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Epithelial Cysts in the Intrapancreatic Accessory Spleen that Clinically Mimic Pancreatic Cystic Tumor
– A Report of Two Cases –

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Cystic lesions in the accessory spleen are extremely rare and they present a challenging clinical differential diagnosis. We report here on two cases of epithelial cyst of intrapancreatic accessory spleen that mimicked pancreatic cystic tumor. In both cases, the patients underwent distal pancreatectomy under the impression of a benign cystic tumor of the pancreas. Unilocular or multilocular cysts in the pancreas tail were observed, and these were later shown to be epithelial cysts in the accessory spleen located within the pancreatic tail. The cysts were lined by columnar, cuboidal or stratified squamous epithelium.

Key Words : Accessory spleen; Pancreas; Epithelial cyst

Nonparasitic splenic cysts are classified as either epithelial cysts or as pseudocysts based on the presence or the lack of a cellular lining. Epithelial cysts in the spleen are lined with columnar, cuboidal or squamous epithelium. The pathogenesis is controversial and many hypotheses have been proposed.

Cyst formation in an intrapancreatic accessory spleen is very rare, and Davidson *et al*.[1] first reported a case of epidermoid cyst (a type of epithelial cyst lined wholly by squamous epithelium) in an intrapancreatic accessory spleen in 1980 and 13 similar cases have since been described.[2-10] In the present report, we describe two cases of epithelial cysts arising in an accessory spleen located in the pancreatic tail.

CASE REPORTS

Case 1

The patient was a 32-year-old man with a mass in his pancreas that was detected by ultrasonography during a routine physical check-up. He had occasionally complained of left upper quadrant pain after drinking. The patient was admitted for further evaluation and treatment. Abdominal computed tomography revealed a 7.5×6.5 cm sized unilocular cystic mass with inner fluid debris or hemorrhagic fluid in the tail of pancreas (Fig. 1A). Calcifications within the cystic wall suggested a pseudocyst and so spleen-preserving distal pancreatectomy was performed. Of the tumor markers, only the serum CA19-9 was slightly increased to 53 (IU/mL).

The cut surface of the resected specimen showed a unilocular cyst with relatively glistening luminal surfaces (Fig. 1B). Calcifications and attached pancreatic tissues were also identified.

Case 2

A 49-year-old woman presented with abdominal pain and she was found to have a cystic lesion in the pancreatic tail via abdominal computed tomography, which showed a well-circumscribed cystic tumor with septations (Fig. 1C). A serous or mucinous cystadenoma was suspected and so laparoscopic dis-

