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LYMPHOEPITHELIAL CYST- A RARE NON-MALIGNANT CYSTIC LESION OF PANCREAS

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Abstract :
Lymphoepithelial cysts are rare pancreatic lesions of undetermined pathogenesis. The literature on this entity is limited to case reports or small series. We describe a case of 33 year female, with incisional hernia incidentally diagnosed to have cystic lesion in tail of the pancreas that was managed by distal pancreatectomy. An extensive Medline search was carried out for lymphoepithelial cyst of pancreas. Till date less than 100 cases were identified in available literature. This entity has uniform and distinctive clinicopathological features. About half of the reported cases were asymptomatic with most of the lesions diagnosed incidentally. Majority of patients presents with non-specific symptoms making preoperative diagnosis difficult. Lymphoepithelial cyst of the pancreas is a rare benign lesion, which is difficult to diagnose preoperatively. High index of suspicion and preoperative fine needle aspiration cytology may help in making diagnosis and avoiding surgery in asymptomatic patients.

Keyword : Lymphoepithelial cyst (LEC), Cystic lesion of Pancreas, True pancreatic cyst, Distal Pancreatectomy

Introduction:

Cystic lesions of the pancreas are relatively rare [1-3]. In adults, 85%–90% of these lesions are pseudocysts [2, 3]. True cysts of the pancreas, characterized by an epithelial lining, are uncommon cystic pancreatic lesions. Although they constitute a challenging differential diagnosis at the clinical, radiological, and pathologic levels, all patients with pancreatic cystic lesions, whether asymptomatic or symptomatic, must be thoroughly investigated to ascertain the underlying nature of the cyst [4]. Lymphoepithelial cyst (LEC) of the pancreas is an exceedingly rare nonneoplastic entity of uncertain histogenesis. LEC are true pancreatic cysts lined by

squamous epithelium and surrounded by matureymphoid tissue. The cyst arises typically in middle-aged men, and is usually asymptomatic or causes nonspecific abdominal symptoms. There is no specific serologic marker for this entity. None of its radiological characteristics can help differentiate it from other cystic lesions of the pancreas. Fine-needle aspiration cytology may be able to suggest its benign nature. The outcome after surgical excision is uniformly good with good symptom control and no recurrences.Successful management of LEC depends on ability to differentiate them from other cystic neoplasm of pancreas.

Case Report:

A 33 year old female presented with incisional hernia on evaluation incidentally diagnosed to have cystic lesion in tail of the pancreas. There was no history of pain in abdomen, vomiting, hematemesis or malena. Patient was non-smoker & alcoholic. No associated co morbidities. Her physical examination was unremarkable. Her serum Amylase, CEA, CA 19-9 were normal and Ultrasonography abdomen, Contrast enhanced computed tomography abdomen (Fig. 1a & b) were done, found a lesion of 5cm round, well-circumscribed, cystic mass with mild wall enhancement in tail of pancreas. Provisional diagnosis of cystic lesion of pancreas was made and patient was planned for distal Pancreatectomy. The patient underwent exploratory laparotomy and on exploration, a 5cm round, well-circumscribed, cystic mass found in tail of pancreas (Fig. 2a). Distal Pancreatectomy was performed (Fig. 2b).Postoperative course was uneventful; she was started on oral feed after 72 hours and was discharged on 7th postoperative day. Patient is asymptomatic on regular follow-up with CECT and CA19-9 levels. Her postoperative CA 19-9 level is normal. Microscopic evaluation from resected specimen cyst wall lined by squamous epithelium. The subepithelium showed abundant lymphocytes with germinal centre formation. Keratinization was also noted (Fig. 3a & b). Diagnosis of LEC was confirmed.

