Large pancreatic mass with chylous ascites

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ABSTRACT

Primary pancreatic lymphoma (PPL) is a rare entity, most likely to be clinically misdiagnosed as pancreatic adenocarcinoma. The cure rate of PPL is higher than that of adenocarcinoma. We present a case of PPL that presented as a pancreatic mass with chylous ascites and describe the incidence, clinical features, diagnostic approach, and most commonly used treatment regimens for PPL.

KEYWORDS Chemotherapy; lymphoma; pancreatic

Primarily pancreatic lymphoma (PPL) is rare and mimics pancreatic head cancer. We describe a man who presented with a pancreatic mass and chylous ascites and was found to have PPL.

CASE REPORT

A 69-year-old man presented with a 2-month history of reduced appetite, abdominal pain, and distension in the absence of fever, chills, weight loss, or change in bowel habits. He was a nonsmoker, denied alcohol use, and had a non-contributory past medical history as well as no family history of cancer. A farmer, he reported exposure to Agent Orange many years earlier. His abdomen was soft, distended, and nontender with fluid thrill and positive shifting dullness. The only abnormal laboratory test was a mildly elevated lactate dehydrogenase of 254 U/L (normal range 122–225 U/L). Abdominal computed tomography (CT) showed a 20 × 10 × 17 cm soft tissue mass impinging the distal duodenum, replacing the proximal pancreatic parenchyma, and encasing the celiac and superior mesenteric arteries; ascites, lymphadenopathy, and multiple omental nodules were seen (*Figure 1*). No cirrhosis or hepatomegaly was noted.

A bedside paracentesis drained 2.5 L of chylous fluid. The fluid analysis revealed a lymphocytic-predominant cell count with a high triglyceride level of 980 mg/dL; the cytology was negative for malignancy. Acid-fast bacilli cultures and QuantiFERON tests were negative; thus, tuberculosis was ruled out. Endoscopic-guided ultrasound revealed

changes suggestive of mild to moderate chronic pancreatitis and an irregular, hypoechoic pancreatic mass.

Fine-needle aspiration of the mass was performed. Microscopic examination revealed numerous small and large lymphoid cells, with irregular nuclear membrane contours, in a background of pancreatic acinar cells. Immunohistochemical stains showed lymphoid cells strongly positive for CD20 and a few scattered cells stained with CD5 (*Figure 2*). These findings were consistent with possible B-cell lymphoma. A positron emission tomography-CT scan showed the pancreatic mass associated with metabolically active inguinal, aortocaval, juxtadiaphragmatic, and inframammary lymphadenopathy (*Figure 3a*). The low-level fluorodeoxyglucose activity of ascites was suggestive of malignant ascites. The patient underwent a diagnostic laparoscopic biopsy of the mass confirming the diagnosis of B-cell lymphoma of germinal center origin positive for CD10, BCL6, BCL2, and CD45. Fluorescence in situ hybridization analysis showed no evidence of double- or triple-hit lymphoma. The patient received six cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) and tolerated it well. A follow-up positron emission tomography-CT scan showed that the pancreatic mass reduced in size to 10 × 5 cm without areas of new or worsening lymphoma (*Figure 3b*).

DISCUSSION

PPL is very rare and usually mimics carcinoma of the pancreatic head. Extranodal non-Hodgkin's lymphoma (NHL) constitutes 30% to 40% of all NHL cases.¹ PPL comprises <2% of

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Figure 1. Axial view of CT abdomen demonstrating the large pancreatic mass.

Figure 2. Staining of pancreatic B-cell lymphoma: (a) hematoxylin and eosin; (b) CD20.

Figure 3. Positron emission tomography scans (a) before and (b) after chemotherapy.

extranodal malignant lymphomas and 0.5% of all pancreatic masses.¹ The diagnostic criteria for PPL include a dominant pancreatic mass without superficial or mediastinal lymphadenopathy on imaging, absent hepatosplenic involvement and with a normal blood leukocyte count.²

More than 80% of the cases of PPL have a predominant involvement of the pancreatic head. Two different morphological patterns of pancreatic involvement have been described: a localized, well-circumscribed tumor pattern and a diffusely enlarged pattern, as in the current case. The clinical presentation of PPL is very nonspecific without classic B symptoms of nodal NHL like fever, chills, and night sweats. Abdominal pain is the most common symptom (83%), followed by abdominal mass and weight loss.³ Other clinical presentations include jaundice, acute pancreatitis, and small bowel obstruction; on rare occasions, PPL can present with recurrent hypoglycemic episodes due to destruction of pancreatic tissue by tumor infiltration.^{3,4} The laboratory tests for PPL are also nonspecific. High tumor burden, beta-2-microglobulin levels >2 mg/L, and high lactate dehydrogenase levels are poor prognostic markers. In our case, the tumor markers were normal and lactate dehydrogenase was elevated.

Different imaging modalities such as transabdominal ultrasonography, CT, and magnetic resonance imaging can be used for tumor localization. Identification of certain radiological findings like a bulky, localized pancreatic head tumor without Wirsung duct dilatation, invasive and infiltrating growth involving the retroperitoneum or upper abdominal organs, and enlarged lymph nodes below the level of the renal veins help point toward PPL rather than pancreatic adenocarcinoma.⁵ Although endoscopic ultrasound can find morphologic differences between PPL and pancreatic adenocarcinoma, differentiating the two based solely on imaging characteristics is difficult, and tissue needs to be obtained to confirm the diagnosis. With the advent of flow cytometry and immunocytochemistry, which are independent of tissue architecture and cytomorphology, there has been an increase in the diagnostic yield with fine-needle aspiration in cases of lymphoma.⁶ Sampling with a core needle is also possible using endoscopic ultrasound.⁷

The different treatment options for pancreatic lymphoma include surgery, chemotherapy, and radiation, alone or in combination. Most patients can be managed without surgical intervention, and long-term remission can be obtained with chemotherapy alone, as occurred in our case.³ The most common regimen is CHOP, and the addition of rituximab, a chimeric monoclonal antibody against CD20 B-cell antigen, has shown overall survival improvement in patients with diffuse large B-cell lymphoma. A cure rate of up to 30% has been reported in patients with PPL using these treatment options.⁸

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