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CASE REPORT | PANCREAS

Primary Anaplastic Large Cell Lymphoma of the Pancreas

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ABSTRACT

Anaplastic large cell lymphoma is an exceedingly rare subtype of non-Hodgkin lymphoma with fewer than 10 cases reported in the literature. We present a 64-year-old woman who presented with a pancreatic head mass, causing acute pancreatitis, which was eventually diagnosed as an anaplastic large cell lymphoma. The disease is very responsive to chemotherapy, further highlighting the importance of early recognition and treatment.

INTRODUCTION

Pancreatic malignancies are the 12th most common cause of cancer in the United States, but the fourth leading cause of cancer deaths.[1] Because 85% of pancreatic malignancies are comprised of adenocarcinoma, physicians are generally less familiar with other types of pancreatic neoplasms.[2,3] We report a case of primary pancreatic anaplastic large cell lymphoma (ALCL), a rare subtype of non-Hodgkin lymphoma, and one of the subtypes of T-cell lymphoma, presenting as a pancreatic mass causing acute pancreatitis. Recognition of ALCL is important because the disease is highly sensitive to chemotherapy and thus carries a very good prognosis.[4]

CASE REPORT

A 64-year-old woman presented to the emergency department with 1 week of abdominal pain radiating to the back and with nausea. Physical examination revealed right upper quadrant abdominal tenderness. Laboratory examinations were significant for alanine transaminase 271 U/L, aspartate transaminase 234 U/L, alkaline phosphatase 706 U/L, total bilirubin 2.5 mg/dL, and lipase 115 U/L. Computed tomography (CT) demonstrated pancreatic head edema with a poorly defined area of hypoenhancement within the head. Magnetic resonance imaging showed a 1.5-cm stricture in the mid-distal common bile duct (CBD) and a 1.6-cm area of increased signal focus, concerning for possible mass or pancreatitis in the pancreatic head. Esophagogastroduodenoscopy was performed, which revealed a 5-mm gastric body polyp with superficial erosion (Figure 1).

Endoscopic ultrasound revealed a 2-cm hypoechoic pancreatic head mass with invasion in the mid-CBD and the portal vein (Figure 2). Biopsies from the gastric body polyp and pancreatic head mass resulted in an anaplastic lymphoma kinase (ALK)-positive ALCL, suggestive of a primary pancreatic lymphoma with metastasis to the stomach (Figure 3). She underwent endoscopic retrograde cholangiopancreatography, which confirmed stricture of the mid-CBD on fluoroscopy, brushings obtained were positive for malignant cells. She was decompressed using a metallic stent. The patient was started on a cytarabine, hydroxydaunorubicin, vincristine, and prednisone (CHOP) regimen with the addition of pegfilgrastim with a significant reduction in both the size of the pancreatic head and gastric masses on repeat CT abdominal imaging at 6 months postdiagnosis. She continues to follow with oncology after recent completion of CHOP and is scheduled to undergo repeat imaging soon.

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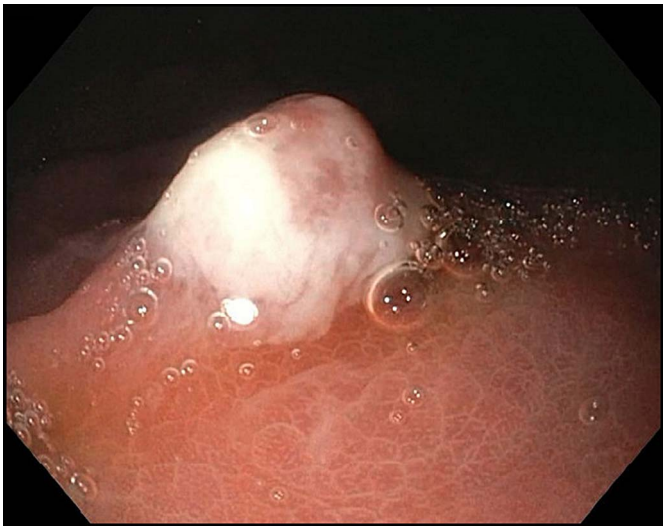

Figure 1. Esophagogastroduodenoscopy showed a gastric polyp that was positive for an anaplastic lymphoma kinase-positive anaplastic large cell lymphoma.

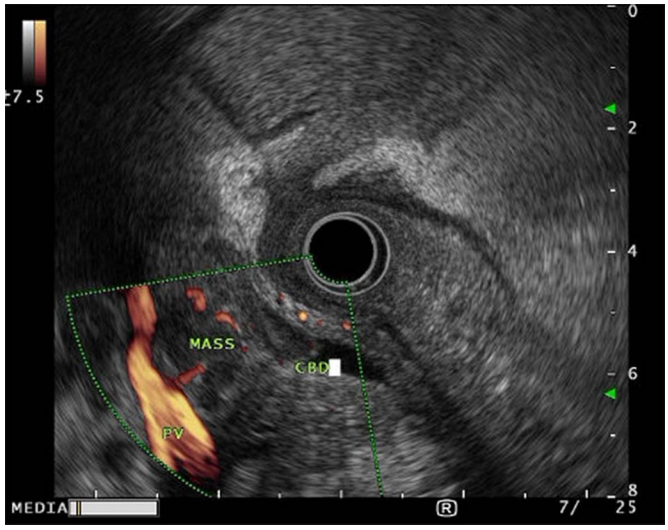

Figure 2. Endoscopic ultrasound showed a pancreatic anaplastic large cell lymphoma invading the portal vein and the mid common bile duct.