Discussion:

LEC of the pancreas is rare true pancreatic cyst, lined by stratified squamous epithelium. In this review, we searched all the cases of lymphoepithelial cyst of the pancreas published in the literature, from the first case reported by Luchtrath et al [5] in 1985and characteristics of LEC become apparent, they are seen in middle-aged patients (mean age, 56 years; range, 20–82 years) predominantly in men (M:F, 4:1). The most common symptom at presentation is abdominal pain. Other complaints at presentation include anorexia, weight loss, vomiting, back pain. Many cases were diagnosed incidentally during work-up for other diseases. LEC are often rounded and have a well-defined wall that sharply demarcates it from the pancreas and surrounding adipose tissue. The average size of LEC is 4.5 cm (range, 1–12 cm). They can be multilocular or unilocular. These lesions seem to be equally distributed in head, body and tail region of the pancreas.**Pathogenesis of LEC** The histogenesis of LEC is unknown. A proposed mechanism of pathogenesis for LEC is the development from epithelial remnants in lymph nodes. The histological characteristics of LEC are unique, and were first described by Lüchtrath and Schiefers [5], who noted the microscopic similarity of the cyst to the branchial cleft cysts of the lateral neck. Microscopically, the LEC are characterized by cysts lined by stratified squamous epithelium and immediately adjacent dense

subepithelial lymphoid tissue that contains lymphoid follicles. The lesion is separated from the pancreatic parenchyma by a capsule of thin fibrotic tissue.

Differential Diagnosis

The clinical differentiation of LEC from other cystic lesions of the pancreas can be challenging. LEC have a macrocystic appearance and are thus clearly distinguished from microcystic lesions such as serous microcystic adenoma. Their distinction from macrocystic lesions at clinical level could be problematic. Chemical analysis of aspirated cyst fluid has proved to be useful in the differential diagnosis of pancreatic cysts in general, the aspirated fluid from LEC has squamous epithelial lining (to differ from pseudocyst) which is rich in lymphoid cells (to differ from mucinous & malignant cystic lesions) & sometimes it may also contain "cheesy" or "caseous" appearance characteristic of keratinaceous debris [11].

The traditional markers such as CEA, CA19-9, CA-125, and fluid viscosity would be expected to be significantly lower in LEC than in mucinous neoplasms [8, 9]. Only few cases presented with elevated serum levels of CA 19-9 [7, 9, 10].

Role of EUS +/FNAC in Lymphoepithelial Cyst of Pancreas:

The main issue in the treatment of this benign lesion is its differentiation from other cystic lesions of the pancreas, most importantly pseudocysts and cystic neoplasm [11]. Since the treatment options and the prognosis of these entities is different. In these situations, fine needle aspiration (FNA) of the lesion may be able to suggest the benign nature of the lesion and as a true cyst of the pancreas [11]. Cytological material obtained from LEC reveals nucleated or anucleated squamous epithelial cells, occasional histiocytes and rare lymphocytes,without evidence of neoplastic cells. Recently endoscopic ultrasound (EUS) and EUS guided FNA plays important role in the evaluation of cystic lesions of the pancreas and in diagnosis of LEC preoperatively, and thus avoiding unnecessary surgery in patients [11, 12, 13]

The presence of squamous material and lymphocytes on cytologic examination after EUS guided FNAC is diagnostic of LEC. Aspirate CEA level may be elevated and should be considered in conjunction with cytologic results to avoid misdiagnosis as a mucinous cystic neoplasm. In radiologically benign appearing lesions, EUS+FNA confirmation of a negative cytology and low fluid CEA can further provide evidence to support a monitoring approach and deferral of surgical intervention [11]. The classical finding of LEC on EUS is hypoechoic uniloculated or multiloculated cystic lesion. Occasionally, fine or coarse sludge like hyperechoic echo architecture is also seen likely due to debris within the cyst [13].

Management Options:

Surgical management of the LEC has been variably described in the literature from conservative and regular follow-up in asymptomatic patients to classical Whipple's procedure (pancreatoduodenctomy) in few patients. No recurrences or progression into lymphoma or carcinoma have been documented in the cases of LEC in which follow-up information was available. Thus, if the tumor can be diagnosed preoperatively, the option of "wait and watch" may be clinically acceptable. However, in most cases, the possibility of another type of pancreatic cystic neoplasm is difficult to rule out with the current investigative methods. After reviewing the available literature we suggest following recommendations regarding management of lymphoepithelial cyst of the pancreas:

1. Preoperative FNA or EUS and FNA should strongly be considered in a high surgical risk and asymptomatic patient.

2 If the FNA establishes the diagnosis of a LEC, the operation can be avoided and the patient may be followed with serial imaging.

