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CASE REPORT

초음파 내시경 유도하 세침 흡인술을 이용하여 진단된 췌장의 림프상피성 낭종

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Pancreatic Lymphoepithelial Cysts Diagnosed with Endosonography-guided Fine Needle Aspiration

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Although lymphoepithelial cysts (LECs) of the pancreas are benign lesions, most of them have been treated with surgical resection due to diagnostic difficulty. We report a 66-year-old woman diagnosed with pancreatic LECs. Abdominal ultrasound revealed two masses in the pancreas, which were not visible on the abdominal computed tomography. In an abdominal magnetic resonance imaging, pancreas lesions showed solid tumors, which revealed a low signal intensity on T1-, moderate high signal intensity on T2 weighted images, and homogeneous delayed enhancement in the portal venous phase. Endosonography (EUS) revealed two hypoechoic round masses measuring 1.5 cm and 4.5 cm in the body and tail of the pancreas, respectively. EUS-guided fine needle aspiration (FNA) revealed squamous cells, amorphous keratinous debris, and lymphocytes. The patient was diagnosed with LECs of the pancreas. For the duration of the follow-up period of two years, imaging studies were unchanged. EUS-FNA is useful in making a definite diagnosis and avoiding unnecessary surgery. This is the first case of pancreatic LECs diagnosed with EUS-FNA in Korea. **(Korean J Gastroenterol 2017;69:253-258)**

Key Words: Pancreatic cyst; Pancreas; Endosonography; Fine needle aspiration

INTRODUCTION

Lymphoepithelial cysts (LECs) of the pancreas are rare benign lesions. More than 150 cases of LECs have been published in the literature since the first case, which was reported in 1985.[1] However, the term, LEC was coined two years later in 1987.[2] Although specific radiological features have not been identified, an abdominal computed tomography (CT) scan is the most reliable imaging modality for diagnosing patients with LECs.[3] The majority of cases are macrocystic lesions with mixed patterns of solid and cystic components, depending on the keratin composition.[4] However, a few cases with solid lesions, due to compacted keratin contents, have been reported.[5] Recently, pancreatic lesions have accurately been diagnosed with endosonography-guided fine needle aspiration (EUS-FNA).[5] The treatment methods of pancreatic LECs have changed with the improvement of diagnostic yields. In most patients with LECs, surgical resection

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has been the treatment of choice prior to the EUS era.[6] However, in recent years, conservative treatment has been used in most cases.[5]

Herein, we report a curious case of an asymptomatic 66-year-old woman with LECs of the pancreas detected incidentally. Hypoechoic lesions, which were not visible on the abdominal CT, were revealed on the abdominal ultrasound. Abdominal magnetic resonance imaging (MRI) and EUS revealed two solid masses in the body and tail of the pancreas. Definite diagnosis was obtained using EUS-FNA, and the patient was managed conservatively. We reported an atypical case of pancreatic LECs with a literature review.

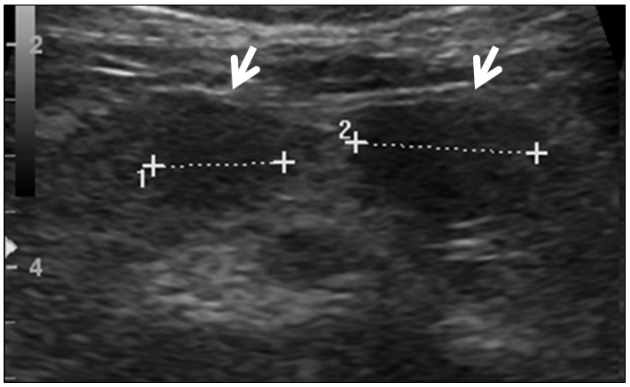

Fig. 1. Abdominal ultrasonography revealing approximately 1.2 cm and 1.6 cm sized low echoic round lesions (arrows).

