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A Case of Epidermoid Cyst of the Intrapancreatic Accessory Spleen

A 54-year-old woman presented with a huge palpable mass on left upper quadrant of the abdomen. After preoperative work-up, a cystic disease of pancreatic tail or accessory spleen was initially suspected. We performed exploratory laparotomy and resected both the spleen and a 15×11 cm-sized huge cystic mass containing a part of solid component which extended continuously to the pancreatic tail. The solid component, comprising the upper portion of the resected cyst, was reddish brown and granular like as normal splenic tissue. The inner surface of the cyst was smooth and was filled with yellowish white material. Histologic examination showed an epidermoid cyst originating in the accessory spleen of the pancreatic tail lacking hair or skin appendages.

Key Words: *Epidermal Cyst; Splenic Neoplasms; Pancreas*

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INTRODUCTION

About 10% of the population is reported to have accessory spleens, which are usually located at or near the splenic hilum. However 16% of the accessory spleen is found in the pancreatic tail (1, 2). The epidermoid cyst formed in the accessory spleen within the pancreatic tail is an extremely rare disease. It has been reported only about ten times worldwide since initial report by Davidson et al. (3) in 1980. It could be misdiagnosed as cystic tumor or pseudocyst of pancreatic tail, thus requiring differential diagnosis. We here report a case of epidermoid cyst of accessory spleen within the pancreatic tail.

CASE REPORT

A 54-year-old female patient was admitted with complaints of epigastric discomfort, fullness, nausea, vomiting and weight loss for one month. On physical examination, a 8.0×9.0 cm-sized, palpable mass with tenderness was noted in the left upper quadrant of the abdomen.

Abdominal ultrasonography found a huge cystic mass located at the pancreatic tail. On abdominal computed tomography, a 15.0 cm-sized, huge cystic mass connected to the pancreatic tail was compressing the pancreas (Fig. 1A). The mass was mostly composed of a cystic component, and a small solid component in the left lateral and upper part. The solid component of the mass, forming a wall in the left lateral and upper portion of the cyst, showed the same homogenous attenuation as in the spleen (Fig. 1B). On the magnetic resonance imaging, the cystic component showed low-signal intensity on T1-weighted images, and high on T2-weighed images. The solid component showed intermediate low-signal intensity on T1-weighted images and intermediate high on T2-weighted images. The solid component showed the same signal change as the normal splenic tissue on contrast study (Fig. 2).

Surgical exploration was performed under the assumption that it was either a cystic disease of the pancreas or accessory spleen. According to operative findings, a 15.0×11.0 cm-sized, huge cystic mass connected to the pancreatic tail, containing a 2.7×2.2 cm-sized, brownish fibrous tissue at the left upper part, was found. The spleen had normal location and appearance, and measured 8.5×5.2×2.6 cm. There was no connection between the spleen and the cystic mass. The frozen section of the fibrous tissue of the cyst revealed to be of the same nature as normal splenic tissue. The cystic mass with surrounding pancreatic tissues was resected by distal pancreatectomy, along with splenectomy.

On pathologic examination, the huge cystic mass in the pancreatic tail measured 15.0×11.0×3.5 cm and weighed 192 g. The cyst was a single cyst without any loculation. A portion of the outer surface of the cyst was covered with the accessory spleen in the pancreatic tail.

