



# Large lymphoepithelial cyst of pancreas: a rare entity managed using lumen-apposing metal stent—case report and review of the literature

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Received: 22 March 2019 / Accepted: 8 April 2019 / Published online: 12 April 2019  
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## Abstract

Lymphoepithelial cysts (LECs) are extremely rare nonmalignant cysts of the pancreas. Asymptomatic LECs are managed conservatively, but symptomatic LECs have traditionally been managed with surgical resection. We report the first case of symptomatic infected LEC of the pancreas successfully managed with EUS guided endoscopic drainage with lumen-apposing metal stent. We also review the relevant literature and discuss the diagnosis and management of this rare cyst of the pancreas.

**Keywords** Lymphoepithelial cyst of pancreas · Pancreas cyst · Lymphoepithelial cyst · Lumen-apposing metal stent

## Introduction

Lymphoepithelial cysts (LECs) are extremely rare nonmalignant cysts of the pancreas [1]. Diagnosis of these cysts is difficult, but it is important to differentiate them from malignant cysts. Endoscopic ultrasound (EUS) and fine needle aspiration (FNA) along with cyst fluid analysis has been used to help to guide diagnosis of a benign vs. malignant cyst. The cytological yield of EUS FNA of pancreas cyst is low [2], and an accurate determination of LEC is generally made after the surgical excision of these cysts. Majority of LECs are detected incidentally on imaging, but they can present with symptoms such as abdominal pain, nausea, vomiting, fever or weight loss [3]. We report the first case of a large infected LEC of pancreas diagnosed and managed with the placement of EUS-guided lumen-apposing metal stent (LAMS).

## Case report

A 61-year-old male patient presented to the hospital with severe upper abdominal pain and fever. Past medical history was significant for hypertension, hyperlipidemia, and GERD. He had a history of tobacco use but denied any alcohol use. He had previous episodes of pancreatitis. On examination, the patient was febrile, tachycardic and tender to palpate in the epigastrium without any rebound or rigidity. Laboratory examination revealed mildly elevated white cell count (12,000/ $\mu$ L), otherwise complete blood count, basic metabolic profile, liver function tests, lipase, and amylase were normal. CT scan of the abdomen showed 11.1  $\times$  10.5  $\times$  8.8 cm cystic lesion in the body of the pancreas with multiple foci of internal gas suspicious for an infected pseudocyst (Fig. 1).

EUS revealed an anechoic lesion suggestive of a cyst in the body of the pancreas measuring about 11 cm  $\times$  10 cm in maximal diameter. One pass was made into the cyst with a 19-gauge fine needle using a transgastric approach. Approximately 5 mL of cheesy, white material was aspirated suspicious for a lymphoepithelial cyst or a pseudocyst. No mucin was seen, therefore, the suspicion for a mucinous cystic neoplasm was low. Pancreatic duct was normal in caliber, and no pancreatic duct connection was identified; hence the possibility of the main duct IPMN was low. Moreover, no suspicious solid lesion was identified.

Because of patient abdominal pain and signs of infection a decision was made to drain the cyst endoscopically. Cyst gastrostomy was then performed with placement of