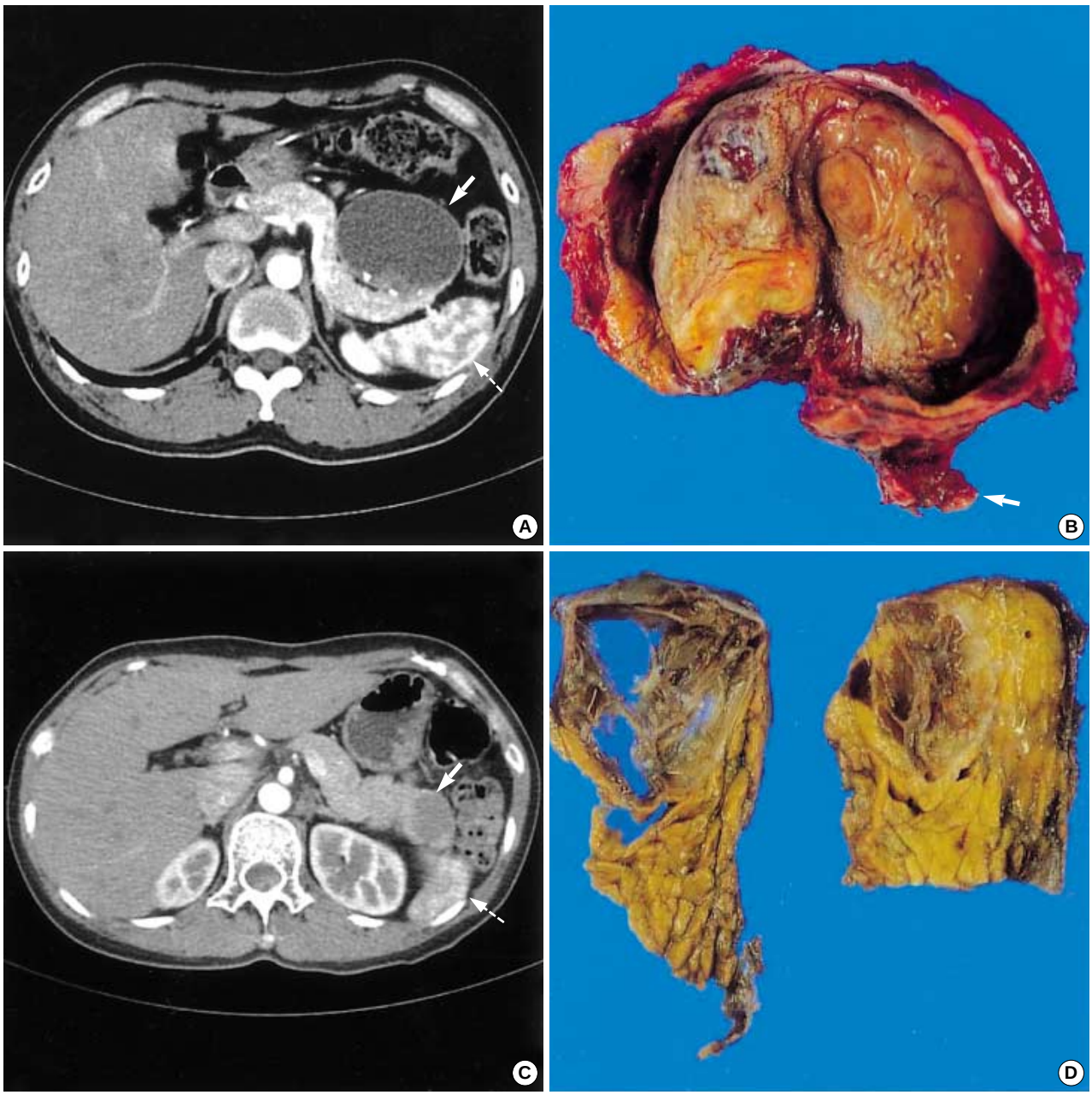

Fig. 1. (A) The abdominal contrast-enhanced computed tomography showing a well circumscribed cystic mass in the pancreatic tail without relation to the spleen (mass: arrow, spleen: dotted arrow) (case 1). (B) A unilocular cyst with glistening surfaces adheres to pancreatic tissues (arrow). The wall seems to be slightly rigid due to calcifications (case 1). (C) A cystic tumor with septations is visible in the pancreatic tail portion (tumor: arrow, spleen: dotted arrow) (case 2). (D) A multilocular cyst with thin walls is well-demarcated from the surrounding pancreatic parenchyma (case 2).

tal pancreatectomy with splenectomy was performed. The preoperative laboratory findings, including CEA and CA19-9, were within normal limits.

Upon gross examination, a 2.0×2.0×1.0 cm sized multilocular cystic mass that was firmly attached to the pancreatic tail was observed. No demonstrable solid component existed (Fig. 1D).

The histological examinations demonstrated an epithelium-lined cystic wall in both cases. In case 1, a unilocular cyst was surrounded by splenic parenchyma with a thick fibrous layer in the intervening stroma. The cystic wall was lined by non-keratinizing squamous epithelium and, in some parts, flattened or cuboidal epithelial cells that continued to the stratified squamous epithelium (Figure not shown). Ciliated columnar cells