CASE REPORT

A 66-year-old woman was referred to our hospital for further evaluation of pancreatic masses. One week prior to her visit, two hypoechoic round masses were detected in her pancreas using an abdominal ultrasound during a health surveillance (Fig. 1). She had no relevant prior medical, surgical, smoking, or alcohol abuse history, and no specific symptoms related to the pancreatic lesions. Her physical examination was unremarkable. Laboratory data were: white blood cell

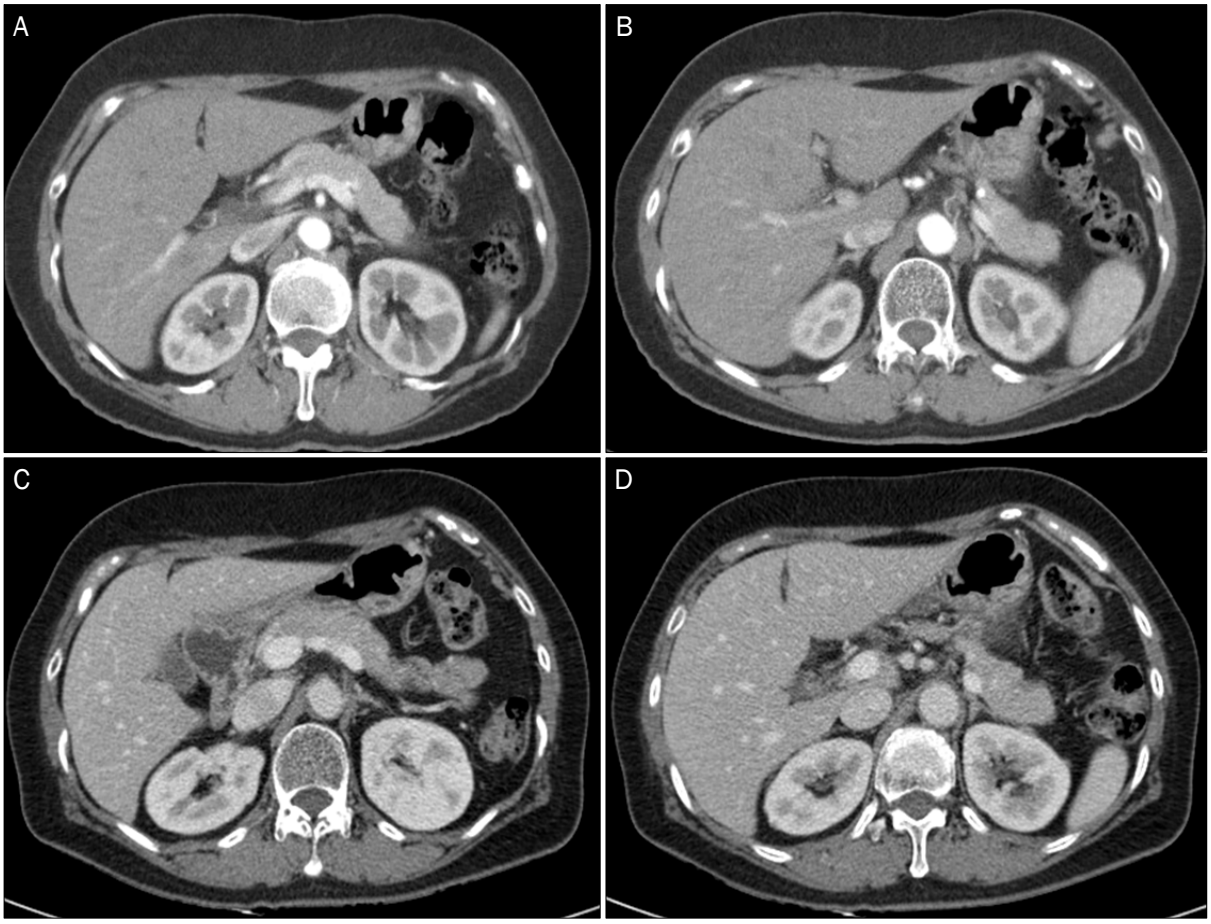

Fig. 2. Axial contrast-enhanced abdominal computed tomography image showing no evidence of abnormal lesions and a normal pancreatic duct (A, B, arterial phase; C, D, delayed phase).

