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Pathology

LYMPHOEPITHELIAL CYST OF PANCREAS: A RARE ENTITY

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ABSTRACT Pancreatic cystic lesions can be neoplastic or non-neoplastic in nature. Majority of these especially those in the body or tail or pancreas are asymptomatic and detected incidentally on routine imaging. Pancreatic lymphoepithelial cyst [LEC] is an extremely rare, benign cystic lesion that is difficult to differentiate preoperatively from other cystic pancreatic lesions. We report a case of LEC of the pancreas in an elderly male who had a synchronous carcinoma of the Stomach. Imaging showed a cystic lesion in the tail of pancreas extending to the splenic hilum. He underwenta radical gastrectomy with distal pancreatosplenectomy. The histopathological examination showed cyst lined by benign keratinised stratified squamous epithelium atop dense lymphoid tissue.

KEYWORDS : Lymphoepithelial cyst, pancreatic cyst, benign

INTRODUCTION:

Lymphoepithelial cysts (LECs) are rare, benign cysts that include ~0.5% of all the pancreatic cysts and occur most frequently in middle-aged men [1, 2].

They were first described by *Lüchtrath* and Schriefers in 1985 in a cyst that seemed histologically like a branchiogenic cyst [3, 4].

Pseudocysts are the most common cystic lesions of the pancreas and frequently occur as a complicationof pancreatitis [5]. True cysts of pancreas, in contrast, are most often neoplastic [6]. Macroscopically, LEC is a true cyst filled with keratinous material and histologically, the wall of the cyst is lined by keratinizing squamous epithelium with granular layer, and surrounded by lymphoid tissue. The size of LECs ranges from 1 to 15 cm, with an average size of 4.5–6 cm, and can arise in any part of the pancreas [1,2, 7, 8].

Approximately half of the patients present incidentally, with the remaining patients being associated with non-specific symptoms such as nausea, diarrhoea, abdominal pain, weight loss, and fatigue [9].

They differ from other pancreatic cysts in their malignant potential and prognosis. Therefore, correct diagnosis of pancreatic cystic lesions is very significant, and physicians must be aware of the different types of cystic lesions and their characteristics before the choice of treatment [1,10]. Herein, we report this rare entity of pancreatic LEC.

Case Report:

A CT scan of the abdomen incidentally showed a 5.5 x 5 cm distal pancreatic cyst in a 70 year old male who was evaluated for increased urinary frequency. In addition, the CT scan showed enlarged perigastric lymphnodes. An Endoscopic Ultrasound guided FNAC from pancreatic tail cystic lesion showed abundant necrotic debris and anucleated squames while parigastric lymph node was reported as metastatic adenocarcinoma. An upper GI endoscopy and biopsy confirmed an adenocarcinoma of stomach. An interim PET-CT evaluation following neoadjuvant chemotherapy showed a relatively increase in the size of the cyst to 8 cms (Figure 1). The tumor markers viz serum CA19.9 (53 U/mL) and Serum CEA level (7.27 ng/mL) were mildly elevated. The patient subsequently underwent a radical gastrectomy with a distal pancreatosplenectomy. Grossly, a large smooth surfaced rounded cyst measuring 9.5 x 7.5 x 6cm was seen in the tail of pancreas, bulging into the peripancreatic tissue and the splenic hilum. On cutting open, a uniloculated cyst with few fine ridges was seen and filled with yellowish turbid fluid along with whitish flakes. The wall thickness ranges from 1-3mm. The fluid and whitish flakes was collected in a container and the cytospin slides were prepared. The cytospin smears showed squames, keratin and necrotic debris(Figure 2). On histology, the cyst was lined by keratinised stratified squamous epithelium with granular layer, with orderly maturation and no atypia. The subepithelium showed a thick band ofsmall lymphocytes including lymphoid follicles with germinal centres, (Figure 3). The rest of the pancreatic parenchyma was unremarkable.

DISCUSSION:

PLECs are rare and benign lesions comprising about 0.5% of all pancreatic cystic lesions. They occur in the 5th and 6th decades of life with male predominance. The body or tail of the pancreas is where two-thirds of the cysts arise, while one-third originates from the head. [9] Most of these PLECs are asymptomatic until they grow to large sizes. Abdominal pain and discomfort are the most common symptoms [1].

The exact etiology of LECs is unknown. There are numerous etiological theories that have been considered, including the formation of pancreatic ducts from squamous metaplasia, derivation from epithelial remnants in lymph nodes, displacement of branchial cysts that fuse with the pancreas during embryogenesis, or a form of teratoma [1].