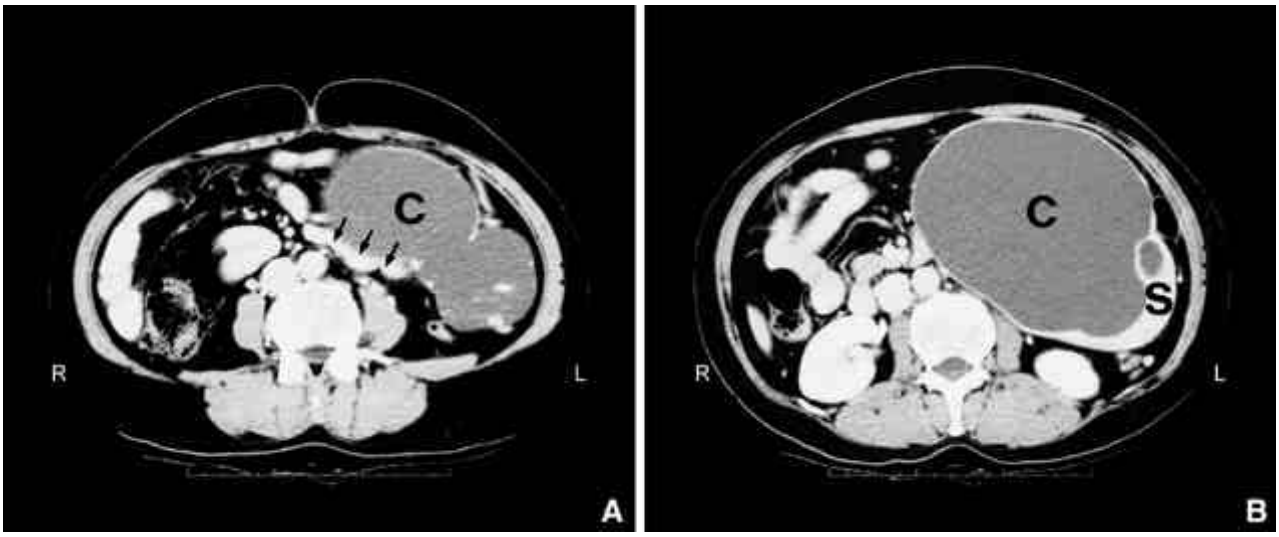

Fig. 1. Abdominal computed tomography. A huge cystic mass is connected to the pancreatic tail, compressing the pancreas (arrows) (A). Solid component, which was later revealed to be an accessory spleen, shows the same homogenous attenuation as the spleen (B). C, cystic component of the mass; S, solid component of the mass.

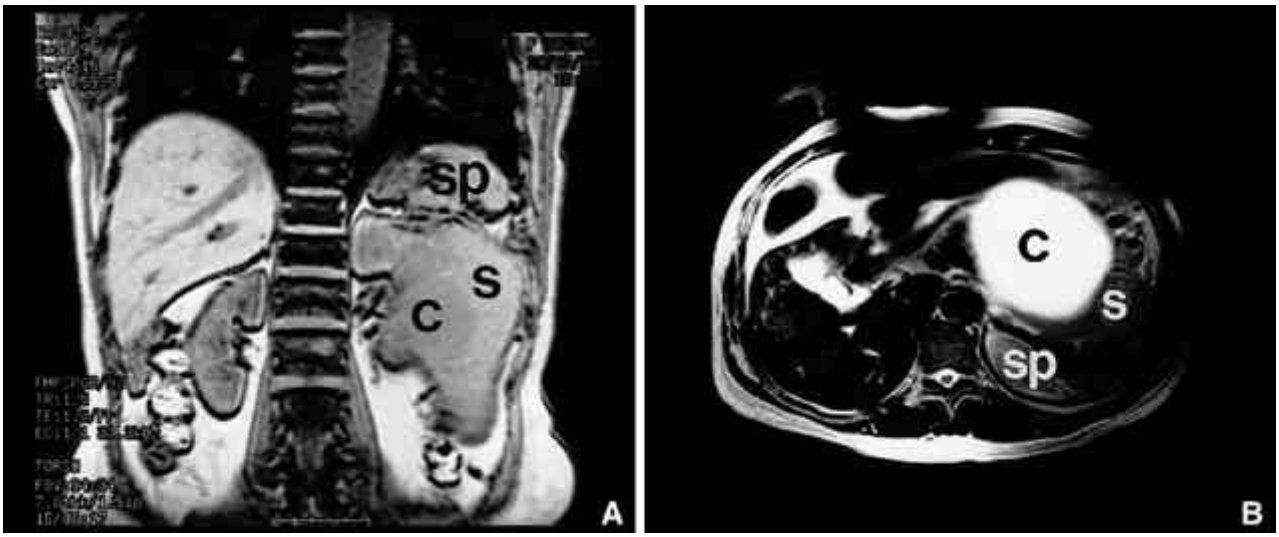

Fig. 2. Coronal T1-weighted (A) and axial T2-weighted (B) magnetic resonance images. Solid component of the mass shows the same signal changes as the normal splenic tissue, but separate completely from the spleen. C, cystic component of the mass; S, solid component of the mass; SP, spleen.

