

## Primary pancreatic lymphoma: A diagnostic conundrum

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### To the Editor,

Primary pancreatic lymphoma (PPL) is an extremely rare non-epithelial tumor constituting far less than 1% of all pancreatic neoplasm [1, 2]. Clinical and imaging findings are otherwise non-specific in differentiation of pancreatic lymphoma from other pancreatic cancers. Herein, we report a case of PPL presenting as multiple masses in the pancreatic head, body and tail.

An 86-year-old female presented to the emergency department for evaluation of a three-week history of progressive fatigue and night sweats without anorexia, abdominal pain or weight loss. She had a medical history of diet-controlled type II diabetes mellitus and denied use of alcoholic beverages or tobacco products.

On admission, physical examination revealed a mildly pale woman without palpable adenopathy. She had a mildly distended abdomen, without tenderness, organomegaly or fluid thrill. Laboratory examination showed normal platelets and white blood cell count, hemoglobin 10.2 g/dL (11.5–15.5 g/dL), hematocrit 30.7% (34–36%), predominantly direct hyperbilirubinemia 2.7 mg/dL (0.1–1.0 mg/dL), lactate dehydrogenase of 327 U/L (100–190 U/L), alkaline phosphatase 313 U/L (38–135 U/L), normal amylase, lipase of 638 U/L (50–290 U/L), and marginal elevation in transaminases: AST 71 U/L (14–36 U/L), ALT 53 U/L (9–52 U/L). She had

normal complement levels, negative hepatitis panel, non-specific antinuclear antibody elevation, and an elevated CA 19-9 level at 102 U/mL (<55 U/mL). Abdominal ultrasound reported intra-hepatic and common biliary ductal dilatation, cholelithiasis with a small amount of ascites, and a questionable mass in the pancreatic tail. Magnetic resonance cholangiopancreatography reported multiple cystic lesions in the uncinate process, pancreatic body, and tail associated with a non-dilated pancreatic duct.

Computed tomography (CT) scan of the abdomen and pelvis were not suggestive of widespread adenopathy but revealed extensive infiltrative appearance in the retroperitoneum suspicious for lymphoma associated with pleural effusions and ascites. Evaluation with endoscopic ultrasound revealed two heterogeneous hypoechoic cystic mass lesions in the pancreas: a 3.2x2.2 cm lesion in the distal body and a 3.5x3.2 cm multicystic lesion in the pancreatic head each of which were biopsied (Figure 1). Additional findings included common biliary ductal dilatation, multiple hypoechoic cystic lesions in the



Figure 1: Hypoechoic pancreatic head mass on EUS in patient with primary pancreatic lymphoma

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pancreatic body and tail, and duodenal obstruction from the pancreatic head mass. Histopathological analysis of the specimen revealed a CD20 positive diffuse large B-cell lymphoma (Figure 2). Immunohistochemistry was additionally positive for the following tumor markers: CD45, CD10, B-cell lymphoma (BCL) 6, and BCL-2; and negative for cytokeratin AE1/AE3 cocktail, chromogranin, synaptophysin, CD56, cyclin D1, CD23, CD3 and CD5. Ki-67 was reported as equivocal.

Though no surgical intervention was performed, she successfully had percutaneous biliary drainage with stent placement and started systemic chemotherapy with rituximab, cyclophosphamide, vincristine, and prednisone (R-CVP) within days of diagnosis. She was dose reduced for the first cycle due to obstructive jaundice with peak total hyperbilirubinemia of 5.8 mg/dL (0.1–1.0 mg/dL) which subsequently resolved. CT imaging after three cycles of systemic chemotherapy showed interval decrease in biliary ductal dilatation, near complete resolution of bilateral pleural effusions and ascites, and decrease in size of pancreatic lesions. She has completed six cycles of therapy per NCCN guidelines with a partial response expected at the least based on clinical, laboratory, and imaging parameters.