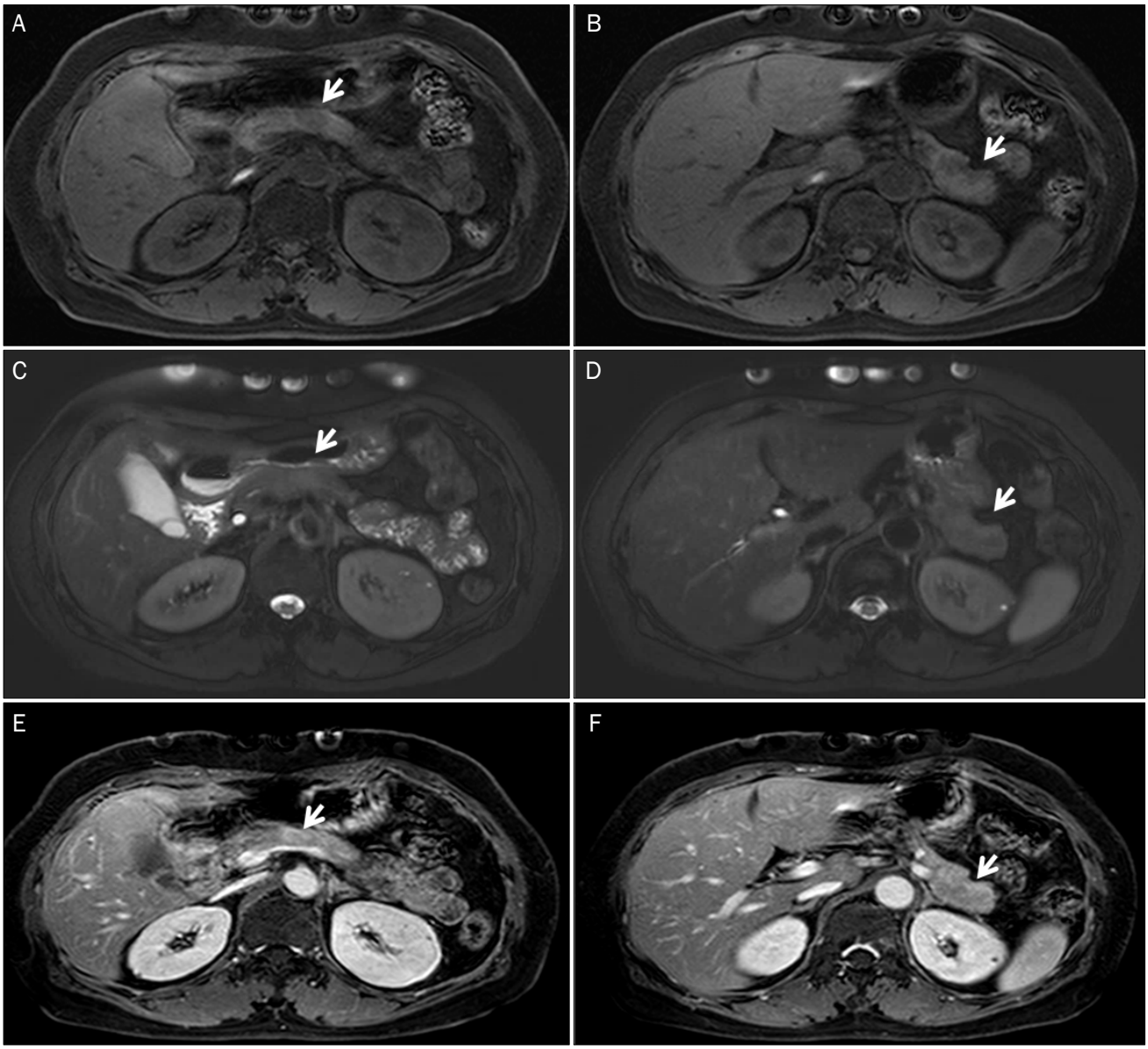

Fig. 3. Abdominal magnetic resonance images. Pre-contrast T1 weighted axial magnetic resonance images showing hypointense masses, 1.5 cm (A, arrow) and 4.5 cm (B, arrow) sized round shapes in the body and tail of the pancreas, respectively. They show moderate high signal intensity on T2 weighted images (C, D) and homogeneous delayed enhancement in the portal venous phase after contrast enhancement (E, F), suggesting solid tumors (arrows).

