Pancreatic Lymphoepithelial Cyst Showing Multiple Floating Ball-like Appearances

Hiroyuki Matsubayashi1, Yoshiko Aikawa2, Teiichi Sugiura2, Keiko Sasaki3, Kinichi Hotta1, Hiroyuki Ono1

INTRODUCTION

A lymphoepithelial cyst (LEC) is a rare pancreatic lesion, histologically showing squamous epithelia, dense lymphoid tissues, and a keratin substance. Cross-section images of the pancreatic LEC typically show a well demarcated unilocular or multilocular cyst without a solid component. Here we report a rare case of pancreatic LEC in which multiple floating ball-like components were depicted via endoscopic ultrasound. The ball-like components were also depicted by various imaging methods such as computed tomography (CT) showing low-density components, T1-weighted magnetic resonance imaging (MRI) and diffusion-weighted imaging (DWI) showing high-intensity components, and T2-weighted MRI showing low-intensity components. The ball-like components in all images were not well enhanced. Laparotomic cyst resection was performed, and the surgical material revealed keratin balls inside the pancreatic LEC. Keratin components of a pancreatic LEC can take a liquid, sludge, or solid form. Clinicians must be aware of the variations in imaging to facilitate the differentiation and management of pancreatic cystic lesions.

Key words: pancreas – lymphoepithelial cyst – ball-like – keratin substance – endoscopic ultrasonography.

ABSTRACT

A lymphoepithelial cyst (LEC) is a rare pancreatic lesion, histologically showing squamous epithelia, dense lymphoid tissues, and a keratin substance. Cross-section images of the pancreatic LEC typically show a well demarcated unilocular or multilocular cyst without a solid component. Here we report a rare case of pancreatic LEC in which multiple floating ball-like components were depicted via endoscopic ultrasound. The ball-like components were also depicted by various imaging methods such as computed tomography (CT) showing low-density components, T1-weighted magnetic resonance imaging (MRI) and diffusion-weighted imaging (DWI) showing high-intensity components, and T2-weighted MRI showing low-intensity components. The ball-like components in all images were not well enhanced. Laparotomic cyst resection was performed, and the surgical material revealed keratin balls inside the pancreatic LEC. Keratin components of a pancreatic LEC can take a liquid, sludge, or solid form. Clinicians must be aware of the variations in imaging to facilitate the differentiation and management of pancreatic cystic lesions.

Key words: pancreas – lymphoepithelial cyst – ball-like – keratin substance – endoscopic ultrasonography.

Abbreviations: CA 19-9: carbohydrate antigen 19-9; CEA: carcinoembryonic antigen; DWI: diffusion-weighted image; LEC: lymphoepithelial cyst; IPMN: intraductal papillary neoplasm; MCN: mucinous cystic neoplasm.

CASE REPORT

A 67-year-old man with mild epigastric pain was referred to our hospital to investigate an abdominal cyst located between the pancreas tail and the gastric body. He had a past history of gallstones, and he had undergone laparoscopic cholecystectomy eight years before. During those eight years, the size of the cyst, confirmed by computed tomography (CT), increased from 1 cm to 2.5 cm (Fig. 1). His blood examinations...
showed no remarkable abnormality including CEA or CA 19-9. Endoscopic ultrasonography (EUS) revealed multiple high-echoic components with partial acoustic shadows floating in the cyst, which appeared to be contiguous to the pancreas tail (Fig. 2). Magnetic resonance imaging (MRI) (Fig. 3) demonstrated multiple ball-like lesions depicted in a high-intensity signal through a T1-weighted image and a diffusion-weighted image (DWI) and in low intensity by T2. Magnetic resonance cholangiopancreatography (MRCP) showed a slightly high intensity signal at the cyst, though lower than that of a renal cyst. Dynamic MRI demonstrated a faint enhancement only at the cystic wall and no enhancement at the ball-like components. 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) showed no abnormal uptake around the pancreas.

For a preoperative diagnosis, several candidates were listed. A LEC and an epidermoid cyst were considered if a pancreatic origin was assumed or a duplication cyst and enteric cyst for a gastric origin; however, a definitive diagnosis could not be obtained because of the difficulty in interpreting the intracystic ball-like lesions and the cyst’s origin. EUS-FNA was not performed, in order to avoid critical complications due to leakage, such as chemical peritonitis [5] or tumor dissemination [6].