3 In the symptomatic patients with acceptable surgical-risk, an exploration of the upper abdomen should be undertaken.

4 A frozen section biopsy of the cystic mass should be obtained during the operation. If the microscopic evaluation verifies the diagnosis of a LEC, a simple cyst enucleation should be sufficient treatment.

5 In symptomatic patients, if lesion is situated in body or tail of pancreas, then simple distal pancreatectomy with splenic preservation should be performed, only if simple enucleation of the cyst is not feasible.

6 In the cases where cyst involved the head of the pancreas, and either involving or compressing duodenum or the common bile duct, enucleation or drainage procedure rather than resection procedure (Whipple's or pylorus preserving pancreatoduodenctomy) should be performed.

Conclusion

True pancreatic cysts lined by stratified squamous epithelium are rare. We present a case of lymphoepithelial cyst and review of the present literature. These lesions, despite their rarity, must always be kept in the differential diagnosis of a cystic pancreatic lesion. It requires a high degree of suspicion and fine needle cytology (EUS +/FNA) in all patients to diagnose this rare entity. All patients with pancreatic cystic lesions, whether asymptomatic or symptomatic, must be thoroughly investigated to ascertain the underlying nature of the cyst. Regular follow-up is all that is required in asymptomatic patients with proven diagnosis. Enucleation of the cyst avoids the unnecessary resection in these patients. However due to uncommon nature and difficulty in radiological diagnosis most cases will continue to be identified on pathological examination after resection.

References:

1. Brugge WR, Lauwers GY, Sahani D, Fernandez del Castillo C, Warshaw AL. Cystic neoplasms of the pancreas. N Engl J Med. 2004;351:1218–1226.

2. Kloppel G, Solcia E, Longnecker DS, Capella C, Sobin LH (1996) Histologic typing of tumors of the exocrine pancreas. In: World Health Organization—international histologic classification of tumors. Vol 2. Geneva, Switzerland.

3. Adsay NV, Klimstra DS, Compton CC. Cystic lesions of the pancreas. Semin Diagn Pathol. 2000; 17:1–6.

4 Gracea G, Ong SL, Rajesh A, Neal CP, Pollard CA, Berry DP, et al. Cystic lesion of the pancreas. A diagnostic & management dilemma. Pancreatology. 2008;8(3):236–251.

5 Luchtrath H, Schriefers KH. A pancreatic cyst with features of a so-called branchiogenic cyst. Pathology. 1985;6:217–219.

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

7 Barbaros U, Erbil Y, Kapran Y, Bozbora A, Ozarmagan S, Bilgic B. Lymphoepithelial cyst: a rare cystic tumor of the pancreas which mimics carcinoma. JOP. 2004;5:392–394.

8 Younus S, Bleibel W, Bleibel H, Hernady N. Lymphoepithelial cyst of the pancreas. Dig Dis Sci. 2007;52 (11):3136–3139.

9 Idetsu A, Ojima H, Saito K, Hirayama I, Hosouchio Y, Nishida Y, et al. Lymphoepithelial cyst of the pancreas: report of a case. Surg Today. 2008;348(1):68–71.

10 Yamaguchi T, Takahashi H, Kagawa R, Takeda R, Sakata S, Yamamoto M, et al. Lymphoepithelial cyst of the pancreas associated with elevated CA 19-9 levels. J Hepatobiliary Pancreat Surg. 2008;15 (6):652–654.

11.Scheiman JM. Management of cystic lesions of the pancreas. J Gastroenterol Surg. 2008;12(3):405 –407. doi: 10.1007/s11605-007-0350-5.

12 Attasaranya S, Pais S, LeBlanc J, McHenry L, Sherman S, DeWitt JM. Endoscopic ultrasound-guided fine needle aspiration and cyst fluid analysis for pancreatic cysts. JOP. 2007;8(5):553–563.

13 Ahlawat SK. Lymphoepithelial cyst of the pancreas: role of endoscopic ultrasound guided fine needle aspiration. JOP. 2008;9(2):230–234.