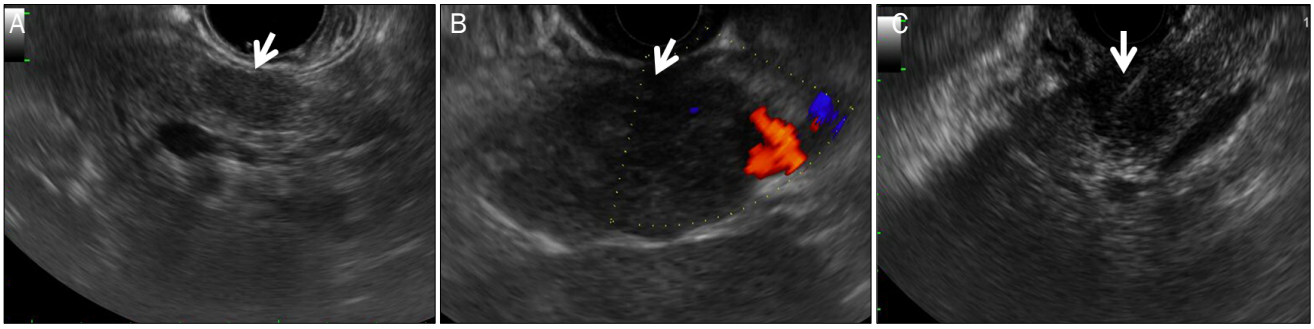

Fig. 4. Endosonography images. Radial endosonography revealing 1.5 cm (A, arrow) and 4.5 cm (B, arrow) sized hypoechoic, well-demarked, and round lesions in the body and tail of the pancreas, respectively. (C, arrow) Endosonography-guided fine needle aspiration using 22G needle.

