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Primary pancreas extranodal NK/T cell lymphoma

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An 80-year-old woman admitted to our department presenting with abdominal cramp pain on epigastrium for 3 days. Laboratory studies showed elevation of total bilirubin 1.56 mg/dL, AST 1561 IU/L, ALT 725 IU/L, ALP 580 U/L, GGT 493 IU/L, amylase 209 IU/L, lipase 697 IU/L, CEA 6.82 ng/mL and CA 19-9 45 U/ml. An abdominal CT showed diffuse enlargement of the pancreas with peripancreatic haziness and a 5.5-cm, ill-defined heterogenous mass, with a central necrosis in the pancreatic head. A MRI using axial dynamic fat-saturated T1-weighted images obtained a diffuse homogeneous less enhancing mass, 5.5-cm diameter, in the pancreatic head with upstream minimally pancreatic duct dilatation and common bile duct dilatation. Endoscopic ultrasound (EUS) examination revealed an irregular mixed echoic mass at the head of pancreas. Peri-portal lymph node (hypoechoic, 10mm diameter) was seen. After contrast injection with Sonazoid, the mass showed early arterial enhancement with late washout. EUS guided fine needle biopsy (FNB) was performed and visible whitish core specimens were obtained. Immunohistochemical staining showed LCA (+), CD3 (+), CD45RO (+), CD 56 (+) but negative for CK-Pan, EMA, synaptophysin, chromogranan-A, CD 20, CD 30, CD 99, TdT. EB virus-encoded RNAs (EBER) show positive results on in situ hybridization. These findings were consistent with extranodal NK/T-cell lymphoma. There was no tumor on paranasal sinus MRI. Subsequent bone mass aspirate and biopsy also showed no involvement. Therefore, primary pancreas extra-nodal NK/T cell lymphoma was confirmed based on radiologic and histologic findings. The patients underwent one cycle cisplatin chemotherapy for CCRT. But, the patient rapidly evolved into irreversible multi-organ failure and expired at day after diagnosis. Primary pancreas NK/T-cell lymphoma (PPNTL) is an extremely rare and rapid progression with only few published case in the literature. EUS-FNB using core needle may be crucial diagnostic modality. Acquisition of core specimens followed by IHC staining and evaluation of the preserved tissue architecture may aid in diagnosis. To date, the optimal treatment is no established.