Lymphoepithelial cysts of the pancreas: The use of endoscopic ultrasound-guided fine-needle aspiration in diagnosis

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Lymphoepithelial cysts (LECs) are rare non-neoplastic lesions that can appear as a complex cyst or a mass in the pancreas. Cytology from endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA) can be helpful in making a diagnosis with the aim of avoiding unnecessary surgical resection. A case involving a 51-year-old woman with lower abdominal pain who was found to have a multiloculated cystic lesion at the junction of the pancreatic body and tail is described. Cytology from EUS-FNA was consistent with a pancreatic LEC. The lesion was managed conservatively and follow-up imaging of the cyst over the following two years was unchanged. The patient remains clinically well. Cytology from EUS-FNA can help distinguish LECs from cystic neoplasms, thus preventing radical surgical resection of this benign pancreatic cyst.

Key Words: Cytology; Endoscopic ultrasound; Lymphoepithelial cysts; Pancreas

Cystic lesions of the pancreas can be divided into true cysts, pseudocysts and cystic neoplasms. A true cyst is distinguished by the presence of an epithelial lining, their benign natural history and developmental origin. Lymphoepithelial cysts (LECs) are a rare type of true cyst that can mimic pseudocysts and cystic neoplasms. Therefore, it is imperative to find a reliable way to diagnose LECs to avoid unnecessary radical surgical resection. The present report describes a 51-year-old woman in whom the diagnosis of an LEC was made on cytology by endoscopic ultrasound-guided fine-needle aspiration (EUS-FNA), who was thus managed conservatively.

CASE PRESENTATION

A 51-year-old woman presented with a one-month history of left lower quadrant abdominal pain. Her medical history was significant for sarcoidosis and type 1 diabetes. There was no history of alcohol use, gallstone disease or acute pancreatitis. An abdominal computed tomography (CT) scan revealed a multiloculated cystic lesion measuring 4.2 cm × 2.6 cm × 3.2 cm in the proximal body of the pancreas, with some minor peripancreatic lymphadenopathy. She had not undergone previous abdominal imaging. A follow-up CT scan one month later showed no change in the size of the lesion, but resolution of the lymphadenopathy was evident. Serum carbohydrate antigen

Les kystes lymphoépithéliaux du pancréas : Le recours à l'aspiration à l'aiguille sous échoguidage pour poser le diagnostic

Les kystes lymphoépithéliaux (KLE) sont des lésions non néoplasiques rares qui peuvent prendre la forme d'un kyste complexe ou d'une masse dans le pancréas. La cytologie par aspiration à l'aiguille sous échoguidage (AAE) peut permettre de poser un diagnostic tout en évitant une résection chirurgicale inutile. Est décrit le cas d'une femme de 51 ans ayant des douleurs abdominales basses chez qui on a découvert des lésions kystiques multiloculées à la jonction du corps et de la queue du pancréas. La cytologie par AAE était compatible avec un KLE du pancréas. La lésion a été traitée de manière prudente, et l'imagerie de suivi du kyste au cours des deux années suivantes n'a pas évolué. La patiente est demeurée bien sur le plan clinique. La cytologie par AAE peut contribuer à distinguer un KLE d'un néoplasme, évitant ainsi la résection chirurgicale radicale de ce kyste bénin du pancréas.

(CA) 19-9 was elevated at 350 U/L (reference range lower than 37 U/L); the remainder of her laboratory results, including a complete blood count, electrolytes and liver enzymes, were normal.

Endoscopic ultrasound was performed, revealing a solid mass at the pancreatic neck measuring 2.5 cm in diameter (Figure 1). There was no accompanying ductal dilation. Several enlarged celiac and peripancreatic lymph nodes were seen. An EUS-FNA of the largest celiac lymph node showed benign lymphocytes, but the aspirate from the pancreatic mass revealed large amounts of well-differentiated squamous epithelium, keratinaceous and amorphous debris, and the presence of lymphoid cells (Figure 2). There were no suspicious features of carcinoma. A subsequent magnetic resonance (MR) image showed a well-defined, multilocular cystic lesion at the junction of the body and tail, measuring 2.4 cm × 2 cm (Figure 3). On fat-suppressed T1 sequence, the cyst showed a high heterogeneous signal and an intermediate signal on T2 MR imaging, with no solid component.

A diagnosis of LEC was made based on the fine-needle aspirate of the pancreatic mass and the MR image. The patient was followed with a repeat MR imaging study six months later that showed no interval change. She continued to undergo abdominal ultrasound every six months for the next two years, with the lesion remaining unchanged. Currently, the patient continues to do well clinically.