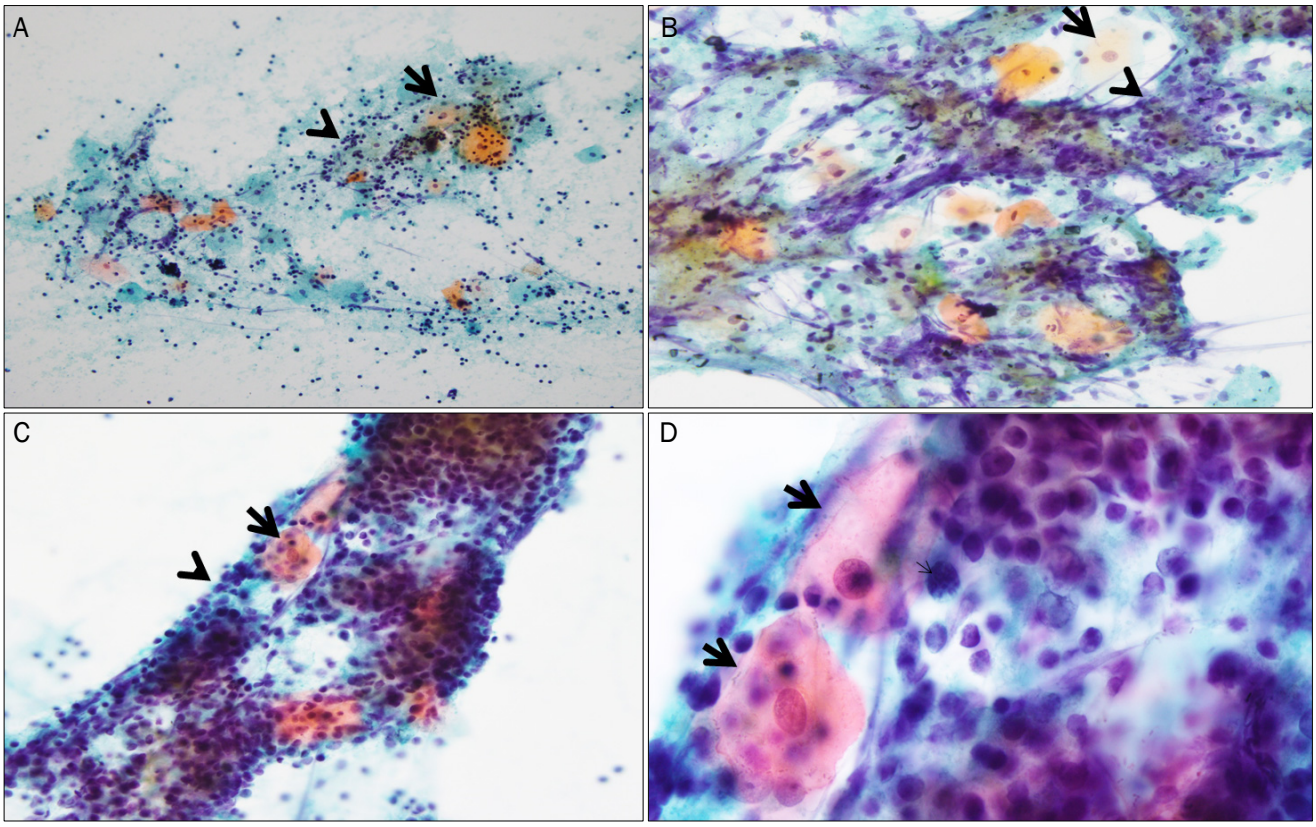

Fig. 5. Cytologic smear obtained with two rounds of endosonography-guided fine needle aspiration (A and B obtained from 1st exam; C and D obtained from 2nd exam). Viable benign squamous cells (arrows) with lymphoid tissues (arrowheads) were seen, which were suggestive of lymphoepithelial cyst (Papanicolaou stain; A, ×200; B, C, ×400; D, ×1,000).

count of 4,500/mm^3, hemoglobin level of 12.2 g/dL, platelet count of 183,000/mm^3, aspartate aminotransferase of 23 IU/L, alanine aminotransferase of 18 IU/L, γ-glutamyl transferase of 52 IU/L, alkaline phosphatase of 112 IU/L, total bilirubin of 0.7 mg/dL, amylase of 31 IU/L, lipase of 21 IU/L, and CA19-9 of 4.69 U/L.

A contrast enhanced dynamic abdominal CT scan did not reveal any abnormal lesions or pancreatic duct dilatation (Fig. 2). MRI revealed two lesions: A 1.5 cm low-attenuated round mass on the pancreatic body and a 4.5 cm elongated mass on the pancreas tail. The two lesions showed low signal intensity on T1-weighted images, moderate high intensity on T2 weighted images, and homogeneous delayed enhancement in the portal venous phase after contrast enhancement, which suggested solid masses (Fig. 3). EUS showed well-demarcated, solid appearing, hypoechoic, and heterogeneous tumors, 1.5 cm and 4.5 cm in the body and tail of the pancreas, respectively (Fig. 4). Two rounds of EUS-FNA with 22G needle (22G Echotip; Wilson-Cook, Winston Salem, NC, USA) were performed at each tumor site. Pathology demonstrated abundant lymphoid tissues, mature, keratinizing squamous epithelia, and keratinized materials, which were compatible with LECs (Fig. 5). Mucinous goblet-like cells and acute inflammation were not seen. Follow-up image studies, including an abdominal CT scan three months after the diagnosis and 2 EUS every six months showed no interval changes. We performed another EUS-FNA at 9 months due to concerns of malignancy, which was consistent with LECs. We finally diagnosed the patients with pancreatic LECs using repeated EUS-FNAs and follow-up imaging studies two years later.