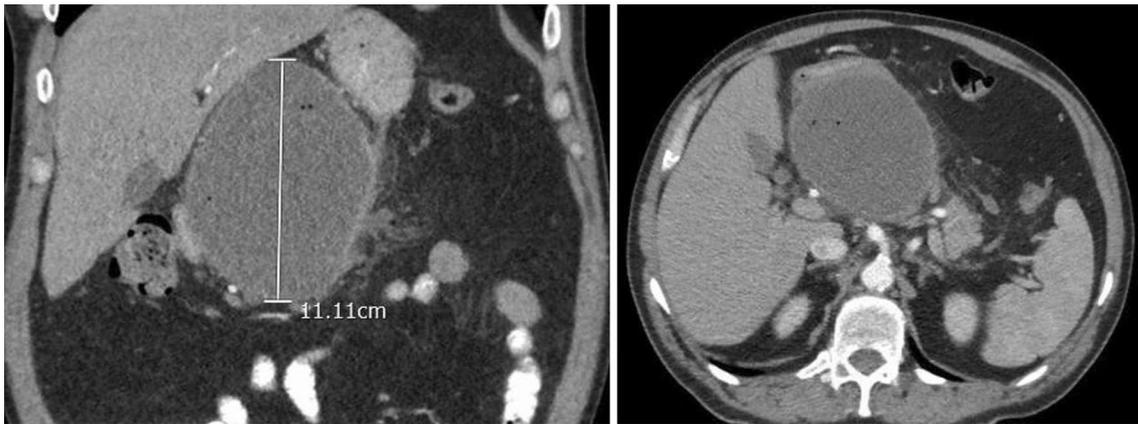
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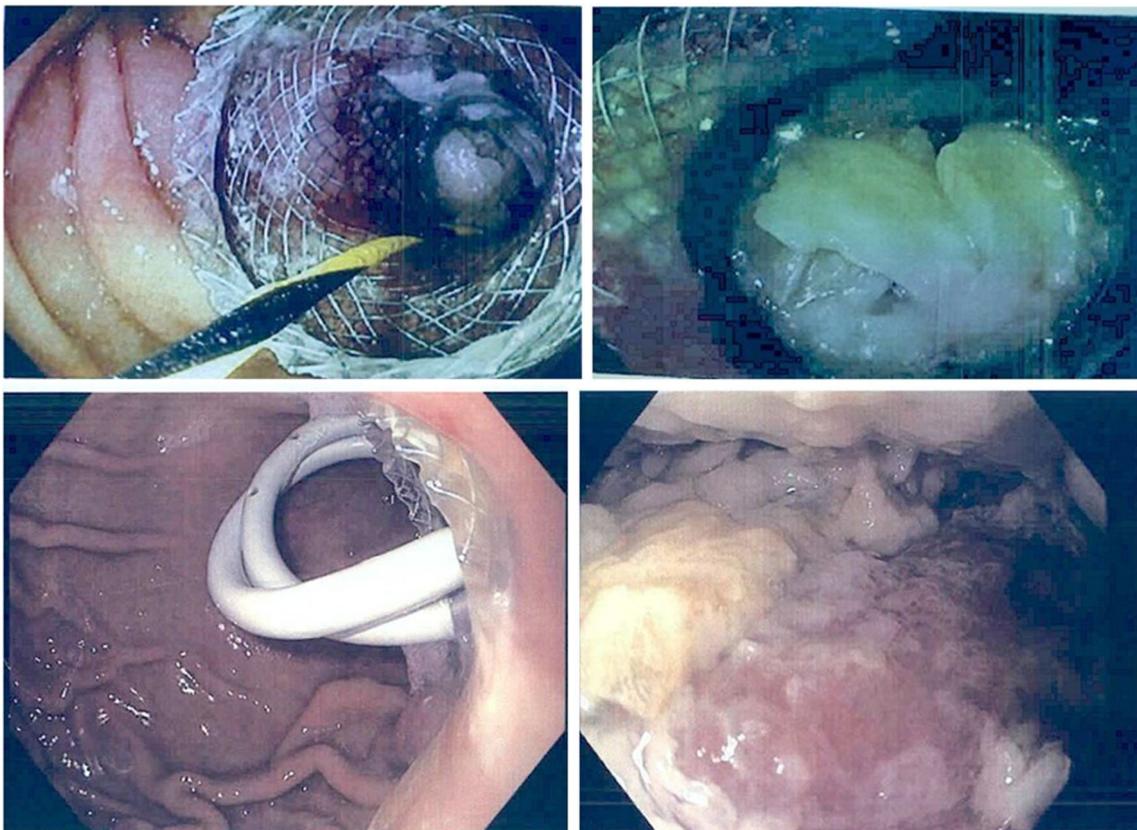


**Fig. 1** CT scan showing a cyst in the body of the pancreas with multiple foci of internal gas

a LAMS (Hot AXIOS, Boston Scientific) of dimensions 15 mm (diameter) × 10 mm (length) under EUS guidance. LAMS lumen was dilated to 15 mm, and then endoscope was advanced in the cyst cavity (Fig. 2). Cyst cavity was lavage, irrigated and cleaned. Two pigtail stents were placed across the LAMS into the cyst cavity. Cultures of cyst aspirate were positive for *Streptococcus anginosus*; the patient

was treated with amoxicillin for 7 days. Post-cyst-gastrostomy patient did well, sepsis resolved, and the patient was discharged home.

