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Knife wielding radiologist: A case report of primary pancreatic lymphoma

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ABSTRACT

Majority of malignant pancreatic neoplasms are epithelial in origin and mostly arise from exocrine gland. Ductal adenocarcinoma compromises the major histological type of such tumors. Primary non-epithelial tumors of exocrine pancreatic gland are extremely rare and incorporate lymphoma and sarcoma. Primary pancreatic lymphoma compromises less than 0.5% of pancreatic malignancies. Primary pancreatic lymphoma can be difficult to differentiate from pancreatic adenocarcinoma and other neoplasms on imaging, and a correct diagnosis is crucial for appropriate patient management.

1. Introduction

Lesions involving the pancreas can broadly be subdivided, radiographically, into cystic versus solid, and non-neoplastic versus neoplastic lesions. Solid appearing neoplastic lesions usually include pancreatic adenocarcinoma as the most common, followed by less common solid pancreatic neuroendocrine tumors, solid pseudopapillary tumors, pancreatoblastoma, and pancreatic metastasis. Primary pancreatic lymphoma (PPL) is an extremely rare solid neoplasm. It constitutes of less than 2% of extranodal lymphomas and 0.5% of all pancreatic masses [1]. Radiographic studies, such as computed tomography (CT), various forms of ultrasonography (US), and magnetic resonance imaging (MRI), may demonstrate key features to help the radiologist offer confident reports to guide the workup and management of patients with pancreatic masses, as current treatment strategies and prognosis are substantially different for neoplastic and nonneoplastic lesions and for type of neoplastic lesion. Therefore, we report a case of PPL, focusing on current literature regarding key imaging features and clinical presentation, and why the ability to differentiate at the stage of diagnostic imaging can influence further management.

2. Case presentation

A 36-year-old female with history of tobacco and alcohol abuse presented with 4 weeks of progressive constant circumferential pain across the upper abdomen and lumbar region accompanied by nausea, vomiting, diarrhea and unintentional 11-pound weight loss.

2.1. Investigations

A contrast enhanced CT (CECT) of the abdomen and pelvis was performed with series obtained in the late arterial and delayed phases, revealing a large hypodense lesion centered within the junction of the neck/body of the pancreas with mild dilation of the pancreatic duct (Fig. 1). Adjacent to the lesion was a bulky conglomerate of mesenteric lymph nodes extending below the level of the renal veins. It was concluded that a primary malignancy with central necrosis was the primary concern. However, with the clinical history, additional considerations were given to inflammatory or infectious causes. The patient was clinically treated as having pancreatitis and provided with parental fluids, bowel rest, and pain control with improvement of symptoms and tolerance of oral intake after four days of inpatient management. Therefore, the patient was discharged home with arrangement made for an outpatient endoscopic ultrasound (EUS) and potential biopsy.

Within two days of discharge, the patient was unable to tolerate oral intake and had a return of symptoms. A repeat CECT with late arterial and delayed phases revealed interval enlargement of the hypoenhancing mass and adjacent bulky mesenteric adenopathy. The patient was transferred to a tertiary center and admitted for further workup.

Upon admission to the referring hospital, an abdominal US demonstrated a complex avascular cystic lesion surrounded with thick soft tissue rim (Fig. 2). There was mild dilatation of the extrahepatic biliary

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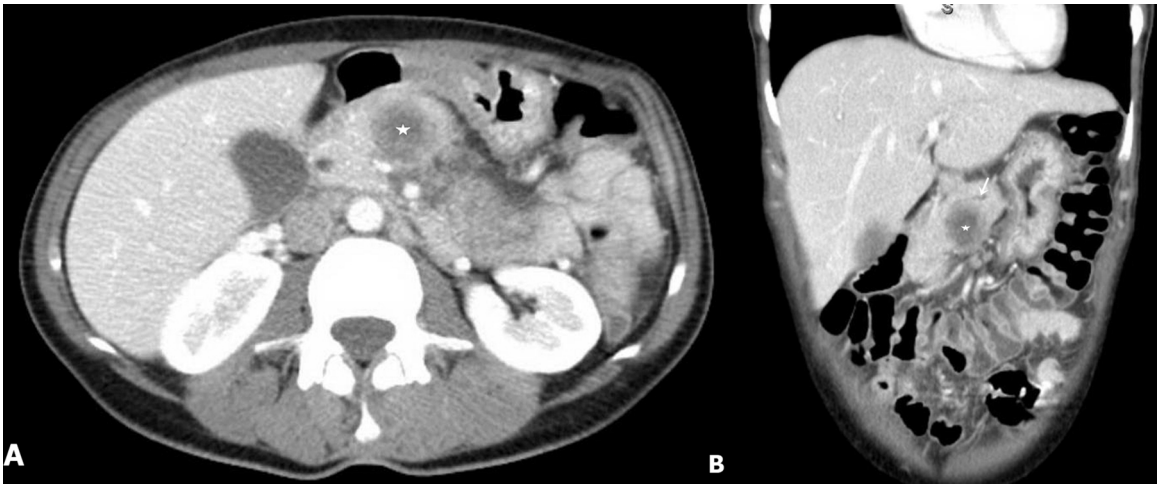

Fig. 1. (A) Contrast enhanced axial, and (B) Coronal CT scan demonstrates a hypodense mass within the proximal pancreatic body (star). Pancreatic duct is minimally dilated despite the large size of mass (arrow).