The inner surface of the cyst was smooth and was partly covered with grayish white lamellated keratin materials (Fig. 3). The cystic wall measured 0.1 cm in thickness and the intrapancreatic accessory spleen measured 1.7 cm in maximal thickness. On microscopic examination, the cyst was lined by stratified squamous epithelium with keratinization, but lacking hair and skin appendages. These findings were corresponded to the epidermoid cyst (Fig. 4A). The intrapancreatic accessory spleen showed the regression of white pulps and was separated from the pancreas only by a thin fibrous capsule (Fig. 4B). The spleen was unremarkable.

The patient's postoperative course was uneventful and the patient was discharged on the ninth postoperative day. During the follow-up of several months, the patient was well and enjoying a normal life.

DISCUSSION

Heterotopic splenic tissue can be categorized into two types, splenosis and accessory spleen. Splenosis occurs when the splenic tissue is autotransplanted through surgical intervention or traumatic splenic rupture. Accessory

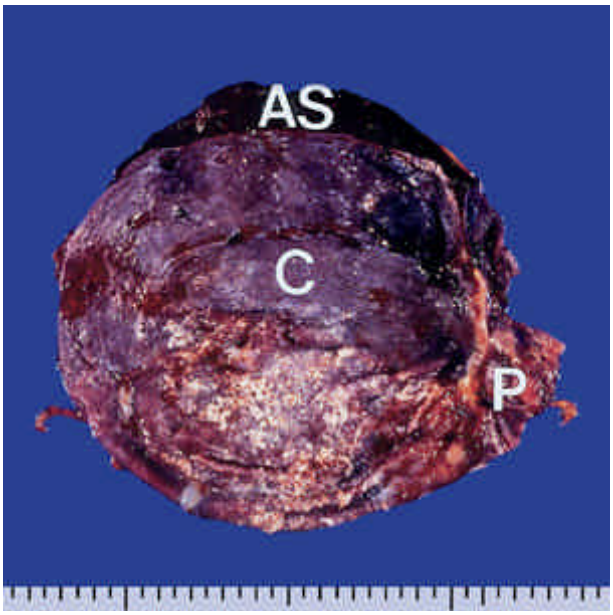

Fig. 3. Gross finding of the cystic mass in cut section. A portion of the outer surface of the cyst is covered with the accessory spleen in the pancreatic tail. C, cyst; AS, accessory spleen; P, pancreatic tail.

spleen is a congenitally duplicated splenic tissue in heterotopic location. Reportedly, accessory spleens are found in approximately 10% of the population, and the 80% of accessory spleens is located around the splenic hilum, although they could be located anywhere in the intraperitoneal cavity from the diaphragm to the pelvis. They occur specifically in the gastrosplenic or splenorenal ligaments, in the mesentery, or even in the pelvis or scrotum (1, 4). However, accessory spleens which are located within the pancreas are infrequent, and only 16% of the accessory spleens is within the pancreatic tail as an autopsy study showed (1).

Embryologically, the spleen is a solid organ derived from the mesoderm and has been known to have a less frequency rate of cystic diseases (0.5-2.0%) than other organs (5). Splenic cysts are almost always benign, and are classified into primary (true) or secondary (pseudo) cysts, according to the presence of cellular lining of the inner surface of the cyst wall (6). The 75% of the splenic cysts is post-traumatic pseudocysts, and the rest of them are true cysts which are further classified into parasitic and non-parasitic cysts (7). Non-parasitic cysts include hemangiomas, lymphangiomas, epidermoid cysts and dermoid cysts. The echinococcal cyst is one of the parasitic cysts (7).

