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CASE REPORT | PANCREAS

Endoscopic Ultrasound-Guided Needle-Based Probe Confocal Laser Endomicroscopy (nCLE) of Intrapancreatic Ectopic Spleen

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Abstract

Accessory spleens and splenosis represent the congenital and acquired type of ectopic splenic tissue. Generally, they are asymptomatic entities posing as solid hypervascular masses at the splenic hilum or in other organs, such as the pancreas. Intrapancreatic ectopic spleen mimics pancreatic neoplasms on imaging studies, and due to the lack of radiological diagnostic criteria, patients undergo unnecessary distal pancreatectomy. We present the first case of intrapancreatic ectopic spleen in which the concomitant use of needle-based probe confocal laser endomicroscopy and fine-needle aspiration supported the final diagnosis.

Introduction

Ectopic splenic tissue is a rare entity that manifests as accessory spleen or splenosis and is frequently misdiagnosed as a pancreatic hypervascular neoplasm.[1] The role of imaging studies such as computed tomography (CT), magnetic resonance imaging (MRI), and endoscopic ultrasound (EUS) is crucial for the identification of pancreatic masses. However, the information these studies provide is limited when differentiating malignancy. As there is no radiographic criteria established for ectopic spleens, use of needle-based probe confocal laser endomicroscopy (nCLE) prior to EUS-guided fine-needle aspiration (FNA) in the diagnosis of pancreatic masses may increase diagnostic accuracy.

Case Report

A 24-year-old woman was referred for an EUS evaluation after a CT scan showed a 3 x 2.9-cm round hypervascular mass in the tail of the pancreas (Figure 1). She presented with 1 month of worsening dull epigastric pain radiating to upper right and left quadrants and the lumbar region. Her past medical history included thrombotic thrombocytopenic purpura (TTP), and she underwent splenectomy 5 years ago for profound thrombocytopenia. Her family history was significant for a second-degree relative with pancreatic cancer. Laboratory data was normal, with peripheral smear with Howell Jolly bodies 1+ and negative tumor markers.

EUS revealed a 2.8 x 2.9-cm round, well-defined homogenous hypoechoic mass in the pancreatic tail with no other endosonographic pancreatic abnormalities (Figure 2). A 19-gague needle was preloaded with an AQ-Flex 19 (Mauna Kea Paris, France) probe, and nCLE was performed using the probe. The mass was punctured and 2.5 mL of 10% fluorescein sodium was injected. Findings on the nCLE demonstrated numerous thick white bands with floating small, black particles inside the bands, suggesting the presence of blood vessels, and floating erythrocytes were identified (Figure 3). Subsequently, 4 passes of FNA using a 22-gauge needle were obtained. Side-by-side

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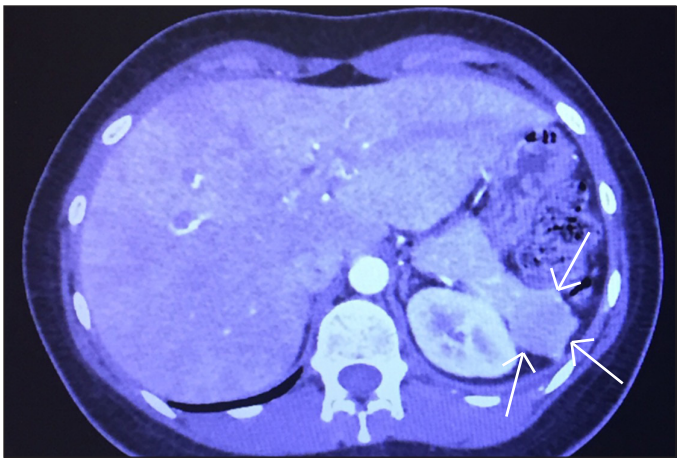

Figure 1. Computed tomography showing a 3 x 2.9-cm round hypervascular hypodense mass in the tail of the pancreas (arrow).

pathology and endomicroscopy review supported the final diagnosis of intrapancreatic splenic tissue (Figure 4). There were no complications following these procedures. During her 9-month follow-up, the pain resolved with proton pump inhibitors prescribed once daily, her platelets maintain between normal ranges, and no hematologic recurrence signs.