A laparotomic enucleation of the pancreatic cyst was performed. The surgical material revealed multiple cheesy balls inside the cyst (Fig. 4). Microscopically, these balls were composed of keratinized substances, and the cyst wall was mostly covered by stratified squamous epithelium and partially by columnar epithelium. Within the cystic wall, many lymph follicles and sebaceous glands were seen, but no hair follicles (Fig. 5). A small amount of the pancreatic parenchymal tissue was recognized beside the cyst, and the pathological diagnosis was pancreatic LEC.
DISCUSSION

A LEC is a rare pancreatic cystic lesion, and the entity of this disease has been established by the accumulation of individually reported cases or small numbers of series. Mege et al. [2] extracted and summarized the critical information from 117 previous studies in the literature. According to this review, a pancreatic LEC is often recognized in middle-aged to elderly males (age average: 55 years; range: 20–82 years old; gender: 91 males vs. 26 females) and is sometimes accompanied by abdominal pain (43%) and an elevation of serum tumor marker CA19-9 (55%). The LECs form as both unilocular (40%) and multilocular (60%) cysts, uniformly distributed in the pancreatic head, body and tail [1, 2, 7]. These findings are not really specific to this particular disease, however, often being recognized in pancreatic neoplastic cystic lesions such as mucinous cystic neoplasm (MCN) or intraductal papillary neoplasm (IPMN). An accurate preoperative diagnosis has been obtained in only 25% of pancreatic LECs [2].

Meanwhile, a pancreatic LEC has some histological characteristics such as rich keratin substances expelled from the squamous epithelia covering its cystic wall, with another feature being the aggregation of lymph cells within the wall. This keratin substance can be determined by EUS-FNA, and this, coupled with a cyst fluid concentration of CEA and CA19-9, is diagnostic to some degree [4, 8]. However, this also has several diagnostic pitfalls [4, 8]. Besides, FNA from pancreatic cystic lesions can rarely cause leaks of the cystic fluid [5, 6], which result in severe events such as tumor dissemination and chemical peritonitis. Therefore, image diagnosis is important.

Cross-sectional images of the pancreatic LEC typically show a well-demarcated cyst without a solid component [9], often protruding from the pancreas [3], as in our case. Kim et al. [3] compared CT images in the shapes of various types of pancreatic cystic lesions including serous cystic neoplasm (30 cases), MCN (11), pseudocysts (9), and LEC (8 cases); the researchers demonstrated that an extrapancreatic location (protruding out from the pancreas) is significantly more readily recognized in LEC than in other types of cysts. Images of the intracystic fluid of a LEC vary depending on the level of keratin formation, especially using ultrasonography (US) and MRI. When using US, the cyst fluid usually appears in a low echo but rarely in a solid-like pattern [5, 10]. With MRI, in comparison with water, the cyst fluid of a pancreatic LEC is often demonstrated in higher intensity by T1W and DWI images and in lower intensity by T2W images [9].

When using these standard images, we must bear in mind the possible variations of a pancreatic LEC. The current case demonstrated multiple ball-like components floating inside the cyst, a phenomenon that has rarely been reported to date. These ball-like components were histologically proven to be keratin substance, which can form as a fluid, as sludge, or as a solid. In another rare form of pancreatic LEC, Maekawa et al. [11] reported an intracystic nodule projection that histologically consisted of a lymphoid proliferation, depicted by an abnormal uptake of FDG. Raval et al. [7] reported that 78% of LECs contain epithelial cells producing mucin, which affects the cystic fluid viscosity and the corresponding clinical...
images. Such factors as keratinization, lymphoid tissue, and mucin may alter the image of a pancreatic LEC into atypical appearances.

**CONCLUSION**

Pancreatic LECs may form various types of images depending on the proliferation levels of mucin, lymphoid tissue, and keratin substance. Therefore, clinicians must be aware of the possible variation in images of LEC as well as of the features of other pancreatic cystic lesions [12].

**Conflicts of interest and sources of funding:** None to declare.

**Authors’ contributions:** H.M. clinically managed the patient and wrote the manuscript; Y.A. and T.S. performed surgery; K.S. worked on the pathological diagnosis; K.H., T.S. and H.O. reviewed the manuscript for its intellectual content. All authors read and approved the final version of the manuscript.

**REFERENCES**