Incidental Pancreatic Neuroendocrine Neoplasm Diagnosed by Confocal Laser Endomicroscopy: A Case Report and Brief Literature Review

by Ritu Raj Singh, Mariajose Rose De Leon, Neil R. Sharma

While a majority of the pancreatic cystic lesions (PCL) are benign incidentalomas, many of them can be neoplastic and preneoplastic. Endoscopic ultrasound-guided fine needle aspiration is commonly utilized for the diagnosis of these lesions; however, the sensitivity and specificity are suboptimal. Confocal laser endomicroscopy (CLE) is a more reliable diagnostic tool that is being increasingly used for the diagnosis of PCL. Cystic neuroendocrine neoplasms (NEN) of the pancreas are thought to contribute to a very small proportion of all PCL. Nonetheless, they have been found more commonly in recent studies than previously reported. Here, we present a case report of a patient with cystic NEN in the pancreatic tail that was incidentally found, and CLE was used for diagnosis.

INTRODUCTION

With widespread use of abdominal imaging for diagnostic and screening purposes, the prevalence of pancreatic cystic lesions (PCL) has surged, ranging from less than 1% in an earlier study to 2.6% in a more recent report.[1,2] Endoscopic ultrasound-guided fine needle aspiration (EUS-FNA) is utilized for the diagnosis of these lesions, however, the diagnostic yield is often limited by inadequate cells for analysis in the cystic fluid aspirated. Confocal laser endomicroscopy (CLE), also known as optical biopsy, is a technique that utilizes low frequency laser beam to obtain real-time pictures of the tissues that mimics histological images. While there is emerging evidence of the application of CLE in the diagnosis of mucinous PCL with sensitivity, specificity and accuracy of approximately 90%, there are only few reports of the use of CLE in the diagnosis of cystic neuroendocrine neoplasms (NEN) of the pancreas.[3-5]

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Here, we present the case history of a patient with incidentally detected PCL who underwent CLE and was diagnosed to have a NEN of the pancreas.

Case Report

A 76-year-old male with past medical history of arthritis, diabetes type II, hypertension and neuropathy, presented to emergency department for evaluation of acute onset right flank pain and nausea, without vomiting. Physical exam revealed normal vital signs and positive findings included tenderness in the right costovertebral angle. There was no history of weight loss or fever. There was no history of tobacco, alcohol, or drug use. Family history was significant for multiple types of cancer in siblings, including skin, breast, pancreas, and bladder.

Computed tomography (CT) of the abdomen and pelvis revealed right nephrolithiasis with hydronephrosis, and an incidental finding of a low-density lesion in the tail of the pancreas measuring 1.7 x 2.1 x 2.3 cm^3 (Figure 1). Magnetic resonance

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cholangiopancreatography (MRCP) showed a round rim-enhancing cyst along the pancreatic tail measuring 17 mm x 17 mm axially and 21 mm craniocaudally without definitive communication with the pancreatic duct (Figure 2). Lab results were unremarkable other than elevated serum lipase of 186 U/L and CA19-9 of 54.7 IU/ml.

Following treatment of his nephrolithiasis he was referred for an endoscopic ultrasound (EUS) for further evaluation of the pancreatic lesion.

EUS with CLE, FNA and Wall Biopsy

EUS showed normal main pancreatic duct and a 20.5 mm x 19.4 mm well circumscribed anechoic cystic lesion with atypical wall thickening in the tail of the pancreas without internal septations, solid component or mural nodules within the cyst. Under ultrasound guidance, a 19 g Boston Scientific needle was used for cyst puncture. This has been illustrated in Figure 1. Advanced imaging with CLE (Cellvizio) was performed. The wall of the cyst was surveyed and cluster of cells with trabecular pattern was seen consistent with NEN, as shown in Figure 2. Biopsy of the wall was performed with micro-forceps and the cyst was aspirated.

Pathology revealed neuroendocrine cell proliferation with cells positive for synaptophysin (Figure 3), chromogranin, and pancytokeratin by immunostains, Ki-67 proliferation was 1% and mitosis was not identified favoring well-differentiated neuroendocrine tumor, WHO grade 1.

