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CASE REPORT

Recurrence of a Pancreatic Cystic Lymphangioma After Diagnosis and Complete Drainage by Endoscopic Ultrasound with Fine-Needle Aspiration

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ABSTRACT

Context Lymphangiomas are uncommon benign neoplasms that result from a blockage of the lymphatic system. Pancreatic cystic lymphangiomas are extremely rare. Pancreatic cystic lymphangiomas have been classically diagnosed on histopathologic examination following surgical excision, but recent reports have demonstrated successful diagnosis using endoscopic ultrasound with fine-needle aspiration (EUS-FNA). Data on the natural history of these lesions following drainage via EUS-FNA are lacking. We present a case of successful initial diagnosis and drainage of a pancreatic cystic lymphangioma using EUS-FNA, with unfortunate recurrence of the lesion four months later. **Case report** A 50-year-old female was evaluated for epigastric abdominal pain and nausea. CT scan revealed a 4 cm retroperitoneal cystic lesion. EUS-FNA was performed with complete drainage of the lesion using a 22-gauge needle. Twenty-five mL of chylous white fluid was obtained with laboratory analysis consistent with a pancreatic cystic lymphangioma. The patient was symptom-free for 4 months following drainage, but eventually had symptoms again with a CT scan confirming recurrence. **Conclusions** While EUS-FNA is effective for the diagnosis of pancreatic cystic lymphangiomas, its role in the management of these lesions is questionable. Temporary relief of symptoms can be achieved after EUS-guided drainage, but recurrence is a concern. Definitive cure likely requires complete surgical excision.

INTRODUCTION

Lymphangiomas are uncommon benign neoplasms that result from a blockage of the lymphatic system [1]. They are most often found in the head and neck region, and occur most commonly in children [2]. Lymphangiomas in the abdomen are very rare. Lymphangiomas are considered to be of pancreatic origin if they are within the parenchyma, adjacent to the pancreas, or connected to it by a pedicle [3]. Pancreatic cystic lymphangiomas are extremely uncommon, representing less than 1% of all lymphangiomas and about 0.2% of pancreatic neoplasms [4].

Conventional radiology has generally been inadequate for the evaluation of cystic lesions of the pancreas, and pancreatic cystic lymphangiomas have been classically diagnosed on histopathological examination following surgical excision [5]. However, there have been, recent reports of successful diagnosis of pancreatic cystic lymphangioma using endoscopic ultrasound with fine-needle aspiration (EUS-FNA) [4, 5, 6, 7, 8, 9, 10], yet data on the natural history of these lesions following drainage via EUS-FNA are lacking. We present a case of successful initial diagnosis and drainage of a pancreatic cystic lymphangioma using EUS-FNA, with unfortunate recurrence of the lesion four months later.

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Key words Diagnosis; Endosonography; Lymphangioma, Cystic; Pancreatic Cyst; Recurrence

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CASE REPORT

A 50-year-old female was evaluated for a one-year history of epigastric and left upper quadrant abdominal pain. The pain had worsened over the past two months and became accompanied by nausea, vomiting, and decreased appetite. A CT scan (Figure 1) was ordered

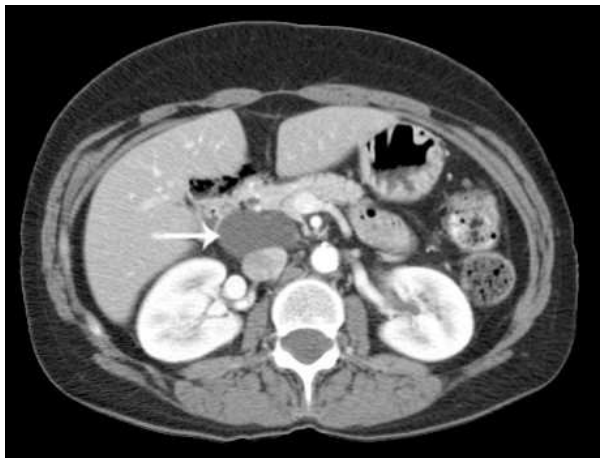

Figure 1. CT scan image showing the cystic lesion (arrow).

and revealed a 4 cm nonspecific retroperitoneal cystic lesion anterior to the inferior vena cava and medial to the second portion of the duodenum. Endoscopic ultrasound (EUS) was subsequently performed to better characterize this lesion. On echo-endoscopic examination, there was noted to be a 3.6 cm cyst adjacent to the head of the pancreas (Figure 2). FNA was performed using a 22-gauge needle, and the cyst was completely drained with one pass. Approximately 25 mL of chylous white fluid (Figure 3) was obtained and sent for cytology and laboratory analysis. Cytology was negative for malignancy and revealed fibrinous fluid with numerous and relatively uniform small lymphocytes present. Fluid analysis revealed an amylase level of 123 U/L (reference range: 25-115 U/L), CEA of 2.98 ng/mL (reference range: 0-3 ng/mL), and a markedly elevated triglyceride level of 3,379 mg/dL (this value is clearly recognizable as being markedly elevated although there is no defined reference range for triglyceride level in pancreatic cysts in our laboratory). This was consistent with a diagnosis of a chylous pancreatic cystic lymphangioma.

The patient reported complete resolution of her symptoms following the procedure. Due to the benign nature of these lesions, it was decided to initially proceed with expectant management with close clinical follow-up in hopes of sparing the patient potentially unnecessary surgery. The patient did well and remained symptom-free for the next four months, but then unfortunately began having recurrent epigastric pain and nausea. A CT scan was repeated and demonstrated recurrence of the lesion at the same previous location. The patient was subsequently referred for surgical management.

