Introduction

Approximately 10% of the population will have an accessory spleen in various parts of the body\(^1\). When an accessory spleen arises from the pancreatic tail, it occasionally mimics pancreatic solid tumors\(^2,4\), resulting in unnecessary surgical resection. However, because of recent developments in radiological diagnostic modalities and in our understanding of this condition, careful follow-up is now starting to be chosen first, rather than surgery.

However, when the accessory spleen is accompanied by cystic changes, diagnosing the lesion becomes quite challenging. One of the differential diagnoses of cystic lesions is epithelial (epidermoid) cysts formed in the intrapancreatic heterotopic spleen\(^5,10\). A literature review of previous cases suggests that radiological findings vary among cases and that the condition often mimics mucinous cystic neoplasm (MCN), in that both have thick walls, a solid component (similar to mural nodules), and no communication with the pancreatic duct, and patients have elevated serum CA19-9 levels\(^5\). Unfortunately, patients are often treated with surgery, despite the fact that specific treatment is not required for asymptomatic epithelial cysts\(^5,10\).

Another differential diagnosis of cystic lesions in the pancreatic tail could be peliosis of the intrapancreatic heterotopic spleen\(^11,12\). Peliosis is also quite a rare condition and is characterized by multiple cyst-like blood-filled cavities. Peliosis occurs most commonly in the liver and rarely in the spleen. The pathogenesis, natural history, and treatment of splenic peliosis remain unclear, and the accumulation of information from more cases is necessary for a better understanding of this rare condition.

Case presentation

A 44-year-old Japanese man with a medical history of chronic hepatitis C virus infection was referred for the treatment of a pancreatic cystic lesion. When the patient was 42 years of age, abdominal ultrasonography (US) first identified a 1.8-cm cystic mass lesion arising from the pancreatic tail. Although the lesion had been followed-up as a benign pancreatic cyst, the most recent abdominal US indicated that the mass lesion had increased...
over a period of 10 months to a maximum diameter of 3.2 cm.

The patient did not have a history of pancreatitis, abdominal trauma, alcohol abuse, or other drug abuse. The patient was asymptomatic and physical examination was unremarkable. Laboratory studies revealed hepatobiliary, pancreatic enzyme, and tumor markers all within the normal range. Contrast-enhanced abdominal computed tomography (CT) revealed a 3.5-cm oval cystic lesion with a clear, thick, and smooth wall in the pancreatic tail. The cyst wall was mildly enhanced, whereas the inner part of the cyst was not (Fig. 1). Magnetic resonance imaging revealed high signal intensity for the inner contents of the cyst on T1-weighted images and low signal intensity on T2-weighted images, suggesting a mucinous element (Fig. 2A, B). T1-weighted imaging clearly demonstrated the septum of the cyst, which was not clear on CT scans. Although preoperative endoscopic ultrasound revealed the septum of the cyst, the result was not diagnostic. There was no communication between the main pancreatic duct and the cyst identified on endoscopic retrograde cholangiopancreatography, and the lesion was diagnosed as MCN of the pancreatic tail. The patient underwent distal pancreatectomy because of the malignant potential of the lesion (based on the increase in size of the cystic tumor and the nodule within the cyst). Fine needle aspiration biopsy was not performed because of the potential risk of implanting malignant cells along the biopsy needle.

On laparotomy, intraoperative US showed a well-demarcated round mass in the pancreatic tail with septa (Fig. 3). On the resected specimen, the cystic lesion was dark red in color and filled with fluid (Fig. 4A). The cyst was multilobular with a red colored inner surface and filled with old blood (Fig. 4B). Pathologically, the cyst was located in the intrapancreatic accessory spleen. There were also other cystic lesions filled with old blood but without an apparent epithelial lining (Fig. 5).
Fig. 3  Intraoperative ultrasound
The contents of some cavities were highly echogenic (white arrowhead), whereas others had low echogenicity. The septa of the cyst are indicated by the white arrow.

