¹⁰ The presence of an epidermoid cyst in an intrapancreatic accessory spleen is an extraordinarily rare finding. Differentiating these preoperatively from concerning malignant lesions such as a neuroendocrine tumour, mucinous cystic neoplasm (MCN), intraductal papillary mucinous neoplasms (IPMN) or solid pseudopapillary tumour of the pancreas remains difficult.

The similar density of the solid component of the cystic lesion to the spleen on enhanced CT and MRI has been postulated to assist in correct identification of an epidermoid cystic lesion within an intrapancreatic accessory spleen. However, only a few case studies have reported if this finding was present.^{11–13} Bastidas *et al*¹⁴ describe successful identification of intrapancreatic ectopic spleen with use of an endoscopic ultrasound-guided needle-based probe confocal laser endomicroscopy which permits high-resolution visualisation at microscopic

levels through the Fine Needle Aspiration (FNA) needle.¹⁵

Identification of a distal pancreatic tail lesion concerning for a premalignant cyst that is greater than 3 cm is often treated with surgical resection.

This is a case of a pancreatic tail lesion concerning for MCN based on available imaging, which after final histopathological evaluation was found to be an epidermoid cyst in an intrapancreatic accessory spleen.

CASE PRESENTATION

A 17-year-old Caucasian woman with medical history significant for obesity, depression and supraventricular tachycardia presented with 5 months of persistent nausea and intermittent vomiting, not associated with meals. She endorsed a 10 lb unintentional weight loss during this period. She denied abdominal pain, melena, constipation or diarrhoea. She also denied a history of pancreatitis, diabetes or trauma. There was no significant family history of pancreatitis or malignancy.

INVESTIGATIONS

Serological tests were performed and a complete blood count, sodium, potassium, chloride, bicarbonate, blood urea nitrogen, creatinine, glucose and hepatic function panel were all within normal limits. Cancer antigen 19-9 was <15. Physical examination revealed no abnormality. CT of the abdomen and pelvis revealed a 3.6×1.9×2.8 cm cystic lesion arising in the tail of the pancreas (figure 1).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of a cystic pancreatic tail lesion includes MCN, IPMN, solid pseudopapillary tumour of the pancreas and cystic pancreatic neuroendocrine tumour.

TREATMENT

Due to concern for a premalignant cystic lesion of the pancreas greater than 3 cm, surgical intervention was recommended and discussed with the patient and guardian. They elected to proceed with robotic distal pancreatectomy with splenectomy. Intraoperatively, there was no evidence of metastatic disease. Ultrasound was used to ensure presence of cystic lesion in the tail of the pancreas. The surgical resection specimen was sent for pathologic analysis.

OUTCOME AND FOLLOW-UP

The patient's postoperative course was uncomplicated and she was discharged home on postoperative

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Figure 1 CT of the abdomen and pelvis revealed a 3.6×1.9×2.8 cm cystic lesion arising in the tail of the pancreas.

day 3. Histological examination revealed a benign epithelial cyst lined by squamous epithelium arising in an accessory spleen, which was found in the soft tissue between the pancreas and the spleen. The accessory spleen measured 4.3×3.9×0.9 cm, and weighed 12.7 g.

DISCUSSION

The presence of an intrapancreatic epidermoid cyst within an accessory spleen is a rare finding. van Dijk *et al*¹⁶ reviewed 57 published cases of epidermoid cysts in intrapancreatic accessory spleens since they were initially described in 1980. In all cases,

like ours, the lesion was found in the pancreatic tail. Unlike our current case, the majority were middle-aged women of Asian descent. All patients reviewed underwent either an open or distal pancreatectomy with or without splenectomy.

In conclusion, an epidermoid cyst of an accessory intrapancreatic spleen is a rare finding that is hard to identify on diagnostic imaging and often misinterpreted as a premalignant or malignant lesion. Though our current patient had symptoms that seem to have improved since surgery, further investigation into how to improve preoperative diagnosis is warranted.

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Learning points

- ▶ One in six accessory spleens occur in the tail of the pancreas.
- ▶ Accessory spleen should be included as a differential diagnosis of pancreatic tail lesions.
- ▶ Splenic epidermoid cysts are true cysts lined by squamous epithelium, and though rare can be found within accessory spleens.
- ▶ Endoscopic ultrasound-guided needle-based probe confocal laser endomicroscopy may provide an advantage versus endoscopic ultrasound-guided fine-needle aspiration in obtaining an accurate preoperative diagnosis of pancreatic lesions.
- ▶ Treatment of pancreatic lesions with robotic assistance should be strongly considered due to the benefits to the operating surgeon and additionally decreased postoperative pain, length of stay and associated mortality and morbidity.

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