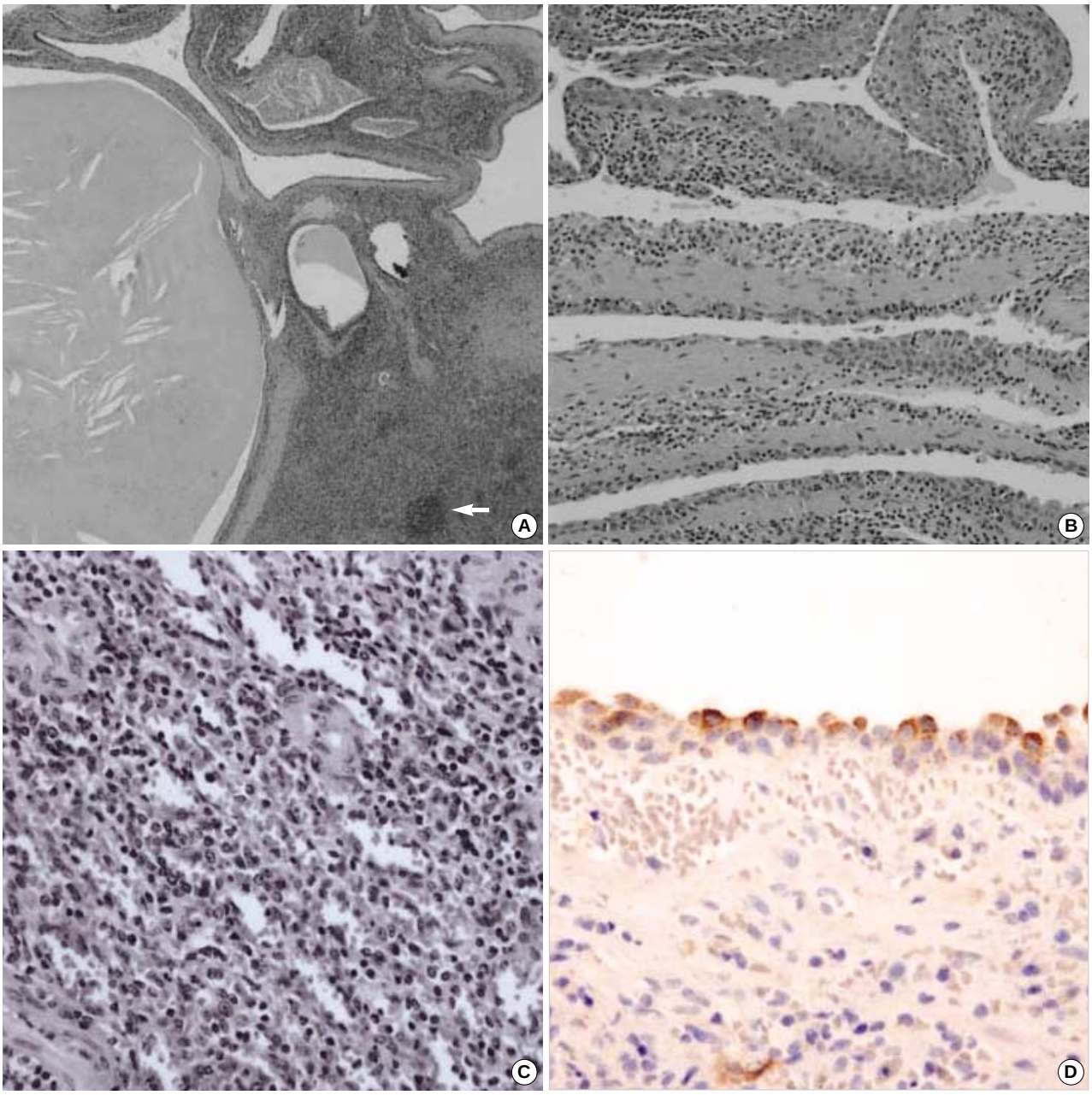

Fig. 2. (A) Multiloculation septated by epithelial cell lined-walls. Note the vague white pulp (arrow) in the thick septum (case 2). (B) Diverse epithelial cells including stratified squamous epithelial cells, ciliated cells and cuboidal cells coexist (case 2). (C) Red pulps rich in capillaries and penetrated by sinuses are identified in cyst walls (case 2). (D) Immunohistochemical staining for calretinin showing focal positivity in the epithelial cell lining of case 1.

were also seen, which were occasionally undermined by stratified squamous epithelial cells (Figure not shown). Similar histological features were found in case 2. In this case, the cyst was multilocular and it was demarcated from the pancreas by thick connective tissue. It had septations that consisted of various epithelial cells with or without splenic red pulps in the intervening stroma (Fig. 2A). The epithelial lining showed a mixture of flattened mesothelial-like cells, ciliated cuboidal cells and stratified squamous epithelial cells (Fig. 2B). Red pulps in the cystic walls were identifiable (Fig. 2C). No hair or skin appendages were identified. To clarify the nature of these epithelial cells, they were immunostained for pancytokeratin and calretinin. In both cases, the epithelial lining was strongly positive for pancytokeratin, and only focal immunoreactivity for calretinin was seen in case 1 (Fig. 2D).

