Lymphoepithelial cyst of the pancreas: a case report

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BACKGROUND: **Lymphoepithelial cyst of the pancreas is a rare lesion of undetermined pathogenesis that had been documented almost exclusively in males. The literature on this entity is limited to reports of single or a small number of cases.**

METHODS: **The case we described herein was compared with a total of 36 cases reported elsewhere.**

RESULTS: **The 37 cases of lymphoepithelial cyst of the pancreas including our case were reviewed. Lymphoepithelial cysts have uniform and distinctive clinicopathologic features. Approximately 46% of the reported cases were asymptomatic with the lesions found incidentally, and their symptoms were non-specific.**

CONCLUSIONS: **Lymphoepithelial cyst is a rare benign lesion of the pancreas. Fine-needle aspiration biopsy (FNAB) is a rapid and reliable technique that can be used as the first diagnostic step in cases of cystic lesions of the pancreas.**

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KEY WORDS: **lymphoepithelial cyst; pancreas**

Introduction

Lymphoepithelial cyst of the pancreas is a rare lesion of undetermined pathogenesis that had been documented almost exclusively in males.[1] The literature on this entity is limited to reports of single or small numbers of cases. The disease presents as a painless enlargement of one or both parotid glands and does not appear to be associated with other disease processes within the head and neck, or with polycystic disease of the kidney, pancreas, or congenital fibrosis of the liver.[2] Preoperative diagnosis is of quite difficulty.[3] We describe our case herein and compare it with 36 cases reported elsewhere.

Case report

A 43-year-old woman was admitted to our hospital because of 1-month vague epigastric pain without nausea, vomiting, diarrhea, fever, jaundice, and weight loss. She had no significant medical history. Physical examination demonstrated mild tenderness in the epigastrium but no evidence of an acute abdomen. Laboratory studies including functional tests of the pancreas, liver and kidney showed normal values. The levels of serum carbohydrate antigenic determinant (CA19-9) and carcinoembryonic antigen (CEA) were not elevated. Ultrasonography showed multicystic masses with heterogeneous internal echo in the head and tail of the pancreas, measuring 3.8×3.9 cm and 4.0×4.4 cm. CT showed multilocular cystic lesions in the pancreatic tail and body and abnormal contour of the pancreas (Fig. 1). Endoscopic retrograde cholangiopancreaticography (ERCP) demonstrated deviation of the pancreatic duct branches because of the masses, but no stenosis or dilatation was detected, nor communication between the pancreatic ducts and the cyst. In order to further define the masses, EUS-guided fine-needle aspiration biopsy (FNAB) was performed using a 21-gauge Chiba needle mounted on a 10-ml disposable syringe. Histological examination revealed that tissue threads were composed of keratinizing squamous epithelial cells, and some of which showed no nucleus and a few lymphocytes, thereby confirming the diagnosis of lymphoepithelial cyst of the pancreas (Fig. 2). The patient was not subjected to an operation for the cyst because of her mild symptom. She is living well.

Review of the literature

Altogether 45 patients with lymphoepithelial cyst of the pancreas were reported from 1985 to 2001. We reviewed 36 patients with detailed data and the present patient. The cysts were located at the head (7 patients), body (16), tail (11), and peri-pancreas (3). The male

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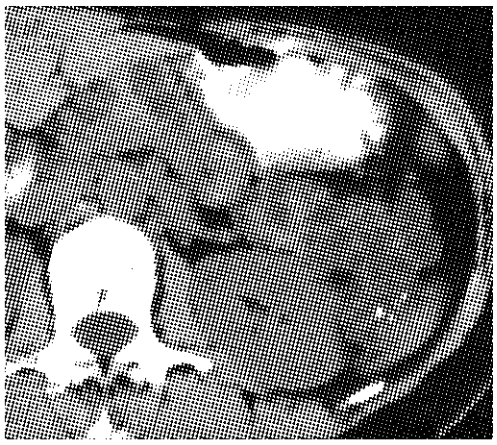

Fig. 1. CT showing a multilocular cystic lesion in the tail and body of the pancreas.