EUS-FNA cytology was inconclusive, showed benign inflammatory cells but no mucin. However, the cyst fluid CEA levels were elevated to 12,000 with low Amylase (< 100). One week later an upper GI endoscopy was



**Fig. 2** Endoscopic drainage with cyst gastrostomy with a lumen-apposing metal stent (LAMS)

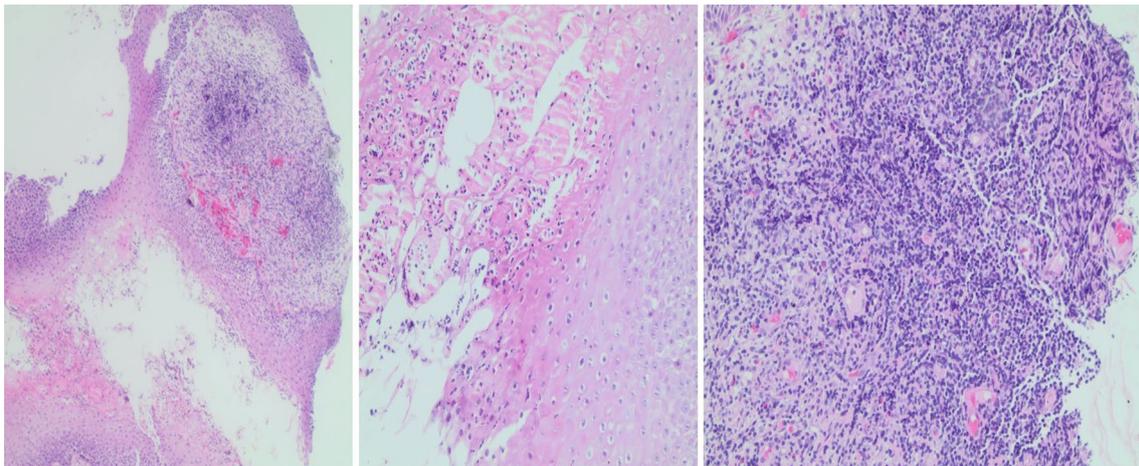
then performed; pigtail stents were removed, and the endoscope was advanced into the cyst cavity through the LAMS. Direct cyst biopsies were obtained using a standard radial jaw biopsy forceps (2.2 mm outer diameter). Overall three passes were made with the biopsy forceps, and adequate tissue was obtained. LAMS was then removed at the end of the procedure. Biopsies showed stratified squamous epithelium lining with an underlying prominent lymphocytic infiltrate containing mainly mature appearing lymphocytes as well as reactive spindle cells; these findings were consistent with a lymphoepithelial cyst (Fig. 3).

At 8 months follow remained asymptomatic without any abdominal pain. MRI done showed a decrease in the size of the cyst to 4 cm (Fig. 4).