DISCUSSION

Although most pancreatic tumors are due to adenocarcinoma, a very small proportion (0.5%) is due to primary pancreatic lymphomas. Of these, at least 80% of primary pancreatic lymphomas are due to the diffuse large B cell lymphoma subtype.[3] ALCL, which is a T-cell subtype of non-Hodgkin lymphoma, is exceedingly rare, with only 8 other cases of primary pancreatic ALCL reported in the literature to date.[4–11] Most of the information on ALCL is drawn from the extragastrointestinal literature, and as noted in the case presented, the pancreatic form of ALCL tends to follow this clinical morphology. ALCL can be further subdivided into ALK-positive and ALK-negative tumors, a distinction which has implications for prognosis, with a 5-year overall survival ranging from 70% to 90% for ALK-positive vs 40% to 60% for ALK-negative ALCL. It has a male predominance (3 men for every 1.2 women), with age of onset occurring younger in ALK-positive (0–30 years) and older in ALK-negative (40–65 years).[12]

Our case is unique in that it represents the only documented case in which an older patient presented with an ALK-positive subtype. Multiple pathogenetic mechanisms for the disease have been identified, with one of the most common having to do with t(2;5) translocation, leading to unregulated production of ALK in lymphoid cells.[5] Although most ALCL tends to present with the classic B symptoms of fever, weight loss, and night sweats, it should be noted that ALCL of the pancreas appears to present with gastrointestinal-related symptoms such as abdominal pain and vomiting.[12] Laboratory results are heterogeneous, but usually demonstrate some combination of elevated aspartate transaminase and alanine transaminase with bilirubin and alkaline phosphatase.

Radiologic imaging via CT or magnetic resonance imaging is nonspecific and usually demonstrates a large mass in the pancreas, most often noted in the pancreatic head in 7 of the 9 cases.[3] In one case, a patient underwent a Whipple based on radiologic findings alone, before the histopathologic diagnosis of ALCL was performed, further underscoring the poor specificity of radiologic imaging and large similarity between different pancreatic malignancies on imaging.[1,4] Diagnosis by histopathology demonstrates CD-30-positive T-cells with prominent cytoplasm and nuclei, and classic "hallmark cells" with horseshoe or kidney-shaped nucleus with prominent eosinophilic cytoplasm.[5,12] Although surgery is sometimes performed for tumor debulking or management of complications of tumor invasion, the definitive treatment for ALCL is chemotherapy. Patients with ALK-positive disease usually respond positively to a CHOP regimen, whereas patients with ALK-negative disease sometimes will require higher doses of chemotherapy, followed by stem cell transplantation.[12] As demonstrated, most patients experience remission with therapy, again highlighting the importance of early detection and diagnosis of this disease.

DISCLOSURES

Author contributions: B. Hughes wrote the manuscript and is the article guarantor. N. Habib edited the manuscript and reviewed the literature. K. Chuang edited the manuscript.

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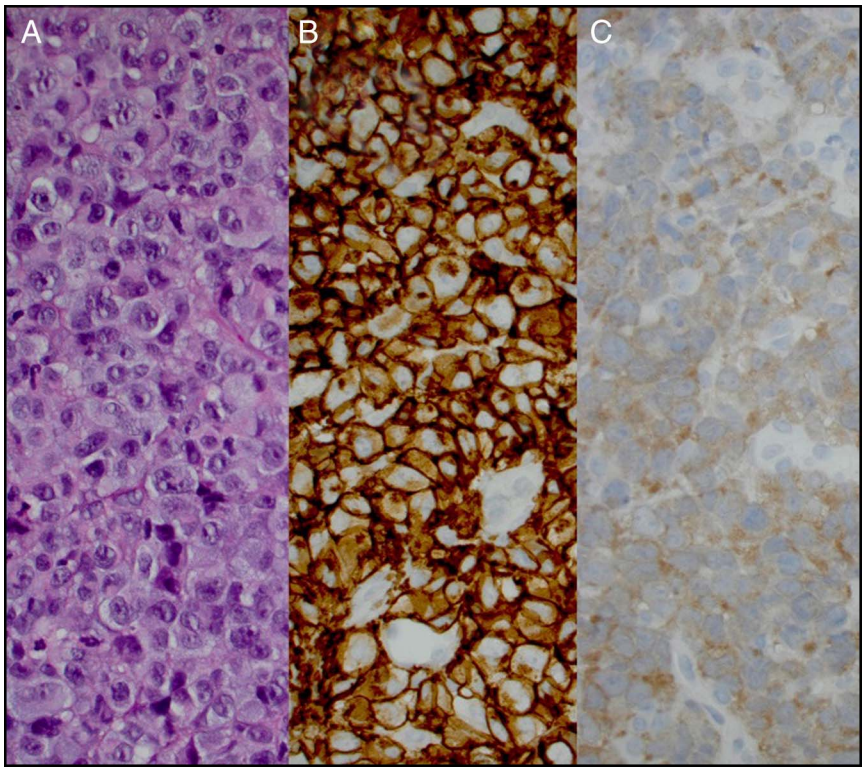

Figure 3. Anaplastic lymphoma kinase-positive anaplastic large cell lymphoma involving the stomach and pancreas. (A) Hematoxylin & eosin stain with lymphoma cells exhibiting dense eosinophilic cytoplasm and large nuclei, (B) CD-30-positive stain, (C) and anaplastic lymphoma kinase-positive stain.

Informed consent was obtained for this case report.

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