Most reported cases of PPL reference a median age of presentation of 55–60 years with a male preponderance. Clinical presentation and imaging studies are often very similar to that of pancreatic adenocarcinoma [2, 3]. Primary pancreatic lymphoma often presents as an isolated pancreatic mass mimicking adenocarcinoma, but without marked pancreatic ductal dilatation on imaging (Table 1). Unlike other pancreatic cancers, obstructive jaundice, as was seen in our patient, is an uncommon presenting feature of pancreatic lymphoma [3]. Histopathological analysis by fine-needle aspiration (FNA) is required for the diagnosis of this rare variety of non-Hodgkin's lymphoma [4, 5]. Surgery is often

Table 1: Differences between primary pancreatic lymphoma and pancreatic adenocarcinoma

	Primary Pancreatic Lymphoma	Primary Pancreatic Adenocarcinoma
Origin	Non epithelial tumor arising from lymphatic tissues	Arises from epithelial cell
Clinical presentation	Obstructive jaundice is an uncommon feature	Obstructive jaundice is a common feature
Diagnosis	Bulky localized tumor mass without marked pancreatic ductal dilatation on imaging is suggestive	Typically presents with marked pancreatic ductal dilatation on imaging
Treatment	Potentially curable with chemotherapy primarily. Role of surgery and radiotherapy are controversial	Treatment primarily by surgical resection. Radiotherapy and chemotherapy may play adjuvant roles
Prognosis	Very good	Poor

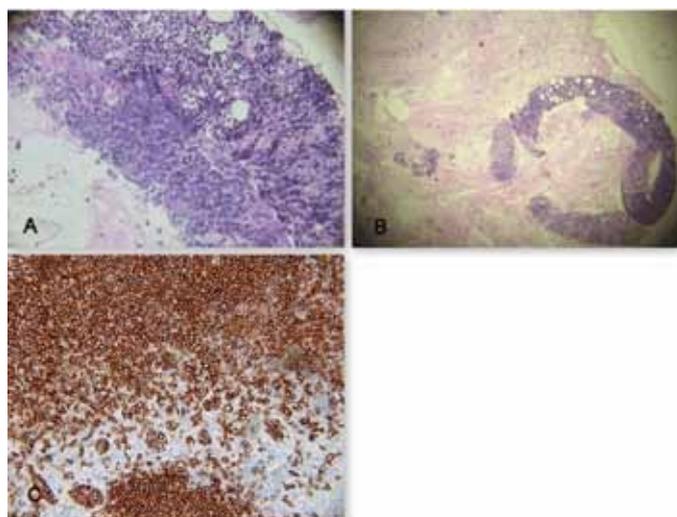


Figure 2: Photomicrograph slides of pancreatic tissue in primary pancreatic lymphoma, (A, B): H&E stain of FNA sample of pancreatic tissue under high and low power view, (C) CD 20 positive B-cells.

reserved for the rare instance when percutaneous or endoscopic biopsies are not diagnostic, or to bypass biliary obstruction [1]. Lactate dehydrogenase (LDH) and beta-2 microglobulin levels might be helpful tools in its differentiation from other common pancreatic cancers. While pancreatic adenocarcinoma is often associated with a grave prognosis, PPL is potentially treatable [1, 2]. The cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) based regimen is the most commonly used therapy. Addition of rituximab, given CD20 positivity to this regimen has been reported to increase complete response rate and prolong both overall and event free survival in recent years [1]. Doxorubicin was not used in the index case to prevent risk of therapy related cardiotoxicity given the patient's age at presentation. This report highlights the need for a heightened clinician awareness of PPL. Though clinically difficult to differentiate from pancreatic adenocarcinoma, its potential curability with therapy makes accurate diagnosis worthwhile. The role of surgery and radiotherapy in treatment remains controversial [1, 4, 5].

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Amara Okoli – Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

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### Guarantor

The corresponding author is the guarantor of submission.

### Conflict of Interest

Authors declare no conflict of interest.

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