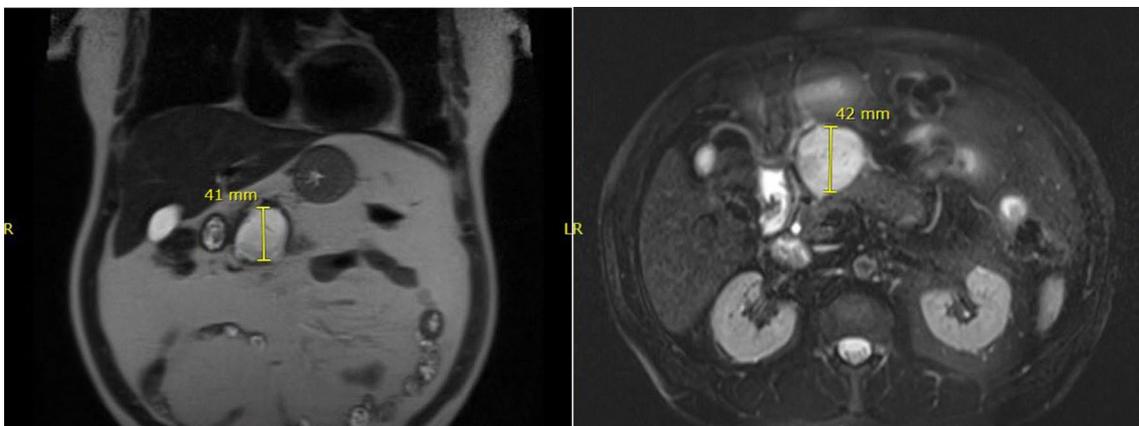
## Discussion

Pancreatic cysts are being increasingly identified because of the increased use of cross-sectional imaging [4]. The incidence of pancreatic cysts in the US population is estimated to be between 3 and 15% [5]. Pancreas cysts have a broad differential diagnosis. In general, they can be categorized into non-neoplastic (e.g., pseudocysts) and neoplastic cystic lesions. Cystic neoplasms of the pancreas can be broadly classified into mucinous (that produce a mucin-rich fluid) and non-mucinous (that do not produce mucin). It is important to differentiate the mucinous pancreas cyst [mucinous cystic neoplasm (MCN), intraductal papillary mucinous neoplasm (IPMN)] from the non-mucinous cysts like serous cystadenoma.

LECs of the pancreas is an uncommon type of benign pancreatic cysts that account for approximately 0.5% of



**Fig. 3** Histopathological examination of pancreatic cyst biopsies demonstrating squamous epithelium and underlying lymphoid tissue



**Fig. 4** Follow up MRI at 8 months showed significant decrease in the size of the cyst from 11 to 4 cm

all pancreatic cysts [6]. Pathogenesis of pancreatic LECs is unknown, but some hypothesize that they form from the squamous metaplasia of the pancreatic ducts, epithelial remnants in lymph nodes, displacement of branchial cysts that go on to fuse with the pancreas during embryogenesis, or the possibility that LECs are a form of teratoma [1, 6].

A total of 236 cases (including our case) of LECs of the pancreas have been reported in the literature so far [7–11]. Table 1 summarizes the characteristics of LECs described in the literature. LECs are more common in males than females, and the mean age of diagnosis is 55 years. LECs are mostly solitary, multiloculated and located in the tail/body of the pancreas [11]. Only half of the patients developed

symptoms related to LECs; abdominal pain was the most common presenting symptom. Our patient presented with abdominal pain and fever with signs of infection of the large LECs which is extremely rare.

EUS-FNA can help in accurate diagnosis, but FNA is inconclusive in most cases because of the acellular specimen. The overall yield of EUS or CT guided FNA or biopsy was approximately 63% [11]. Macroscopic examination of the aspirate may reveal cheesy, white, greasy porridge-like material with keratin debris and cholesterol or fat deposits, occasionally clear/ straw yellow color aspirate can also be seen. Microscopically they are characterized stratified squamous epithelium lining with an adjacent subepithelial layer

**Table 1** Summary of characteristics of LEC of the pancreas described in literature ( $N=236$ )

Characteristic	
Male:female	4:1
Mean age (years)	55
Presentation	
Symptoms on presentation	50%
Abdominal pain	46%
Weight loss	7%
Nausea and vomiting	8%
Fever	3.5%
Jaundice	2%
No of cyst	
Solitary	98%
Location	
Head/uncinate process	28%
Body	29%
Tail	43%
Mean size (mm)	47
Locularity of cyst	
Multilocular	49%
Bilocular	2%
Unilocular	44%
Pre-operative EUS-FNA	44%
Diagnostic yield of EUS guided FNA or biopsy	63%
Aspirate characteristics	
Macroscopic exam	Clear or straw yellow or cheesy white color porridge-like material with keratin debris and cholesterol or fat deposition
CEA levels	Elevated (> 200 ng/mL)
Amylase	Low
Microscopic	Stratified squamous epithelium with an adjacent subepithelial layer of lymphoid tissue containing lymphoid follicles
Management	
Distal pancreas resection/enucleation	80%
Head of pancreas resection or whipple	8%
Conservative	12%

of lymphoid tissue containing lymphoid follicles. Fluid amylase levels are low, but CEA levels are the most commonly elevated (> 200 ng/mL) likely due to the goblet cells or the aberrant immunoreactive squamous epithelial lining [1, 11]. Therefore, LECs can be confused with an MCN.