DISCUSSION

Splenic cysts are rare entities, and they are found in only 0.5-2% of resected spleens. Furthermore, only 2.5% of splenic cysts are nonparasitic cysts.[10] Only about 10% of individuals possess an accessory spleen, and only 16% of accessory spleens were identified within the pancreatic tail in an autopsy study.[12]

An epithelial cyst in the intrapancreatic accessory spleen is extremely rare and most such cases are epidermoid cysts, which are lined by stratified squamous epithelium. On the other hand, the present two cases showed a mixture of epithelial cells, thus, the term epithelial cyst should be employed because the cyst walls were lined with several kinds of epithelium, including low cuboidal, ciliated and nonciliated columnar and stratified squamous epithelium. Only two similar cases have been previously reported.[6,7]

Grossly, these cysts are either multilocular or unilocular. The reported cases showed no characteristic features on the diagnostic imaging, and in most instances, they were not preoperatively diagnosed. Hypervascular blush with a density almost equal to that of the spleen on the postcontrast computed tomography might be helpful for diagnosing ectopic splenic parenchyma.[3] However, this sign may not be useful if the amount of splenic tissue is relatively small. Thus, the preoperative imaging is generally not helpful for making an exact diagnosis, except in the cases with plentiful ectopic splenic parenchyma.

Histologically, these cysts could mimic a number of conditions, including a pancreatic lymphoepithelial cyst. A case occurring in an intrapancreatic accessory spleen that had the appearance of a lymphoepithelial cyst has been reported by Tateyama *et al*.[13] Lymphoepithelial cysts characteristically have a stratified squamous epithelial cell layer and an underlying lymphoid stroma that occasionally shows germinal centers. Epithelial cysts in the intrapancreatic accessory spleen, especially epidermoid cyst with inconspicuous red pulp could be misinterpretated as lymphoepithelial cysts. In our cases, the presence of diverse epithelial cells, the absence of germinal centers and the identification of red pulp were helpful for excluding the possibility of a lymphoepithelial cyst. Additional multiple sections would be needed in such cases to identify the splenic parenchyma.

The histogenesis of epithelial cyst is highly controversial. Ough *et al*.[14] suggested that epithelial splenic cysts are the result of an invagination of capsular surface mesothelium with subsequent cystic expansion and metaplastic changes. In agreement with this idea, Burrig[15] suggested that epidermoid cysts seem to be a variant of mesothelial cysts with focal squamous metaplasia on the basis of the immunohistochemical and ultrastructural findings. Lifschitz et al advocated that these cysts are either of a teratomatous derivation or they originate from embryonic inclusions of squamous epithelium, and that neither squamous metaplasia of the mesothelium nor inclusions of mature squamous epithelium appear to be their source of origin, in view of their cytokeratin profiles.[16,17] In a recent study of epithelial cysts that occurred simultaneously in an intrapancreatic accessory spleen and in the spleen itself, the immunohistochemical profiles and electron microscopic examinations of the two cysts produced conflicting results, which suggested that the two cysts differ in their nature and origin. Specifically, it was suggested that the accessory spleen cyst was caused by an embryonic inclusion of the pancreatic duct and that spleen cyst may have been caused by an inclusion cyst of the mesothelium.[7] In our report, various types of epithelial cell were found to be admixed. In case 1, immunostaining for calretinin showed positivity that faded away in the areas of the stratified squamous epithelium, implying a mesothelial genesis. On the other hand, the epithelial cells of case 2 were absolutely negative for calretinin. These results complicate the rationalization of the origin of epithelial cells in splenic epithelial cysts. To precisely elucidate their histogenesis, more intensive studies are required on more cases.

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