Diffuse Large B-Cell Lymphoma Presenting with Obstructive Jaundice from a Biliary Stricture

Mark Friedman, MD, Ishwaria Mohan, MD, Gregg Brodsky, MD, Vinay Sood, DO
Division of Gastroenterology and Liver Diseases, Albany Medical College and Center, Albany, New York

Abstract

A 34-year-old woman presented with one month of progressive right upper quadrant (RUQ) abdominal pain and one week of jaundice, reported dark urine and light brown stools. The patient had no known medical problems, denied any substance use. Her family history was unremarkable.

Physical examination revealed scleral icterus, jaundice, and tenderness in the right upper quadrant (RUQ) of the abdomen without hepatosplenomegaly. Notable laboratory values were a total bilirubin of 6.3 (normal: 0.1-1.2 mg/dL), a direct bilirubin of 3.8 (normal: 0.0-0.3 mg/dL), an alkaline phosphatase (AP) of 317 (normal: 30-115 U/L), an AST of 86 (normal: 5-45 U/L), and an ALT of 159 (normal: 5-40 U/L). A viral hepatitis panel and CA19-9 were negative.

Endoscopic ultrasound (EUS) showed a 32 mm x 28 mm heterogeneous mass in the dilatation (Figure 2). Also seen was a right adrenal mass and left lower lobe lung mass; in the pancreatic head measuring 3.5 cm in diameter. Mild pancreatic duct dilatation was noted. Total bilirubin was 0.8, direct bilirubin was 0.2, AP was 110, AST was 25, and ALT was 20. Laboratory data was remarkable for an amylase of 150 IU/L and a lipase of 254 IU/L. On physical exam, the patient had mild midepigastric tenderness but no scleral icterus or jaundice. The presenting symptoms of pancreatic lymphoma are usually non-specific and include abdominal pain (43%), abdominal mass (33%), jaundice (37%), acute pancreatitis (12%), small bowel obstruction (12%) and diarrhea (12%)

Imaging and ERCP/EUS Results

Figure 1: ERCP showing a dilated distal and proximal common bile duct with proximal CBD dilation (Figure 1). A 10 mm biliary sphincterotomy and dilation of the CBD stent with the 6 mm balloon was performed. The CBD stricture was brush for cytology which showed benign ductal cells. The CBD biopsy fragments of benign fibrous tissue and glandular epithelium.

Figure 2: CT scan of the abdomen with biliary stent placement: heterogeneous pancreatic mass measuring 3.5 cm with extrapancreatic extension, mild cephalad displacement and mild pancreatic duct dilatation.

Figure 3: Endoscopic ultrasonography (EUS) showed a 32 mm x 28 mm heterogeneous mass in the pancreatic head (Figure 3); an extrinsic deformity in the duodenum which was biopsied. FNA of the pancreatic mass showed atypical large cells suspicious for large cell lymphoma. Duodenal biopsy showed diffuse large b-cell lymphoma involving the small bowel. Cells from both samplings were strongly positive for BCL-6 and negative for BCL-2, CD10, CD14 and MUM-1. CD20 confirmed the B-cell lineage of the malignancy. A bone marrow biopsy showed no evidence of marrow involvement. The patient was started on induction chemotherapy with rituximab-CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisolone) and is now in clinical remission.

Discussion

Malignant lymphoma infrequently involves the pancreas, accounting for less than 1% of NHL cases and between 0.2-4.9% of all pancreatic tumors (6, 7).

The presenting symptoms of pancreatic lymphoma are usually non-specific and include abdominal pain (43%), abdominal mass (33%), jaundice (37%), acute pancreatitis (12%), small bowel obstruction (12%) and diarrhea (12%).

More commonly seen gastrointestinal sites of DLBCL are duodenal- pancreas- and rectum: 10. These patients typically present with a variety of symptoms including watery diarrhea and severe weight loss.

Imaging plays an important role in the diagnosis and staging of pancreatic masses. This is particularly true for pancreatic lymphoma, as treatment and prognosis are significantly different from those for pancreatic adenocarcinoma. A CT scan is the modality commonly used for the detection of pancreatic lymphoma. Ultrasound- or CT-guided fine needle biopsy of the pancreatic mass can also help distinguish pancreatic lymphoma from pancreatic adenocarcinoma.

Definitively diagnosing primary pancreatic lymphoma without a tissue diagnosis is difficult since the clinical signs and symptoms are remarkably similar to those of pancreatic ductal adenocarcinoma. The cornerstone of DLBCL treatment remains chemotherapy with or without radiotherapy. The role of surgery in the management of DLBCL is evolving. About 40% of DLBCL patients present with localized disease that can be contained in one radiation field (14).

With the advent of the ERCP, obstructive lesions are being increasingly managed with biliary balloon dilatation and stenting rather than a radical procedure to resect the mass.

Background

Involvement of the pancreas by NHL has infrequently been reported, and rarely does DLBCL present as obstructive jaundice from a biliary stricture. The clinical signs and symptoms are remarkably similar to those of pancreatic ductal adenocarcinoma.

This is an unusual case report of DLBCL presenting as obstructive jaundice from a biliary stricture. This case is also atypical because initial CBD brushings and abdominal imaging were negative for any malignancy or lesions. However, within 8 weeks, the lymphoma had progressed to a 3.5 cm lesion with extension into the duodenum.

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