Epidermoid cysts are lined with squamous epithelium, but have no skin appendages such as sebaceous glands and sweat glands. The epidermoid cysts of spleen are comparatively rare, accounting for only 10 to 20% of the non-parasitic splenic cysts (5). Furthermore, epidermoid cysts of accessory spleen are so rare that only about 10 cases have been reported since the report by Davidson et al. in 1980 (3). It is interesting that all cases of epidermoid cysts of accessory spleen, including our case, are located in the pancreatic tail. Accordingly the possibility that the epithelium of epidermal cysts of accessory spleen

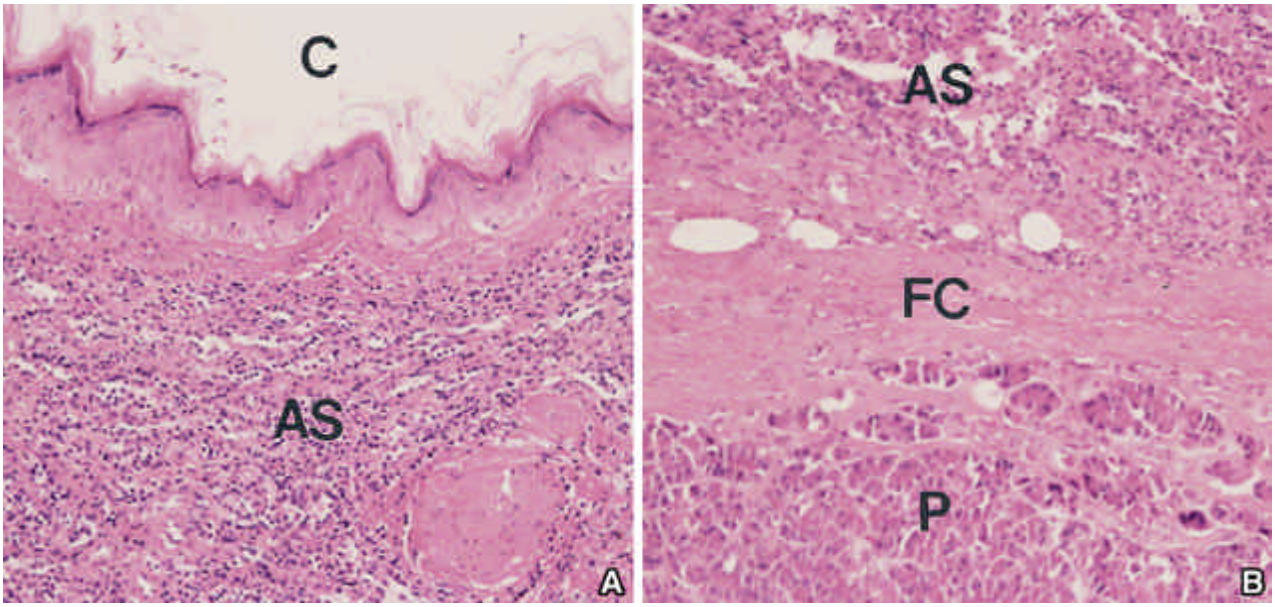

Fig. 4. Microscopic finding of the cyst (H&E, ×100). The cyst is lined by stratified squamous epithelium with keratinization (A). A thin fibrous capsule separate the intrapancreatic accessory spleen from the pancreas (B). C, cyst; AS, accessory spleen; P, pancreas; FC, fibrous capsule.

might have originated from the pancreatic duct has been suggested (8, 9).

The diagnosis of cystic lesions of pancreas and spleen is rather easy with radiologic examination. However, the epidermoid cysts of accessory spleen located within pancreas might be misdiagnosed as pancreatic cysts, so careful evaluation is needed to decide on treatment modality. In our case, the atrophic accessory splenic tissue was revealed to remain in the upper part of the cyst on abdominal computed tomography and magnetic resonance image. Of the cystic diseases of pancreas there are pancreatic pseudocysts, simple pancreatic cysts, cystadenoma, cystadenocarcinoam, and adenocarcinoma with central necrosis.

Surgical resection of cystic tumors of pancreas as well as intrapancreatic accessory spleen is generally considered, in order to confirm benignity or malignancy of the lesions (10). When epidermoid cysts of accessory spleen are small and asymptomatic, surgery might not be necessary. But in practical situations, it is safe and reasonable to perform surgical resection, because the possibility of malignancy cannot be completely excluded. Surgery should employ en-bloc resection of the cyst of accessory spleen, including normal margins of the pancreatic tissue.

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