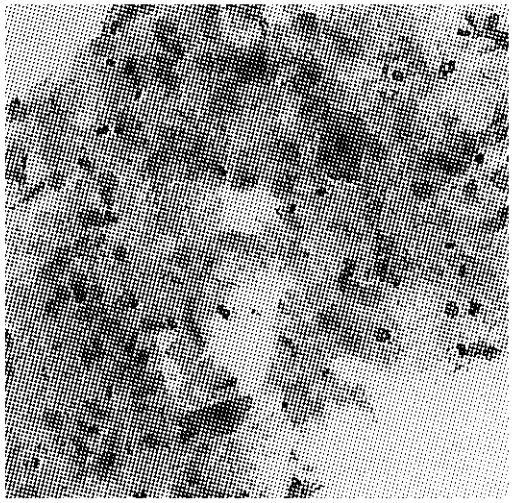

Fig. 2. Tissue threads composed of keratinizing squamous epithelial cells without nucleus but a few lymphocytes (HE original maganification×100).

to female ratio was 3.6:1. The age of the patients ranged from 32 to 75 years (mean 56.7 years), and the course of the disease varied from 6 days to 3 years.

Lymphoepithelial cysts were found by autopsy in 4 patients who died of irrelevant diseases, health examination in 5 patients, and evaluation of diseases irrelevant to the pancreas in 8 patients. Of the remaining 20 patients, 15 complained of abdominal pain, 4 had nausea, vomiting or abdominal malaise, 3 had diarrhea and fever, and 2 had loss of weight and fatigue. Some patients showed more than one symptom.

The levels of serum amylase and lipase in 12 patients were elevated only in 1 patient. Elevated serum levels of CA19-9 were found in 8 of the 15 patients. The level of serum CEA in 5 patients was not abnormal.

Endoscopic retrograde cholangiopancreatography (ERCP) for 14 patients, including the present one, showed the normal duct system in 10 patients, mild changes of the pancreatic duct in 3, and dilatation of the common bile duct in 1. Computed axial tomography (CAT) for 25 patients showed a round mass protruding from the surface of the pancreas into the lesser sac. The lesion of each patient showed a low or water density.

FNAB was performed in 10 of the 33 clinically detected patients, guided by ultrasonography (4), computed tomography (3) and endoscopic ultrasonography (3) respectively. In 4 patients, the fluid of the cyst was taken for laboratory analysis, showing a high level of CEA but an elevation of CA19-9 in 3 patients. In 1 patient, the cystic contents were found to be immunohistologically positive for both CEA and CA19-9. Secondary infection of the cyst was incidentally found only in one patient, who was a 61-year-old man with a 1-week history of epigastric pain, malaise, fever, constipation, abdominal distension, and 7.5×7.2 cm miltilocular cyst in the head of the pancreas revealed by abdominal CT. Postoperative culture of the cystic fluid showed *S. Heidelberg*. Epstein-Barr virus (EBV) in situ-hybridization in one patient showed no evidence of EBV. Surgical resections included local excision in 15 patients, distal pancreatectomy in 12, pancreatoduodenectomy in 2, choledochoduodenostomy in 1, and Roux-en-Y cystojejunostomy in 1. In all the patients, malignancy was not found. Follow-up for 2-6 years showed that symptoms of the cyst disappeared promptly after the operation and all patients were alive and well.

Discussion

Lymphoepithelial cyst of the pancreas was first described by Luchtrath[4] in 1985. In 1987, Truong et al[5] referred it as "lymphoepithelial cyst" in English. To our knowledge, a total of 45 cases have been reported worldwide, but the present case is the first in China.

We reviewed 37 cases with detailed data and found that lymphoepithelial cysts have specific clinicopathological features and mostly affect adult men. Approximately 46% of the reported cases were asymptomatic with the lesions found incidentally at autopsy, physical examination or evaluation of irrelevant diseases. In other cases, the presenting symptoms were non-specific complaints such as abdominal pain, nausea, emesis, diarrhea, fever, loss of weight, and fatigue. But it is unclear whether these symptoms were directly attributable to lymphoepithelial cyst.[6]

The benign biological behavior of all the reported cases suggests that asymptomatic patients or patients with mild symptoms or accurate preoperative diagnosis might be clinically followed up. Conservative surgery should be appropriate for symptomatic patients or when a precise preoperative diagnosis is not achieved.