Discussion

Intrapancreatic ectopic spleen is a rare entity that arises as a result of a birth defect (accessory spleen) or an acquired condition (splenosis). Its presentation is usually asymptomatic, but heterotopic tissue has been reported as incidental findings in patients with upper gastrointestinal disorders associated with nausea and abdominal pain.[2,3] While ectopic spleens are considered benign, their presence is strongly indicative of underlying disease and health progression.

The development of an accessory spleen is congenital, due to an alteration during the differentiation of mesenchymal cells

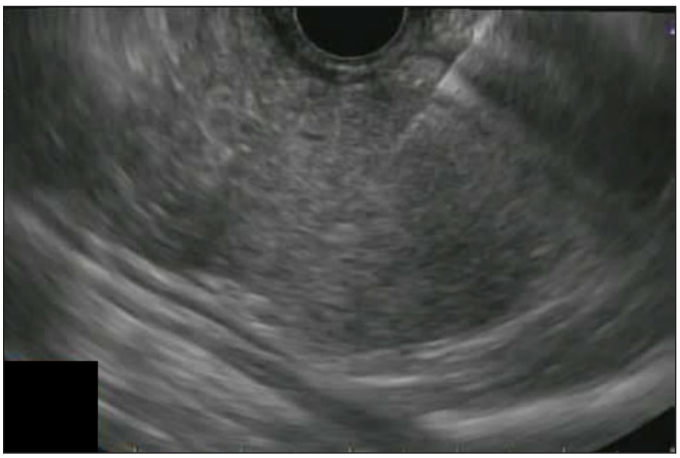

Figure 2. Endoscopic ultrasound showing a 2.8 x 2.9-cm round well-defined homogenous hypoechoic mass in the tail of the pancreas.

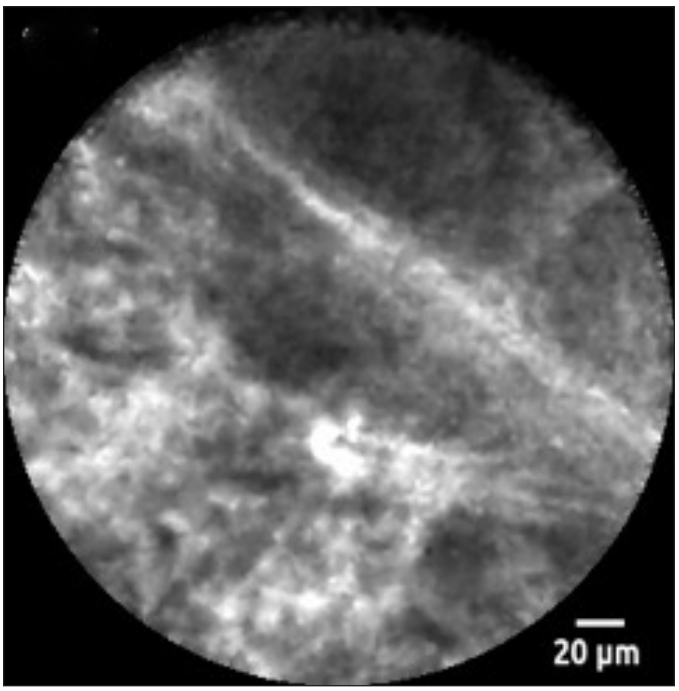

Figure 3. nCLE of pancreatic mass showing numerous thick white bands with floating small black particles inside the bands suggesting the presence of blood vessels and floating erythrocytes.

leading to the formation of splenic tissue along the path of splenic vessels.[4] Their location is limited to their embryological origin, finding them in or near the splenic hilum, pancreas, jejunum, colon, and even pelvis, scrotum, and ovary.[5] In a study of 3,000 autopsies, 80% of accessory spleens reported were found in the splenic hilum, followed by 17% found in the tail of the pancreas.[6] Accessory spleens have the same histological structure and functionality of a normal spleen. They usually present as small, scattered masses supplied by a branch of the splenic artery.[7] Currently, there is no epidemiological study regarding accessory spleens; however, it is estimated that their prevalence is 10–30%.[6]