DISCUSSION

PCL are common and often asymptomatic with a majority discovered as incidental finding on imaging for other indications. They encompass pancreatic pseudocysts, intraductal papillary mucinous neoplasms, serous cystadenomas, and rarely cystic NEN. Imaging features are not characteristic of any cyst and in the absence of specific findings, like presence of a solid component or involvement of the main pancreatic duct, decision to defer further follow up or treatment cannot be made with confidence. Cystic NEN of pancreas account for less than 1% of all PCL (10-17% of NEN).[6,7] The prevalence increases with age, the highest being in those 80 years and older (8.7%).[1]

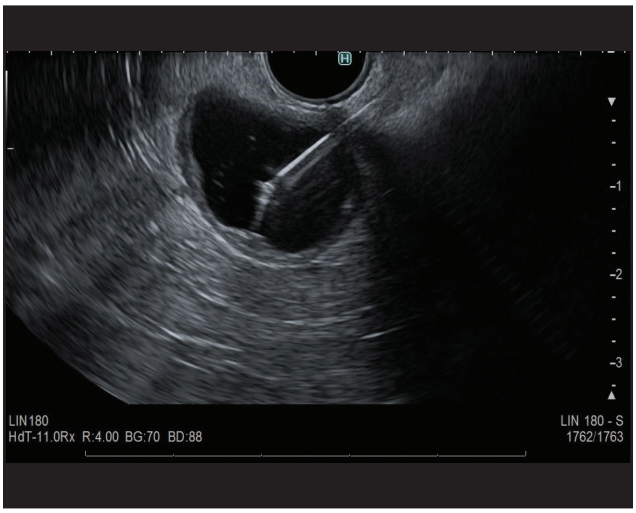

Figure 1.

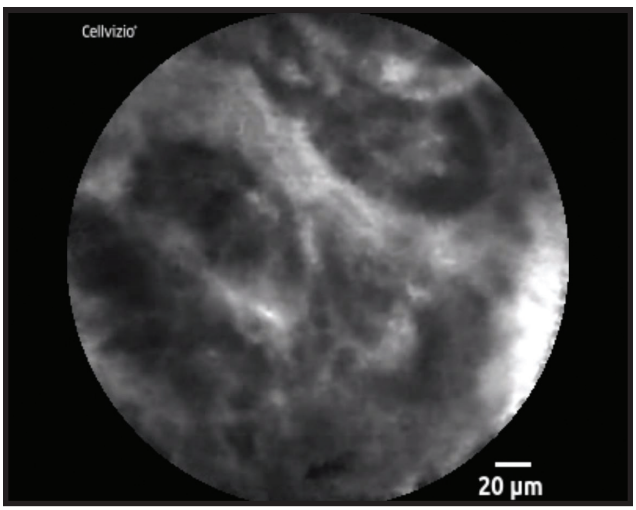

Figure 2.

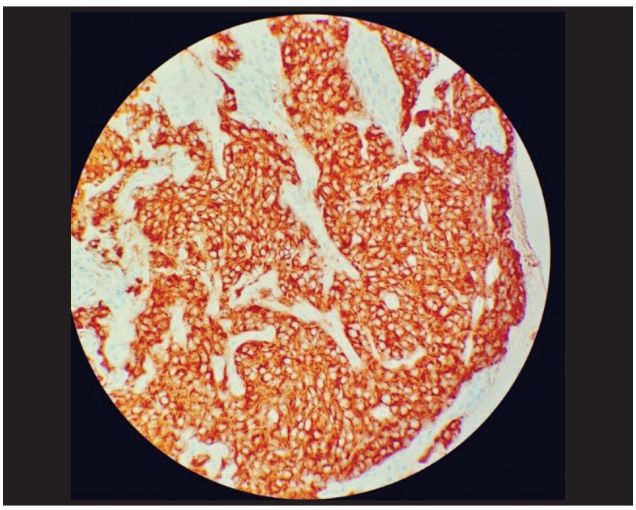

Figure 3.

Current guidelines recommend further evaluation and surveillance of PCL in medically fit patients other than asymptomatic pseudocysts and serous cystadenomas which have very low to no malignant potential. Given rarity of cystic NEN there is no clear guidance, and EUS-FNA is suggested. However, the yield of EUS-FNA is offset by low cellularity and nonspecific nature of the commonly used biomarkers, like carcinoembryonic antigen (CEA).[8]

CLE has developed as a real-time diagnostic tool for PCL. There is limited data on the utility of CLE in cystic NEN due to infrequent finding of these lesions. Nonetheless, the results from other cystic lesions can be extrapolated to these tumors. Our case report illustrates that not all PCL are benign and pancreatic NEN can present as an incidental cyst. The findings on CLE were characteristic of a cystic pancreatic NEN which was verified on histopathology.

CONCLUSION

Cystic NEN are uncommon, however, they are being recognized more frequently than before, and can present as an incidental cyst typically in the pancreatic body or tail. CLE can provide real-time diagnosis of PCL including cystic NEN. Current evidence for CLE in PCL is evolving and the initial results are encouraging. Future investigations of larger scale are expected to support this novel diagnostic modality as a standard of care. ■

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