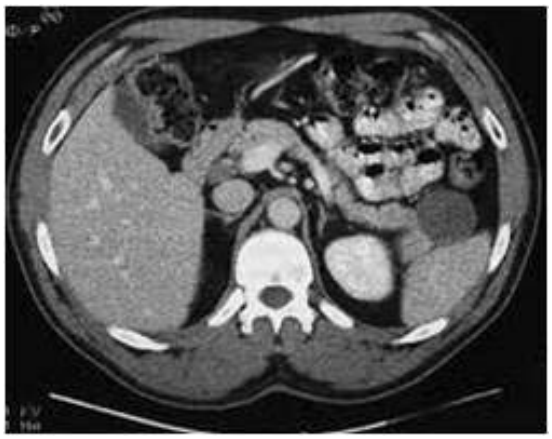
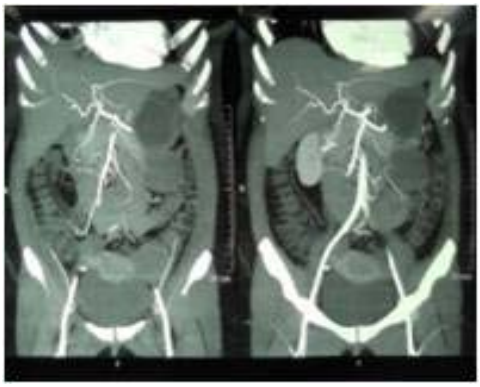
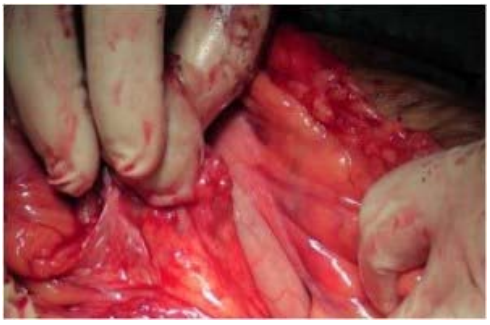


Fig.1a Fig.1b





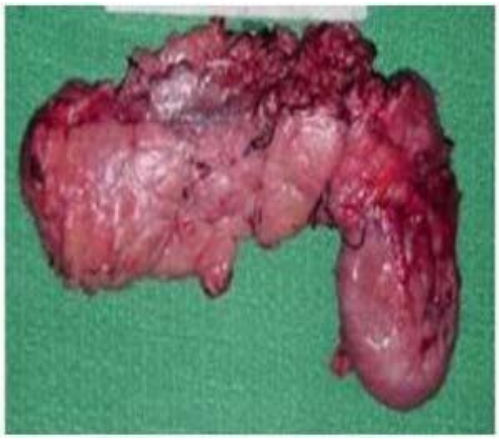
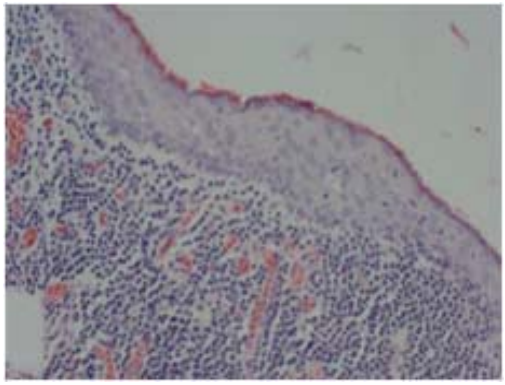


Fig.2a

Fig.2b Fig.3a



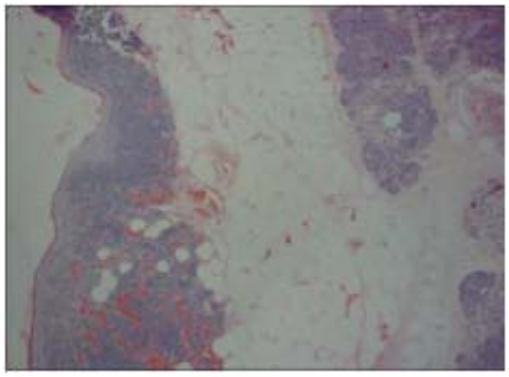


Fig.3b