LECs are usually singlespherical well demarcated cystsin the pancreas with well-defined walls [1]. Cases can be peripancreatic and multilocular also [1]. The content of cysts is often 'cheesy' or 'caseous', which suggests keratinaceous debris. [1].

Microscopically, the cyst wall is lined by mature stratified squamous epithelium, and the subepithelium shows dense lymphoid tissue and lymphoid follicles. The lymphoid cells are a mixture of mature T lymphocytes, with B lymphocytes which forms the germinal centres. Rarely, the cyst may show sebaceous differentiation or mucinous differentiation [1]. If the sebaceous and mucinous components are present in a dominant manner,then the diagnosis is a dermoid cyst [1].

There has been no documented recurrences or malignant transformation of pancreatic LECs, suggesting thatthe cyst is an entirely benign lesion [9].Despite this, due to the difficulty indistinguishing preoperatively from potentially malignant forms of pancreatic cyst neoplasm, such as mucinous cyst neoplasms (MCNs) or intraductal papillary mucinous neoplasmsmany cases are ultimately treated with surgical resection[9].

Preoperative investigations include CT or magnetic resonance imaging, EUS and fine needle aspiration (FNA), and cyst fluid analysis. Radiology imaging may aid to guide a diagnosis, but LECs cannot be reliably distinguished from other cystic pancreatic lesions on imaging alone as they appear similar to a pseudocyst or MCN [9].

On EUS-FNA, the presence of squamous cells and lymphocytes is diagnostic of LEC [8].

LECs have also been shown to express significantly high levels of CEA as well as carbohydrate antigen 19-9, and thus, cyst fluid analysis is not a reliable test to distinguish LECs from malignant cystic lesions [9].

For the diagnosis of pancreatic LECs, surgical excision and histopathological examination still remain the gold standard[5, 6, 7, 8]. For the larger lesions in the head of the pancreas or those encroaching on the spleen, surgical resection by pancreaticoduodenectomy or distal pancreatectomy may be required [4, 7]. If the lesions are small and well-defined, cyst enucleation or drainage may be preferable.[4].

CONCLUSION:

In conclusion, lymphoepithelial cyst of pancreas is a rare and benign pancreatic cystic lesion. On imaging, this entity is difficult to differentiate from other malignant pancreatic cystic lesions. Complete excision and histopathological examination remain the mainstay for diagnosis of LECs.

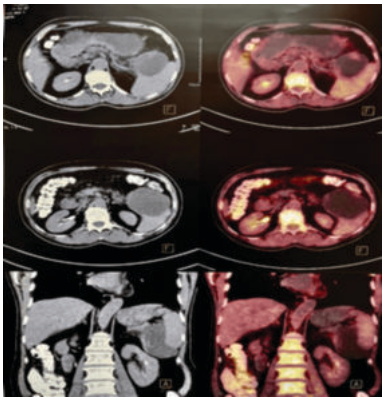
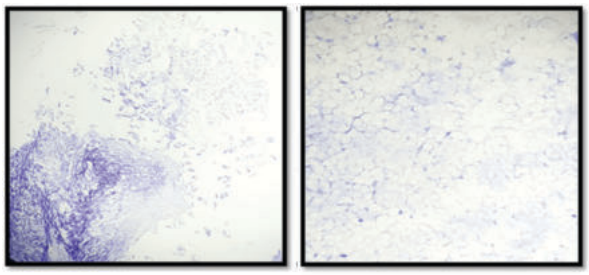


Figure 1: Comparative image of the distal pancreatic cyst on CT and PET images with no appreciable FDG avidity at the pancreatic cyst.



A **B**

Figure 2 :A- The cytospin smear shows keratin and necrotic debris (H&E stain 400x)
B - The cytospin smear shows squames (H&E stain 400x)

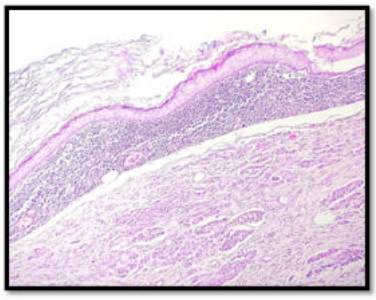


Figure 3: The cyst lined by keratinised stratified squamous epithelium with orderly maturation and no atypia seen. The subepithelium showed a thick band of small lymphocytes and unremarkable pancreatic parenchyma (H&E stain 100x)

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