Uncommon Lymphoepithelial Cyst with Sebaceous Glands of the Pancreas

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ABSTRACT

Context Lymphoepithelial cysts with sebaceous glands of the pancreas are extremely rare, with only 7 cases, including this case, published in English literature. Case report We herein present the case of a 67-year-old Asian man who underwent a resection of a lymphoepithelial cyst of the pancreas during the follow up care for lung cancer. Fourteen years previously he underwent a right lower lobectomy at the right segment nine for lung cancer. A 20 mm mass in the body of the pancreas was identified by CT scan 4 years ago, and the diagnosis was intraductal papillary mucinous neoplasm (IPMN) at that time. Over a 5-year period, this mass grew to 42 mm without dilatation of the main pancreatic duct. The preoperative evaluation, including endoscopic ultrasonographic guided fine-needle aspiration (EUS-FNA), indicated a cystic neoplasm with suspicion of malignancy. Intraoperative frozen section revealed a squamous-lined cyst accompanied by sebaceous glands without any malignant findings. Following this pathological finding, resection of the cyst was performed. Consequently, microscopic examination revealed that it was a lymphoepithelial cyst with sebaceous glands of the pancreas. Conclusions Pancreatic lymphoepithelial cysts can be cured by conservative resection, but if they are asymptomatic and are diagnosed before surgery, no treatment is necessary. To our knowledge, this is the first ever published case of a lymphoepithelial cyst with sebaceous glands of the pancreas, which was found during the follow up care for lung cancer.

INTRODUCTION

Squamous-lined cysts of the pancreas, which are classified as cystic pancreatic lesions, are divided into three types: lymphoepithelial cysts (LECs); epidermoid cysts in intrapancreatic accessory spleen; dermoid cysts [1]. Squamous-lined cysts of the pancreas are generally considered as benign lesions [2, 3, 4]. Etiology of LECs is uncertain, as it is difficult to obtain a correct diagnosis preoperatively, and a subsequent resection is often inevitably performed. Luchtranth and Schriefers [5] described LECs first in 1985, and fewer than 90 cases have been reported since then. From the first case of pancreatic LECs with sebaceous glands published by Fitko et al. in 1994 [6], this lesion represents a rare entity with 7 cases, including the present one, reported in the international articles [6, 7, 8, 9, 10, 11]. Here we present pancreatic LECs with sebaceous glands, in order to promote awareness of this rare entity and the appropriate surgical management.

CASE REPORT

A 67-year-old Asian man with a medical history of lung cancer (well differentiated adenocarcinoma stage I) 14 years previously was followed up at our outpatient clinic after a right lower lobectomy. His serum HIV antibody was negative and there was no other significant medical history. A 20 mm mass in the body of the pancreas, which was a small cystic lesion in 2002, was clearly identified by CT scan and MRI in 2008, 2012, and 2013 respectively (Figures 1 and 2). Subsequent diagnosis of a serous adenoma based on diagnostic imaging and EUS-FNA had been obtained 2 years previously. However, there was a growing tendency, from 20 mm to 42 mm, over the 5-year period, and consequently we decided to resect the lesion. A transition of carcinogenic antigen 19-9 (CA 19-9) during this follow up period
is illustrated in Figure 3. The intraoperative frozen section revealed a squamous-lined cyst accompanied by sebaceous glands without any malignant findings. Subsequently, resection of the cyst was performed. Chemical analysis of the cyst fluid was not performed due to an extremely small amount of fluid.

Macroscopically, the 4.2x2.4x2.2 cm surgical specimen, which weighed 9 g, demonstrated a polycystic lesion filled with soft cheesy material (Figure 4). Microscopic pathological findings revealed that the cysts contained sebaceous glands and were surrounded by squamous-lined epithelium, accompanied by infiltration of lymphocytes and plasma cells with a dense lymphoid follicle inside the cystic wall (Figure 3). These findings were consistent with LECs with sebaceous glands of the pancreas.

His clinical course was stable without complications, and he was discharged after 11 days of hospitalization.

**DISCUSSION**

All cases of squamous-lined cysts (LECs, epidermoid cysts in intrapancreatic accessory spleen, and dermoid cysts) are rare; roughly 90, 50, and 35 cases published in the literature respectively at this moment. To our knowledge, LECs with sebaceous glands of the pancreas are extremely rare with only 7 cases reported in the worldwide articles (Table 1). Interestingly, all cases are middle-aged males: namely, the mean age is 60.7±7.0 year-old. The mean size is 5.4±2.1 cm (range: 4.0-10.0 cm) which is slightly larger than the mean of LECs (4.7 cm) [1].
Intraoperative findings are generally consistent with the diagnostic imaging mentioned above. The cysts are separated from the pancreatic parenchyma with a fibrotic capsule which makes exfoliation of LECs from the pancreas easy. Therefore, it is reasonable not to perform unnecessary surgery such as a pancreaticoduodenectomy or distal pancreatectomy, etc. Given the fact that there is no report that refers to a recurrence of LECs, resection of LECs of the pancreas should be recommended as a surgical procedure at this time. However, if they are asymptomatic and diagnosed correctly before surgery, no treatment is necessary.

It has been pointed out that in one case, that could not obtain a normalization of CA 19-9 after resection, LEC was combined with a pancreatic cancer [15]. Generally, LECs can be considered as benign lesions, but it is clearly important to observe patients closely in order not to overlook a combination of malignant tumors. Furthermore, a simple cystic wall sometimes produces CA 19-9. Following this argument, a serum CA 19-9 level cannot conclude as to whether it is a malignant or benign lesion. Considering our case, the patient has a medical history of lung cancer. Currently, there is no clear correlation between LECs and lung cancer, because a 14-year time lag is significant. However, accumulation of LECs may find pathogenesis in the future.

He will continue to need observation, with particular attention paid to recurrence of LECs or a combination of malignant tumors.

In summary, to date, there are only 7 reports that refer to LECs with sebaceous glands of the pancreas. Although preoperative correct diagnosis may be difficult, characteristic intra-operative findings and frozen section can help in diagnosis of LECs which will prevent unnecessary surgery. When correct diagnosis is obtained preoperatively, pure laparoscopic resection of LECs may be recommended for symptomatic LECs. As a result of this report, we would like to make clinicians more aware of LECs of the pancreas. When recognized and managed appropriately, laparoscopic resection will lead to better surgical management and improved outcome in patients with LECs.
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References