In our patient EUS-FNA fluid was cheesy, white, cream-colored material suspicious for LEC but the cytology was inconclusive. The cyst fluid CEA levels were elevated to 12,000 with low Amylase (< 100) similar to an MCN. Direct visualization of the cyst cavity was performed by introducing a regular endoscope through the LAMS into the cyst cavity. Cyst cavity examination revealed cheesy white colored cyst contents; direct cold forceps biopsies confirmed the diagnosis of LEC of the pancreas. We suggest EUS guided fine needle biopsy (FNB) of the cyst in cases where FNA is inconclusive to increase the cytological yield of the specimen. Direct examination of cyst cavity and biopsies using LAMS can be considered in cases of symptomatic large pancreas cyst/pseudocysts when LAMS is used for drainage and diagnosis remains unclear.

An accurate diagnosis of LECS is essential as their management and prognosis are different as compared to premalignant or malignant mucinous cysts. LECS of the pancreas are benign lesions, and malignant progression has not been reported [1]. They should ideally be managed conservatively, but most patients underwent surgical resection because of the lack of accurate preoperative diagnosis. If LECs can be accurately diagnosed preoperatively, they can be managed conservatively, and surgery can be avoided especially in asymptomatic patients.

Surgical resection can be considered for symptomatic LECs or if the diagnosis remains uncertain and the suspicion of malignancy is high. Surgical management depends mainly on the size, location and the degree of pancreatic and surrounding tissue involvement and included pancreaticoduodenectomy, distal pancreatectomy or cyst enucleation/excision. Alternatively, drainage can be considered for a well-delineated thick wall symptomatic LEC after diagnosis is confirmed as it would be suboptimal management for other mucinous cystic neoplasms [6].

Minimal invasive options such as EUS guided endoscopic drainage can be also considered after confirmation of diagnosis. LAMS has been recently developed for endoscopic management of pancreatic fluid collections, walled-off necrosis, and pseudocysts. LAMS is now also being used to drain benign collections like post-operative fluid collections, gall bladder for cholecystitis, etc. [12, 13]. LAMS has been shown to have a high technical and clinical success rate with low adverse events for mature collections that are favorably located adjacent to the stomach, duodenum [13]. We suggest LAMS can be used well-delineated thick wall symptomatic LEC with caution. We also placed two double pigtail stents in the LAMS. A retrospective study evaluating

the effect of placing a double-pigtail stent in a LAMS on pancreatic pseudocyst showed a significantly decreased rate of pseudocyst infection requiring reintervention when double-pigtail-stent was placed in a LAMS as compared to only LAMS [14]. The authors attributed infections to the presence of undigested food in the cyst and clogging of LAMSs.

We report the first case of symptomatic infected LEC of the pancreas successfully managed with EUS guided endoscopic drainage with LAMS similar to pseudocyst drainage. LAMS should be removed after resolution of symptoms, preferably within 4–6 weeks to minimize complication. LAMS in our patient was removed after 1 week, and the patient was then successfully managed conservatively.

## Conclusion

Lymphoepithelial cysts of the pancreas is a rare type of benign pancreatic cystic lesion that is difficult to diagnose. EUS plays an essential role in the diagnosis and management of these cysts. Asymptomatic LECs are managed conservatively, but symptomatic LECS have traditionally been managed with surgical resection. Alternatively, in selected cases of symptomatic LEC, symptom resolution can be achieved with EUS guided endoscopic drainage with LAMS, after confirmation of diagnosis.

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