Splenosis responds to any process that results in a traumatic or spontaneous splenic rupture. In trauma, multiple and small fragments of viable or degenerating splenic tissue migrate and implant into adjacent structures. Tissue can also disperse hematogenously by forming splenic pulp emboli that travels through the bloodstream until they reach another cavity.[7] Some fragments may preserve normal splenic structure and functionality, but most splenic fragments analyzed have shown distorted architecture with decreased elastic tissue and lack of hilum. Vascular supply usually derives from adjacent or invaded structures. Most splenic fragments are found in the upper left quadrant; however, cases of liver, chest, pelvis, brain, and subcutaneous splenosis have been reported.[2] The risk of splenosis increases in patients undergoing laparoscopic splenectomy. Approximately 76% of patients with lapa-

roscopic splenectomy may develop splenosis due to possible splenic remains left behind during surgery.[8]

Intrapancreatic spleens radiologically simulate pancreatic endocrine tumors, acinar cell carcinomas, solid pseudopapillary neoplasms, and adenocarcinomas. The use of nCLE allows direct tissue architecture evaluation through the endoscope, providing a high-magnification and resolution image with better discernment of pancreatic lesions. Contrast agents may or may not be used, but the administration of intravenous fluorescein presents an image very similar to histology slides without compromising the integrity of real-time imaging.[9] Optical biopsy of normal pancreatic tissue shows acinar cells as dark lobular structures, "coffee beans," and adipocytes as grey oval structures. Usual malignant characteristic findings or common pancreatic lesions are associated with finger-like papillary projections, dark rings with white cores, numerous superficial vascular network, crypt-like structures, cyst wall with epithelial borders, and presence of bright particles.

Intrapancreatic ectopic spleen should be included as a differential diagnosis for all unidentified pancreatic tail masses. A study demonstrated that 10 of 11 intrapancreatic accessory spleens resected were misdiagnosed as pancreatic neoplasms, and only 1 was credited with a correct diagnosis due to the presence of idiopathic thrombocytopenic purpura.[10]

We could not identify whether the presence of ectopic splenic tissue was congenital or acquired. Imaging studies before the patient underwent splenectomy did not show an intrapancreatic mass; therefore, post-splenectomy splenosis diagnosis was not excluded. In addition, both accessory spleens and splenosis fragments have the ability to enlarge and maintain normal splenic function as a compensation for splenectomy.[11,12] As a result, this compensatory mechanism can lead to a high rate recurrence of hematological diseases for which patients underwent splenectomy.[13]

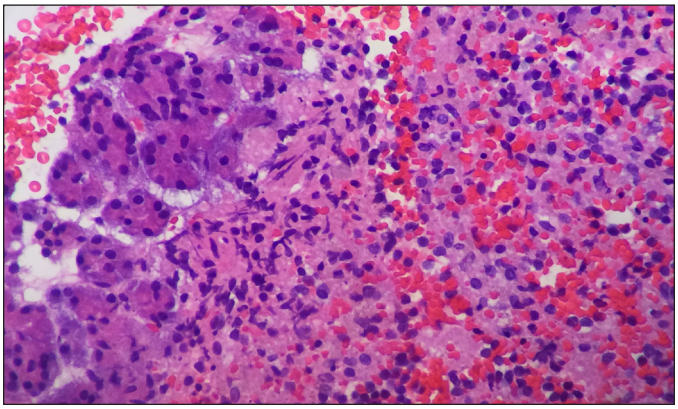

Figure 4. Histology slide showing ectopic splenic tissue with white pulp (darker purple) on the upper left and red pulp in the middle with residual normal pancreas tissue.

The EUS-FNA and pathology gives the final diagnosis of ectopic spleen, but the use of nCLE may significantly reduce post-FNA complications such as hemorrhage and pancreatitis. The addition of nCLE to the usual diagnosis protocol for pancreatic lesions proposes an extra minimally invasive diagnostic mean for physicians. Real-time endomicroscopy provides optimal differentiation of pancreatic masses, potentially reducing malignancy misdiagnosis and thus unnecessary pancreatic resections.

Disclosures

Author contributions: JM Nieto wrote the manuscript, researched the literature, and is the article guarantor. AB Bastidas wrote the manuscript and researched the literature. D. Holloman and A. Lankarani critically revised the manuscript.

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