DISCUSSION

Endoscopic ultrasound has emerged as an extremely useful modality in the diagnosis and management of cystic lesions of the pancreas [11]. EUS can provide details of the outer borders and internal architecture of cystic pancreatic lesions, thereby narrowing the differential diagnosis of pancreatic cysts. In the appropriate clinical scenario, cystic fluid analysis by

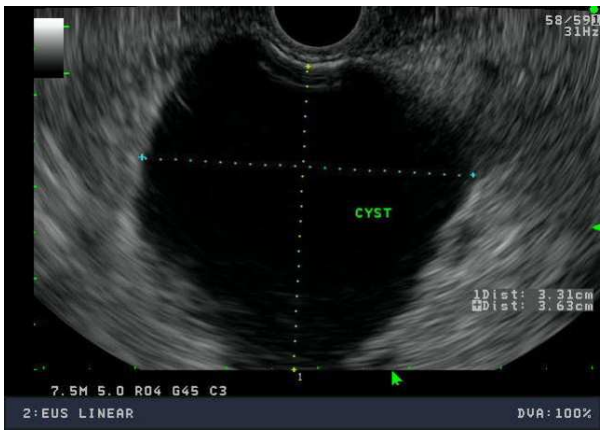

Figure 2. Endoscopic ultrasound image showing the cystic lesion prior to drainage by fine-needle aspiration.

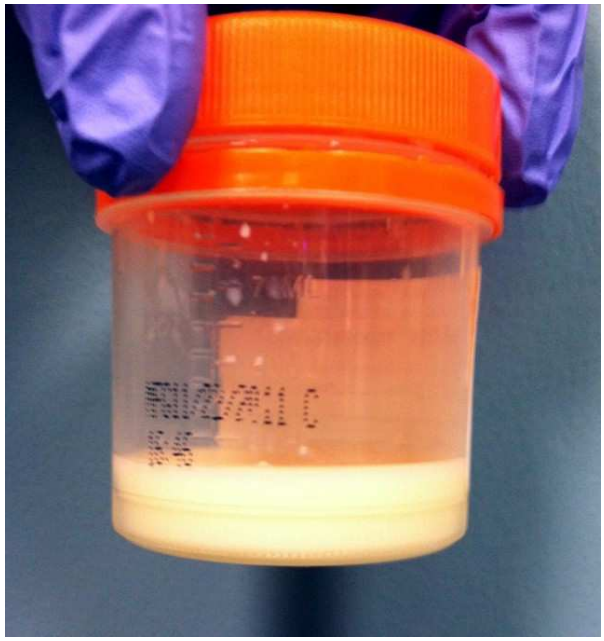

Figure 3. Container showing the white, chylous fluid aspirated from the cystic lesion.

EUS-FNA can increase the utility of EUS. Analyzing fluid for CEA, amylase, and CA 19-9 can help further narrow the differential and may sometimes confirm a suspected diagnosis [12].

The role of EUS-FNA in the evaluation of uncommon cystic lesions such as pancreatic cystic lymphangiomas has been less clear, but recent reports, including this one, have demonstrated the usefulness of EUS-FNA in the evaluation of these rare lesions. Many pancreatic cystic lymphangiomas contain serous fluid, in which case the fluid analysis may not lead to a definitive diagnosis. However, when the fluid is grossly chylous and the triglyceride level is significantly elevated, as in this case, the diagnosis of a pancreatic cystic lymphangioma can be made with confidence. Sampling the collapsed walls of the cyst with the FNA needle may also increase the diagnostic yield [8].

Pancreatic cystic lymphangiomas are most often discovered incidentally in asymptomatic patients, but they can sometimes be symptomatic and may also rarely be associated with complications such as infection, rupture, torsion, bleeding, or obstruction [3, 5]. Pancreatic cystic lymphangiomas are not considered premalignant lesions, and consequently, expectant management with clinical follow-up and surveillance imaging has been proposed as a reasonable approach to management following drainage with EUS-FNA, especially in asymptomatic patients [4, 5, 10]. In symptomatic patients, nonsurgical management is more controversial. One report [6] stated that there was absence of disease progression or complications in two cases 5 months after drainage with EUS-FNA, while another report [7] mentioned that abdominal ultrasound one year after initial drainage with EUS-FNA confirmed the presence of an unchanged lesion with the patient remaining asymptomatic. However, other

authors insist that complete surgical excision is mandatory to prevent recurrence [13, 14]. In this particular case, the patient initially had complete relief of her symptoms following EUS-guided drainage. Unfortunately, this relief was only temporary with recurrence of the lesion and symptoms four months later.

In conclusion, while EUS-FNA is highly effective for the diagnosis of pancreatic cystic lymphangiomas, its role in the definitive management of these lesions is questionable. Temporary relief of symptoms can be achieved after EUS-guided drainage, but as this case illustrates, recurrence is a tangible concern. Expectant management with close clinical follow-up and surveillance imaging may be a reasonable approach in some cases, particularly in asymptomatic patients and poor surgical candidates, but surgical intervention should likely be offered to patients with significant symptoms. In patients who desire less invasive therapy, repeat drainage via EUS-FNA is likely safe and may be an alternative approach. However, definitive cure likely requires complete surgical excision.

Conflict of interest The authors have no conflict of interest to declare

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