DISCUSSION

Pancreatic LECs are true cystic lesions, accounting for approximately 0.5% of all pancreatic cysts.[4] LECs are usually seen in middle-aged men (man-to-woman ratio, 4:1; mean age, 56 years).[7] Patients with LECs are generally asymptomatic, but a few patients have gastrointestinal symptoms, including abdominal pain, vomiting, diarrhea, weight loss, and fever.[6] LECs may occur in any part of the pancreas, and most

of them are single lesion with unilocular (46%) or multilocular cyst (54%), and have a median size of 4.5 cm (range, 0.5-17 cm).[6]

LECs consist of keratinized material lined by mature, keratinizing squamous epithelium, and surrounded by lymphoid tissue.[4] Plausible pathogenic mechanisms include ectopic pancreatic tissues in the peripancreatic lymph nodes, aberrant positioning of branchial cleft cysts at embryogenesis, or squamous metaplasia in intrapancreatic ducts.[4]

Preoperative diagnosis of patients with LECs is difficult. In half of the cases of LEC, serum cancer antigen 19-9 was elevated.[3] A review of 117 patients with LECs reported that an accurate preoperative diagnosis was obtained in only 22% of patients.[6] However, a recent study reported that 11 out of 17 cases were diagnosed with EUS-FNA.[5] Abdominal cross-sectional images showed characteristic features with some variations. Ultrasound image findings revealed mosaic patterns that depended on the degree of keratin formation.[3] Abdominal CT scans showed enhancements of the wall and septum of the cyst, low density cystic lesions without enhancement, and no pancreatic duct dilatation.[3] MRIs often reveal a high intensity of cyst fluids of pancreatic LECs on T1 weighted images, and a lower intensity on T2 and diffusion weighted images, compared with water.[3] Pancreatic LECs were sometimes seen as solid, homogenously hypointense masses on T1 weighted images; gadolinium-contrast administration enhances the rim while the hypointensity of the central core remains constant.[8] Our case had atypical features, including low echoic round lesions on the abdominal ultrasound, which were not visible by the abdominal CT scan. We initially suggested that these LECs were iso-dense masses on the surrounding pancreas, thereby not needing to be identified. However, MRI revealed two solid masses with homogeneous delayed enhancement in the portal venous phase after contrast enhancement. EUS can provide additional image features and thus be very useful in identifying the invisible and iso-dense masses.[5] An EUS case series of 9 patients with LECs reported a solid-appearing hypoechoic and heterogeneous mass with subtle post-acoustic enhancement in 5 cases and pure cystic lesions in 4 cases.[5] According to the density of the keratinized material, LECs can either be pure cysts, mixed, or solid tumors.[4,9] In addition, keratin components of pancreatic LECs can take a liquid, sludge, or solid form.[5] Sometimes, keratin materials inside the cyst can create distinguishing features, including "cheerios-like" appearance[10,11] or a multiple floating ball-like appearance.[12] In our case, EUS revealed clear images of the two hypoechoic round solid lesions at the body and tail of the pancreas.

EUS-FNA is a useful tool to accurately diagnose patients with LECs.[9] Despite atypical imaging findings, definite diagnosis can be achieved using EUS-FNA, which revealed typical LEC features with abundant mature lymphocytes and scattered squamous epithelia, allowing us to avoid unnecessary surgery.[3,9] The number of cases that accurately diagnosed LECs before surgery has been improved by 65%[5] using EUS-FNA. However, cases with high aspirate carcinoembryonic levels or mucin component are difficult to diagnose from mucinous cystic neoplasm or intraductal mucinous neoplasm.[13] Cytological results should be considered in conjunction with EUS findings to avoid misdiagnosis with mucinous cystic neoplasm.

Before the EUS era, most cases of LECs were diagnosed and treated with surgical resection due to the fear of malignancy. A recent report, including more than 100 cases, revealed that half of the patients with LECs had been treated with surgical resection due to difficulties in its preoperative diagnosis.[3] Surgery should not be avoided in cases where malignancy cannot be ruled out.[14] Patients can be misdiagnosed as having pancreatic masses or cystic neoplasms. However, a current EUS-based case series reported that the majority of patients with LECs avoided surgery for these benign lesions using diagnostic EUS-FNA.[5] In our case, findings from abdominal CT or magnetic resonance imaging were not useful in diagnosing our patient with pancreatic LECs. EUS imaging and aspirate provided accurate clues for a definite diagnosis.

