Simultaneous Occurrence of a Rare Pancreatic Lymphoepithelial Cyst and Duodenal Mesenteric Castleman's Disease: a Case Report

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Abstract Pancreatic lymphoepithelial cyst is a rare pancreatic lesion of undetermined pathogenesis, which is a true pancreatic cyst. Castleman's disease is a rare lymphoproliferative disorder, and a mesenteric location is unusual. The simultaneous occurrence of the two diseases are rarer than metachronous ones and has not been reported to date. We present a case report of a patient with simultaneous occurrence of pancreatic lymphoepithelial cyst and duodenal mesenteric Castleman's disease.

Keywords Lymphoepithelial cyst · Pancreas · Castleman's disease · Mesenteric · Histopathological appearance

Introduction

Pancreatic lymphoepithelial cyst (LEC) is a rare pancreatic lesion of undetermined pathogenesis, which is a true pancreatic cyst lined by squamous epithelium and surrounded by mature lymphoid tissue [1]. Castleman's disease (CD) is a rare disorder that presents either as a localized disease or a systemic disease. It represents a morphologically distinct form of "angiofollicular lymph node hyperplasia" [2]. The goal of the study was to report on an interesting and extremely uncommon case of coexistence of the two rare lesions in adjacent anatomical sites.

Case Report

A 30-year-old woman was admitted with a pancreatic mass for further investigation. She had no past history of acute pancreatitis or gallstones, no weight loss, and no family history of tumors. Physical examination revealed mild tenderness on the upper quadrant abdomen. Tumor markers such as CEA and CA19-9 were normal. HIV testing was negative. Abdominal computed tomography (CT) showed an oval-shaped multilocular nonenhanced cyst in the head of the pancreas, and the peripancreatic region has a homogeneous soft tissue mass which was a significant enhancement (Fig. 1). A pancreaticoduodenectomy was performed. Histologically, the cyst of the head of the pancreas demonstrated that the cyst wall was lined with stratified squamous epithelium, which was surrounded by the lymphocytic infiltrate (Fig. 2a). Duodenal mesenteric mass revealed an enlarged lymph node composed of numerous small involuted follicular centers and central hyaline-containing sclerotic vessels surrounded by concentric layers of small lymphocytes (Fig. 2b). A diagnosis of simultaneous occurrence of pancreatic LEC and duodenal mesenteric Castleman's disease (hyaline vascular type) was determined.

Her postoperative course was uneventful, and she was discharged 11 days after surgery. Neither symptoms nor recurrence was found during 1-year follow-up.

Discussion

Cystic lesions of the pancreas can be divided into true cysts, pseudocysts, and cystic neoplasms. LEC of the pancreas is a rare true cyst. It is a slow-growing lesion

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Fig. 1 CT showed a nonenhanced cyst and a significant enhancement soft tissue mass

in the pancreas and was first reported by Luchtrath [3]. The histogenesis of LEC is unknown. A proposed mechanism of pathogenesis for LEC is the development from epithelial remnants in lymph nodes [1]. The histopathological appearance of LEC is characterized by cysts lined by stratified squamous epithelium, which is surrounded by the adjacent dense subepithelial lymphoid tissue with abundant lymphoid follicles [4] and also was demonstrated in our case.

Castleman's disease was first described in 1954 and is a rare disorder of the lymphoid tissue [5]. Although the etiology is unknown, there were observed association of Castleman's disease with HHV-8 infection and overproduction of interleukin (IL)-6 [6]. The histological findings can be divided into hyaline vascular type, plasma cell type, and mixed type. The hyaline vascular type, which is the most common type (90 %), is

characterized by an enlarged lymph node composed of numerous small involuted follicular centers and central hyaline-containing sclerotic vessels surrounded by concentric layers of small lymphocytes [7]. Our case belongs to this variant.

This case showed pancreatic LEC occurring simultaneously with duodenal mesenteric Castleman's disease. We believe that such coexistence must not have the same etiology, but rather a very particular one.

Conclusion

Simultaneous occurrence of pancreatic LEC and duodenal mesenteric Castleman's disease is an extremely rare benign lesion and lacks specific symptoms or signs. Pathologic

Fig. 2 Histopathological appearance of pancreatic LEC (a) and duodenal mesenteric Castleman's disease (b)

examination is essential for diagnosis, and surgical treatment has a good prognosis.

Conflict of Interest The authors declare that they have no competing interests.

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