Intraoperatively, the cyst was primarily spherical,

well-circumscribed, without invading other organs. It was dissected easily from the pancreas, and the non-involved surrounding tissue appeared normal. Grossly, being single, 2-10 cm in diameter, the cut surface of the cyst was well-circumscribed and uni- or multilocular with "caseous", "cheesy", "curd-like" material or with turbid fluid.

Histologically, the cyst had distinctive pathologic features which are essential to its accurate diagnosis. It was composed of a mature, keratinizing, and squamous lining surrounded by lymphoid tissue including a capsule, a subcapsule sinus, germinal centers, and sometimes duct-like structures lined by the squamous or columnar epithelium.[6,7]

The histogenesis of lymphoepithelial cyst is not well understood. Hypotheses include[6] an aberrant branchial cleft remnant that fused with the pancreatic aglage during development; epithelial inclusions or ectopic pancreas in the peripancreatic lymph nodes; squamous metaplasia of an obstructed and dilated pancreatic duct; and an intrapancreatic accessory spleen.

Since lymphoepithelial cyst of the pancreas is a benign lesion, it can be resected locally or by a "wait and see" approach in selected cases. The symptoms attributable to the lesion may disappear immediately after operation and satisfactory prognosis can be ensured.

In summary, lymphoepithelial cyst is quite difficult to distinguish from other cystic neoplasms of the pancreas by clinical manifestations and imaging techniques such as ultrasonography, CT, MRI, EUS, and ERCP. It is diagnosed mostly by postoperative histological examination, and only a few cases could be diagnosed preoperatively by FNAB.[8-11] We consider that FNAB as a rapid and reliable technique may serve the first step in diagnosis of cystic lesions of the pancreas, allowing conservative treatment of symptomatic patients and clinical follow-up of asymptomatic patients.

Competing interest

The author or authors do not choose to response to the statements listed in Instructions for Authors.

References

1 Adsay NV, Hasteh F, Cheng JD, Bejarano PA, Lauwers GY, Batts KP, et al. Lymphoepithelial cysts of the pancreas: a report of 12 cases and a review of the literature. Mod Pathol 2002;15: 492-501.

2 Layfield LJ, Gopez EV. Histologic and fine-needle aspiration cytologic features of polycystic disease of the parotid glands: case report and review of the literature. Diagn Cytopathol 2002;26: 324-328.

3 Fujiwara H, Kohno N, Nakaya S, Ishikawa Y. Lymphoepithelial cyst of the pancreas with sebaceous differentiation. J Gastroenterol 2000;35:396-401.

4 Luchtrath H, Schriefers KH. Pankreaszyste under dem bild einer sogenannten branchiogenen zyste. Pathologe 1985;6:217-219.

5 Truong LD, Rangdaeng S, Jordan PH Jr. Lymphoepithelial cyst of the pancreas. Am J Surg Pathol 1987;11:899-903.

6 Strapko A, Botash RJ, Murthy UK, Landas SK. Lymphoepithelial cyst of the pancreas: a case report and review of the literature. Dig Dis Sci 1998;43:870-874.

7 Worrall NK, Drebin JA. Pancreaticoduodenectomy for lymphoepithelial cyst of the pancreas. Am Surg 2000;66:732-734.

8 Truong LD, Stewart MG, Hao H, Yutani C, Jordan PH. A comprehensive characterization of lymphoepithelial cyst associated with the pancreas. Am J Surg 1995;170:27-32.

9 Tateyama H, Tada T, Murase T, Fujitake S, Eimoto T. Lymphoepithelial cyst and epidermoid cyst of the accessory spleen in the pancreas. Mod Pathol 1998;11:1171-1177.

10 Bolis GB, Farabi R, Liberati F, Maccio T. Lymphoepithelial cyst of the pancreas. Report of a case diagnosed by fine needle aspiration biopsy. Acta Cytol 1998;42:384-386.

11 Liu J, Shin HJC, Rubenchik I, Lang E, Lahoti S, Staerkel GA. Cytologic features of lymphoepithelial cyst of the pancreas: two preoperatively diagnosed cases based on fine-needle aspiration. Diagn Cytopathol 1999;21:346-350.

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