Along with epidermoid cysts (EC) and dermoid cysts (DC), LECs are squamous-lined cysts of the pancreas.[15] Differential diagnosis of these three cystic lesions is difficult in atypical cases. Whereas LEC occurs predominantly in men, EC and DC occur with similar frequency in both sexes. Histopathology reveals distinguishing features for each type of cysts: lymphoid follicles in LEC; splenic tissue in EC; and sebaceous material or hair follicles in DC. Typical features include the same enhancement pattern as the spleen in EC, and all kinds of densities, such as fluid, soft tissue, fat, and calcification in DC. Because these lesions are benign, differential diagnosis from other neoplastic cystic lesions is important.

In conclusion, our patient presented atypical imaging fea-

tures with solid masses on abdominal magnetic resonance imaging, which was not visible on abdominal CT scan imaging. EUS imaging and EUS-FNA are very useful for the diagnosis of patients with LECs. Unnecessary surgery can be avoided when utilizing a proper diagnostic technique, i.e. the use of EUS. We report the first case of pancreatic LECs diagnosed using EUS-FNA in Korea.

REFERENCES

1. Lüchtrath H, Schriefers KH. A pancreatic cyst with features of a so-called branchiogenic cyst. Pathologe 1985;6:217-219.
2. Truong LD, Rangdaeng S, Jordan PH Jr. Lymphoepithelial cyst of the pancreas. Am J Surg Pathol 1987;11:899-903.
3. Terakawa H, Makino I, Nakagawara H, et al. Clinical and radiological feature of lymphoepithelial cyst of the pancreas. World J Gastroenterol 2014;20:17247-17253.
4. Adsay NV, Hasteh F, Cheng JD, et al. Lymphoepithelial cysts of the pancreas: a report of 12 cases and a review of the literature. Mod Pathol 2002;15:492-501.
5. Dalal KS, DeWitt JM, Sherman S, Cramer HM, Tirkes T, Al-Haddad MA. Endoscopic ultrasound characteristics of pancreatic lymphoepithelial cysts: a case series from a large referral center. Endosc Ultrasound 2016;5:248-253.
6. Mege D, Grégoire E, Barbier L, Del Grande J, Le Treut YP. Lymphoepithelial cyst of the pancreas: an analysis of 117 patients. Pancreas 2014;43:987-995.
7. Barbaros U, Erbil Y, Kapran Y, Bozbora A, Ozarmagan S, Bilgiç B. Lymphoepithelial cyst: a rare cystic tumor of the pancreas which mimics carcinoma. JOP 2004;5:392-394.
8. Osiro S, Rodriguez JR, Tiwari KJ, et al. Is preoperative diagnosis possible? A clinical and radiological review of lymphoepithelial cysts of the pancreas. JOP 2013;14:15-20.
9. Nasr J, Sanders M, Fasanella K, Khalid A, McGrath K. Lymphoepithelial cysts of the pancreas: an EUS case series. Gastrointest Endosc 2008;68:170-173.
10. Gao W, Masuda A, Matsumoto I, et al. A case of lymphoepithelial cyst of pancreas with unique "cheerios-like" appearance in EUS. Clin J Gastroenterol 2012;5:388-392.
11. Matrone A, Russo M, Mollica C, et al. Lymphoepithelial pancreatic cyst: an atypical benign pancreatic mass presenting with a “cheerios-like” appearance. JOP 2010;11:170-172.
12. Matsubayashi H, Aikawa Y, Sugiura T, Sasaki K, Hotta K, Ono H. Pancreatic lymphoepithelial cyst showing multiple floating ball-like appearances. J Gastrointestin Liver Dis 2016;25:239-242.
13. VandenBussche CJ, Maleki Z. Fine-needle aspiration of squamous-lined cysts of the pancreas. Diagn Cytopathol 2014;42:592-599.
14. Martin J, Roberts KJ, Sheridan M, et al. Lymphoepithelial cysts of the pancreas: a management dilemma. Hepatobiliary Pancreat Dis Int 2014;13:539-544.
15. Kubo T, Takeshita T, Shimono T, Hashimoto S, Miki Y. Squamous-lined cyst of the pancreas: radiological-pathological correlation. Clin Radiol 2014;69:880-886.