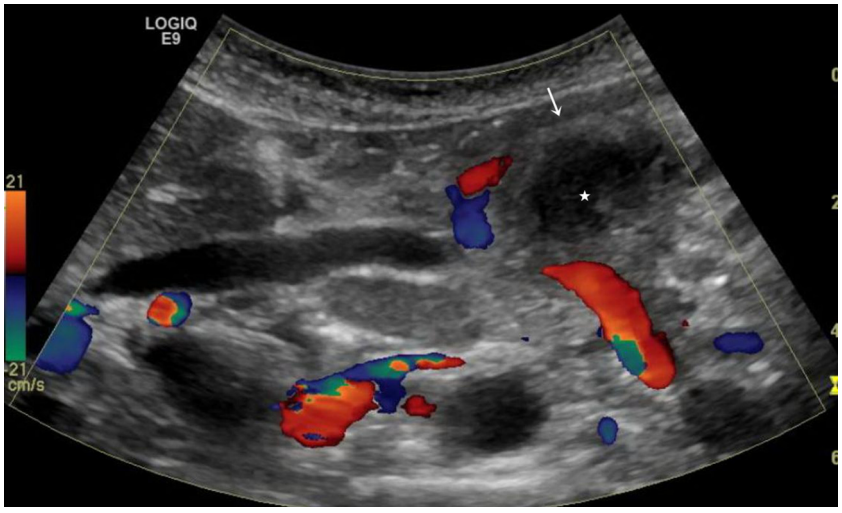

Fig. 2. Targeted US shows a complex avascular cystic lesion (star) surrounded by thick soft tissue rim (arrow).

ducts without a definitive cause. This was followed with further workup of magnetic resonance cholangiopancreatography (MRCP). MR identified a T2W intermediate lesion corresponding to the cyst seen on US (Fig. 3). There was associated mild dilatation and irregularity of the upstream main pancreatic duct. There were multiple enlarged peripancreatic, periportal, and mesenteric lymph nodes with a conglomerate surrounding the superior mesenteric artery (SMA) (Fig. 3). DWI/ADC demonstrated heterogeneous areas of diffusion restriction in the primary mass. However, the mesenteric lymphadenopathy demonstrated definitive and homogenous diffusion restriction (Fig. 4).

2.2. Differential diagnosis

After review of the radiologic studies, and consideration given to the patient's relatively young age, the leading diagnosis for the pancreatic mass and pattern of adjacent lymphadenopathy was a primary B-Cell pancreatic lymphoma.

2.3. Outcome and follow-up

A follow-up EUS guided pancreatic biopsy yielded pleomorphic malignant neoplasm (Fig. 5) strongly positive for ALK and CD30 on immunohistochemical stains (Fig. 6). CD20 was essentially negative, showing very rare cells with nonspecific granular staining, negative for CD3, and negative for cytokeratin AE1/AE3. Genetic analysis demonstrated fusion of the NPM/ALK gene, suggestive of a chromosome (2;5) (p23; q35) translocation. The overall combined findings were most consistent with anaplastic large cell lymphoma.

Further staging workup was performed, including CT of the chest as well as a positron emission tomography (PET). PET revealed increased FDG uptake corresponding with the pancreatic lymphoma and adjacent lymphadenopathy (Fig. 7). The patient was initiated on chemotherapy that included etoposide, prednisolone, oncovin, cyclophosphamide, and doxorubicin (EPOCH) with repeat PET/CT one month later demonstrating complete functional response.

3. Discussion

As mentioned previously, PPL is an extremely rare presentation of extranodal lymphoma and a high level of clinical suspicion must be maintained in order to differentiate from its more commonly encountered counterpart, adenocarcinoma. Current treatment strategies vary, and prognosis are substantially different with primary pancreatic lymphoma having a more favorable prognosis and response to radiation and/or chemotherapy. Therefore, when faced with a solid pancreatic mass, knowing when to direct the next steps in management towards a potential cure without surgical resection [2].

Imaging can suggest the diagnosis of primary pancreatic lymphoma