Acute Pancreatitis: A Rare Case of Primary Peripheral T-Cell Lymphoma-Not Otherwise Specified

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Abstract

Primary pancreatic lymphoma (PPL) represents a rare extranodal lymphoid malignancy, accounting for less than 2% of all lymphomas, 0.1% of malignant lymphomas, and 0.5% of pancreatic tumors. Among its histological subtypes, peripheral T-cell lymphoma (PTCL), particularly PTCL-not otherwise specified (PTCL-NOS), stands out as a rare, highly aggressive variant with a dismal prognosis. PTCL-NOS frequently presents in an extranodal manner, with the pancreas being a scarce site. Symptoms of pancreatic lymphoma, such as those seen in pancreatitis, are heterogeneous and can closely mimic other pancreatic conditions, including adenocarcinoma and autoimmune disorders, complicating the diagnostic process. This case report features a 35-year-old female with recent episodes of pancreatitis, jaundice, and intermittent pain in the epigastric and left upper quadrant abdominal areas. Radiological assessments uncovered three masses in the pancreas’s head (30\times36 mm), neck (21\times25 mm), body (29\times24 mm) dilatation of the pancreatic and common bile ducts, collapse of the splenic and portal veins, and celiac and peripancreatic lymphadenopathy. An open biopsy identified the tumor as PTCL-NOS. Immunohistochemical analysis showed positive staining for CD3, CD4, CD8, CD10, CD45, PD-1, GATA3, and Ki67 (60% of tumoral cells) but negative for CD20, CD30, CD34, TdT, and ALK. Despite six cycles of cyclophosphamide, hydroxydaunorubicin, oncovin, and prednisone (CHOP) chemotherapy, the patient demonstrated central nervous system (CNS) involvement at follow-up. Overall PPL poses significant diagnostic challenges due to its non-specific clinical presentation. Given their varied prognoses and treatment approaches, histopathological examination is vital for distinguishing between clinicopathologic subtypes. With PTCL-NOS, there is a critical need for neurological evaluations to monitor potential CNS involvement and guide appropriate follow-up care.

Keywords: Adenocarcinoma, Autoimmune, Pancreatic lymphoma, Pancreatitis, Primary lymphoma, T-cell lymphoma-not otherwise specified

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