Fig. 4  Resected specimen of the pancreatic cystic lesion
(A) The external aspect of the lesion is dark red (black arrow).
(B) The cross-section of the specimen reveals that some of the cysts are filled with old blood. The cyst is surrounded by pancreatic tissue with fibrous changes (black arrow).

Fig. 5  Pathological examination
(A) The whole structure of the lesion. There are several cavities filled with blood that are not lined by epithelial cells, which is diagnosed as peliosis (P). Other cystic lesions are lined with epithelial cells (small arrowhead) and diagnosed as epithelial cysts. The whole cystic lesion is surrounded by pancreatic tissue with fibrous changes and adjacent to normal pancreatic tissue.
(B) The epithelial cyst is surrounded by columnar epithelium. The thin layer of ectopic tissue is comprised of lymphoid tissue with a germinal center (arrow), sinusoid, and hyalinized fibrous tissue (arrowhead).
(C) The markedly dilated blood-filled spaces (peliosis) lack an endothelial lining.
final pathological diagnosis was peliosis and epithelial cyst of the intrapancreatic accessory spleen without malignant change.

Discussion

Approximately 10% of the population will have an accessory spleen in various parts of the body, including the wall of the jejunum, mesentery, pelvis, and intraparenchymal tissues. However, the formation of epithelial cysts in an accessory spleen is rare and difficult to diagnose preoperatively. Occasionally, malignancy may be suspected because of the solid component of the cyst and elevated CA19-9 levels, even though malignant changes of epithelial cysts have not been reported. It may be possible to correctly diagnose these cysts preoperatively if the solid part of the lesion is of the same intensity as the splenic tissue on CT or MRI. However, when the ectopic splenic tissue is very thin, as in the present case, it resembles the thick wall of neoplastic cystic lesions, making preoperative diagnosis difficult.

In the current case, the cystic lesion was preoperatively diagnosed as MCNs because it had a characteristic feature of MCNs, such as a thick-walled single cyst occurring in the neck to tail of the pancreas, and formation of septa. However, MCNs are predominantly identified in female patients, leaving the other differential diagnosis to be considered. Intraductal papillary mucinous neoplasm (IPMN) of pancreas could be one of them. However, the radiological evaluation of present case did not show dilated main pancreatic duct, bulging of papilla of Vater, communication of pancreatic duct and cysts, multilocular or multifocal cysts. Thus, the cystic lesion in the present case was tentatively diagnosed as MCNs of pancreas harboring possibility of malignant change rather than IPMNs or other epithelial and non-epithelial benign pancreatic cysts.

On pathological examination of the present case, some cystic lesions were identified as peliosis, which is characterized by multiple cyst-like blood-filled cavities within the parenchyma of solid organs. The isolated occurrence of splenic peliosis is quite rare and poorly understood. We consider that peliosis developing in the intrapancreatic accessory spleen is even rarer, and quite difficult to diagnose preoperatively. Peliosis is often described in association with chronic hepatitis C virus, tuberculosis, intravenous drug abuse, chronic alcoholism, hematological malignancies, chemotherapy, or steroid treatment. Although these conditions have been proposed as potential risk factors for peliosis, no solid conclusions have been made as yet. The natural history of peliosis is unclear, and the role of surgery in its treatment has not been determined. However, all patients diagnosed as having peliosis should be provided with information and managed carefully because the spontaneous rupture of peliosis, a potentially life-threatening event, has been reported.

In conclusion, we report on our experience of a patient with rare coexisting peliosis and epithelial cyst in the intrapancreatic accessory spleen. The rapid growth of the pancreatic cystic lesion, possibly due to the peliosis, mistakenly indicated malignant potential, resulting in surgical treatment. A greater understanding of these rare diseases and the development of accurate diagnostic techniques are necessary to reduce unnecessary surgery in patients with pancreatic cystic lesions.

References