Lymphangioma Presents as Peripancreatic Cystic Neoplasm: The Utility of Endoscopic Ultrasound

Heather Pemberton1*, Mohamed Sultan2, Walid Chalhoub2, Shannon Morales3, Mohammed Al-Bugeay2, Jill Smith2 and Nadim Haddad2

1Georgetown University School of Medicine, 3900 Reservoir Road, Washington, DC 20007, USA
2Division of Hepatology and Gastroenterology, MedStar Georgetown University Hospital, 3800 Reservoir Road, Washington, DC 20007, USA
3Department of Medicine, MedStar Georgetown University Hospital, 3800 Reservoir Road, Washington, DC 20007, USA

*Corresponding author: Heather Pemberton, Georgetown University School of Medicine, 3900 Reservoir Road, Washington, DC 20007, USA, Tel: 925-487-3311; E-mail: hep8@georgetown.edu

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Abstract

Pancreatic lymphangiomas are very rare benign cystic lesions that arise from abnormal proliferation of the lymphatic vessels. Although benign, pancreatic lymphangiomas may mimic more concerning cystic neoplasms and therefore require a thorough diagnostic workup. Herein we report a case of a 50-year-old female who presented with a peripancreatic cyst measuring 2.5x3.6x4.7cm that was discovered incidentally during an abdominal ultrasound. She underwent endoscopic ultrasound with fine needle aspiration where chylous fluid was drained. Cytology was negative for malignancy and fluid chemistry demonstrated a significantly elevated triglyceride level consistent with a pancreatic lymphangioma. It is unclear whether asymptomatic pancreatic lymphangiomas are best managed conservatively or with surgical intervention. The patient has not proceeded with surgery at this time.

Keywords: Lymphangioma; Pancreas; Endoscopic ultrasound with fine needle aspiration.

Introduction

A lymphangioma is a rare benign cystic lesion that results from abnormal proliferation of the lymphatic vessels. Specifically, they are thought to arise from congenital lymphatic malformations that result in obstruction of lymphatic flow and the development of lymphangiectasis [1-5]. Cystic lymphangiomas most commonly present in children and frequently affect the head, neck, and axillary regions. Although adult forms of lymphangiomas are uncommon, most cases occur in the mesentery and retroperitoneum with the pancreas being one of the rarest sites of origin [5,6]. Despite their benign nature, pancreatic lymphangiomas may mimic more concerning cystic neoplasms and therefore further diagnostic work-up is often necessary. The use of endoscopic ultrasound (EUS) with fine-needle aspiration (FNA) has proven to be an invaluable tool to establish a definitive pre-operative diagnosis as it provides a safe and effective means for obtaining cystic fluid for cytological analysis [1,3,5].

Case Report

A 50-year-old female with a history of Hepatitis C was referred by her primary care physician for evaluation of her known pancreatic cyst. The cyst was discovered incidentally during an abdominal ultrasound a year prior. A CT scan of the abdomen at that time revealed a non-enhancing cystic structure inferior and adjacent to the pancreatic body measuring 2.5 cm x 3.6 cm x 4.7 cm with no associated pancreatic or intra-hepatic ductal dilation. The patient was asymptomatic and denied any weight loss, abdominal pain, nausea, vomiting, change in bowel habits, and had no family or personal history of gastrointestinal or pancreatic malignancies. Physical examination was unremarkable and no interventions were made at that time. A 12-month follow-up CT demonstrated that the cystic lesion remained unchanged in size. Although the patient remained asymptomatic, she was becoming increasingly anxious about the presence of the mass and was therefore referred for EUS with FNA for further evaluation and to rule-out malignancy. During the EUS, an anechoic cystic lesion measuring 37x24 mm was identified adjacent to the body of the pancreas (Figure 1).

The pancreatic duct and parenchyma appeared normal. A 22-gauge needle was advanced into the lesion without resistance. Upon aspiration, 10 mL of chylous fluid was drained with cytology negative for malignancy and fluid chemistry demonstrating a significantly elevated triglyceride level (>11,000 mg/dL) as well as elevated glucose (309 mg/dL) and total protein (10.6 mg/dL) levels consistent with a peripancreatic lymphangioma (Figure 2).
Definitive pre-operative diagnosis as it provides high quality imaging analysis [1,3,5]. Specifically, EUS-FNA has been reported to have a specificity of 100% and accuracy of 86% in determining the etiology of cystic pancreatic lesions [3,10]. To diagnose a lymphangioma, the aspirated fluid must be chylous appearance and have an elevated triglyceride level similar to that seen in our patient. Aspiration of serous fluid with only a mildly elevated triglyceride level requires further diagnostic workup with possible surgical resection [3]. While malignant transformation of a pancreatic lymphangioma has not been reported in the literature, there is a general consensus that symptomatic lesions are best treated with complete surgical resection, if possible, as it reduces the risk of complications and recurrence [7]. The treatment of asymptomatic lesions such as that seen in our patient, however, remains controversial. Both surgical resection and expectant management with follow-up appear to be reasonable approaches and the risks of surgery must be weighed against the possibility of developing complications such as hemorrhage, torsion, rupture, obstruction and infection in those who prefer a more conservative approach [3,11].

Conclusion

Pancreatic cystic lymphangiomas are exceptionally rare benign tumors that are best distinguished from more concerning cystic neoplasms through EUS with FNA as it is minimally invasive and highly accurate. Complete surgical resection of symptomatic lesions, if possible, appears to be the best therapeutic option to reduce the risk of complications and recurrence. The treatment of asymptomatic lesions however, remains controversial and further studies are needed to delineate if a surgical approach versus watchful waiting is indicated.

Discussion

Cystic lymphangiomas are benign tumors believed to arise from congenital lymphatic malformations that result in obstruction and dilation of the lymphatic vessels. Over 90% of cases are discovered before the age of two with the vast majority presenting in the head, neck, and axillary regions [6-12]. A pancreatic lymphangioma as defined as originating from the pancreatic parenchyma, lying in proximity to the pancreas, or attached to the pancreas by a pedicle are exceptionally rare accounting for less than 1% of all lymphangiomas and 0.2% of pancreatic lesions [3,8,9,13]. As in the case of our patient, the lesions are usually asymptomatic and discovered incidentally when undergoing workup for another disease process [3,5,6]. Patients may present with variable clinical symptoms, however, including abdominal pain, nausea, vomiting, fever, weight loss and/or a palpable abdominal mass [2,3]. While abdominal ultrasonography or an abdominal Computed Tomography (CT) scan may reveal the presence of a lymphangioma, a definitive diagnosis cannot be determined based on imaging alone as these modalities fail to characterize the lesion. Because lymphangiomas can mimic the morphology of more concerning lesions such as mucinous cystic neoplasms, further characterization of the lesion is frequently warranted [2,3]. Currently, the use of EUS-FNA cyst aspiration is recommended to establish a definitive pre-operative diagnosis as it provides high quality imaging of the pancreas and a safe means for obtaining fluid for cytological analysis [1,3,5]. Specifically, EUS-FNA has been reported to have a specificity of 100% and accuracy of 86% in determining the etiology of cystic pancreatic lesions [3,10]. To diagnose a lymphangioma, the aspirated fluid must be chylous appearance and have an elevated triglyceride level similar to that seen in our patient. Aspiration of serous fluid with only a mildly elevated triglyceride level requires further diagnostic workup with possible surgical resection [3]. While malignant transformation of a pancreatic lymphangioma has not been reported in the literature, there is a general consensus that symptomatic lesions are best treated with complete surgical resection, if possible, as it reduces the risk of complications and recurrence [7].

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