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Figure 1) A 10 MHz linear endoscopic ultrasound view of a lymphoepithelial cyst in the body of the pancreas during a fine-needle aspiration biopsy

Figure 2) High-power view of cytology from the pancreatic cyst. Fine-needle aspirate showing well-differentiated squamous epithelium, keratinaceous and amorphous debris, and some lymphoid cells

Figure 3) Fat-suppressed T1-weighted magnetic resonance image showing a well-defined cystic lesion at the junction of the pancreatic body and tail (arrow), emitting a high heterogeneous signal

DISCUSSION

LECs are rare, nonmalignant lesions that were first described by Luchtrath and Schriefers in (1) 1985. Since then, fewer than 90 cases have been reported in the English literature. The etiology of LECs remains unclear. Several hypotheses have been proposed, including that these lesions represent squamous metaplasia of either an obstructed intrapancreatic duct or a peripancreatic lymph node, or that they originate from branchial cleft cysts fused with the pancreatic anlage during embryogenesis (2,3). LECs have been described in the parotid and submandibular glands, lung, thyroid and cervical regions (3). To date, there have been no reports of LECs becoming malignant or recurring after surgical resection. The cysts can occur anywhere in the pancreas, most commonly in the tail and body, followed by the head and neck, and have been described as being up to 13 cm in size (3,4).

Clinically, LECs are usually found incidentally, most commonly in middle-aged men during investigations for abdominal symptoms when symptomatic patients can present with abdominal pain, nausea, vomiting, anorexia and weight loss. Physical examination is usually noncontributory. The differential diagnosis includes pseudocysts and cystic neoplasms.

The main issue in the diagnosis of LECs is differentiating them from other cystic lesions of the pancreas, particularly mucinous cystic neoplasms, because the natural history and management of these lesions are very different. The use of imaging, tumour markers, cytology and histology can be helpful in making a diagnosis. CT findings are often nonspecific and may demonstrate a low-attenuated unilocular or multilocular cystic lesion with a thin enhancing rim, as in the present case. However, LECs can also appear as a low-attenuated solid mass due to the large amount of debris and keratinous material often contained within the cyst (5,6). Transabdominal ultrasound and EUS can be used to further support the cystic nature of these lesions. In an EUS case series by Nasr et al (6), pancreatic LECs were commonly found to appear as solid, hyperechoic, heterogeneous masses with subtle postacoustic enhancement. As such, it has characteristics of both a mass lesion and a cyst. This was also the case in our patient. Fat-suppressed T1-weighted MR imaging can be more helpful because the high keratin content of LECs would show a high signal in T1- and low signal in T2-weighted imaging. However,

in our patient, the lesion showed moderate to high signal in both T1- and T2-weighted images.

Tumour markers are less specific. Serum CA 19-9 has been established as a sensitive marker of biliary and exocrine pancreatic cancer; however, despite several case reports (6,7) of LECs with high serum CA 19-9 levels, including the one presented here, there seems to be no clear correlation between the two. Similarly, there have been conflicting reports (5,7-9) on the utility of cyst fluid analysis for CA 19-9, CA 125, amylase and carcinoembryonic antigen.

Cytology from EUS-FNA is a well-established modality for diagnosing most pancreatic cystic lesions. For many purely cystic lesions, cytology can be difficult because of acellular aspirates. However, for LECs, at least 20 cases have been reported in which the diagnosis was made based on CT or EUS-FNA (4,6,8-10). Cytology classically shows abundant anucleated squamous cells, multinucleated giant cells, mature lymphocytes in a background of keratinaceous debris and a lack of neoplastic cells (4,8,10). Most of these features were visible on the cytology sample from our case (Figure 2); a diagnosis of a pancreatic LEC was made based on these results. Follow-up imaging has thus far confirmed the benign course of this lesion.

Interpretation of LECs by EUS-FNA is complicated by frequent contamination of the aspirate by tissues acquired by the needle during the procedure such as with mucinous and glandular epithelium from intestinal sources, making cystic neoplasm difficult to rule out. Very often, LECs can even contain a thick milky, creamy or frothy aspirate, further confusing the diagnosis (6). In the present case, squamous cells were seen but the presence of keratinaceous debris in the aspirate make contamination from the esophagus unlikely. Furthermore, as described by Renou et al (9), the absence of LEC cytology does not rule out a diagnosis of LEC. The difficulty in preoperative diagnosis still leads many LECs to surgical resection.

LECs of the pancreas are a rare, clinically benign lesion that should be considered in the differential diagnosis of any pancreatic cyst. A definitive preoperative diagnosis can prevent radical surgical resection, but can be difficult because LECs can often mimic neoplastic cysts of the pancreas (11). Our case illustrates the diagnostic utility of EUS-FNA, and supports the conclusion that if a patient is largely asymptomatic and cytology from EUS-FNA strongly suggests the diagnosis of LEC, one can safely opt for conservative management with follow-up imaging.

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