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Cystic lymphangioma of the pancreas

A 46-year old woman was referred for a pancreatic cystic mass found incidentally on abdominal CT scan performed for the evaluation of microscopic hematuria. The patient was asymptomatic and her laboratory tests including a complete blood count, comprehensive metabolic panel, amylase, lipase, CEA, and CA 19-9 were normal. Computed tomography of the abdomen revealed a 6 cm multi-lobed, cystic mass in the body of the pancreas without wall thickening or enhancement (Figure A). An endoscopic ultrasound demonstrated a 5.9 x 2.5 cm multi-loculated, thin walled, anechoic lesion in the body of the pancreas (Figure B). There were no papillary projections. The pancreatic parenchyma and main pancreatic duct appeared normal. There was no peripancreatic or celiac lymphadenopathy. On fine needle aspiration, the lesion was found to contain chylous-appearing fluid (Figure C). Twenty milliliters were collected and sent for cytology and chemical analysis. Fluid CEA was 3.9 ng/ml, amylase was 188 u/l, and triglycerides were 7789 mg/dl. Cytologic evaluation found the fluid to be acellular. A diagnosis of cystic lymphangioma of the pancreas was made and only clinical follow-up was recommended.

Cystic lymphangiomas are rare benign neoplasms considered to be congenital malformations of the lymphatic system. Ninety-five percent of cystic lymphangiomas are located in the head and neck region with less than one percent found in the abdomen (1). Pancreatic cystic lymphangiomas (PCL) are extremely rare lesions with only a few cases reported in literature (2). Pancreatic cystic lymphangiomas are most often solitary lesions
that may be either uni- or multi-locular and contain serous, serosanguineous, or chylous fluid (2-3). Although most PCLs are discovered incidentally, cysts may present with abdominal pain or obstruction of adjacent structures. In addition, complications including hemorrhage, infection, torsion, and rupture have been reported.

The definitive diagnosis of PCL can be made by needle aspiration when the aspirated fluid is grossly chylous and contains a very high triglyceride level. Fluid that is serous or serosanguineous, even in the presence of a mildly elevated triglyceride level is not diagnostic and surgical excision is recommended. In these patients, in whom the diagnosis remains uncertain and in symptomatic patients, surgical resection is curative. If the diagnosis of PCL is made in an asymptomatic patient, further treatment is unnecessary and an acceptable approach is to follow the patient clinically and with periodic imaging.

References:
