Pancreatic Head Mass: Granular Cell Tumor?

Lucas Lambers, MD; Joshua Chemtob, MD; Curtis Gravenmier, MD

Department of Pathology, Orlando Health, Orlando, Florida

Introduction

Granular cell tumor (GCT) is a neuroectodermal neoplasm that commonly arises in the tongue, dermis/subcutis, and breast. The neoplastic cells have abundant eosinophilic cytoplasm containing coarse granules and display infiltrative growth, forming sheets or small nests with a syncytial appearance. GCT is generally positive for S100, CD68 and inhibin, while negative for cytokeratin. PAS with diastase highlights the cytoplasmic granules, which are lysosomes that have accumulated due to loss-of-function mutations in V-ATPase accessory genes involved in endosomal pH regulation.

Rare GCTs occur adjacent to extrahepatic bile ducts or within the pancreas, potentially leading to obstructive jaundice and/or dilatation of the main pancreatic duct. Biomedical imaging shows no characteristic findings and may raise concern for pancreaticobiliary carcinoma. We report of case of GCT arising in the head of the pancreas and associated with abdominal pain.

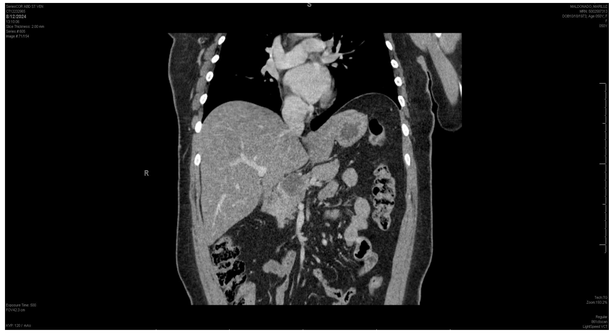

Figure 1. CT with contrast shows a 38 x 26 mm hypodense lesion in the pancreatic head with subtle mural enhancement, although without pancreatic ductal or biliary dilatation.

Case and result

A 50-year-old female presented to the emergency department with right flank pain, at which time abdominal computed tomography noted a 2.6 x 1.8 cm hypodense lesion in her pancreatic head. The main pancreatic duct showed no evidence of obstruction. The lesion appeared stable on imaging obtained during the next two years. Following another emergency room visit for generalized abdominal pain, the patient underwent esophagogastroduodenoscopy with endoscopic ultrasound that identified septate cysts associated with the pancreatic mass, which had grown to 2.9 x 2.7 cm in cross section. Fine needle aspiration was performed and revealed loosely aggregated cells with abundant eosinophilic and granular cytoplasm, small round to ovoid nuclei, and inconspicuous nucleoli. Immunohistochemical stains demonstrated the cells were positive for S100, inhibin, CD56, and synaptophysin (weak) with a low Ki67 proliferation index estimated at less than 1%. PAS with diastase highlighted the cytoplasmic granules in bright pink. The cells were negative for AE1/AE3, chromogranin, PAX8, calretinin, HMB45, and SF1. Beta-catenin showed membranous staining. The patient is currently considering surgical resection of the lesion due to recurrent episodes of severe abdominal pain.

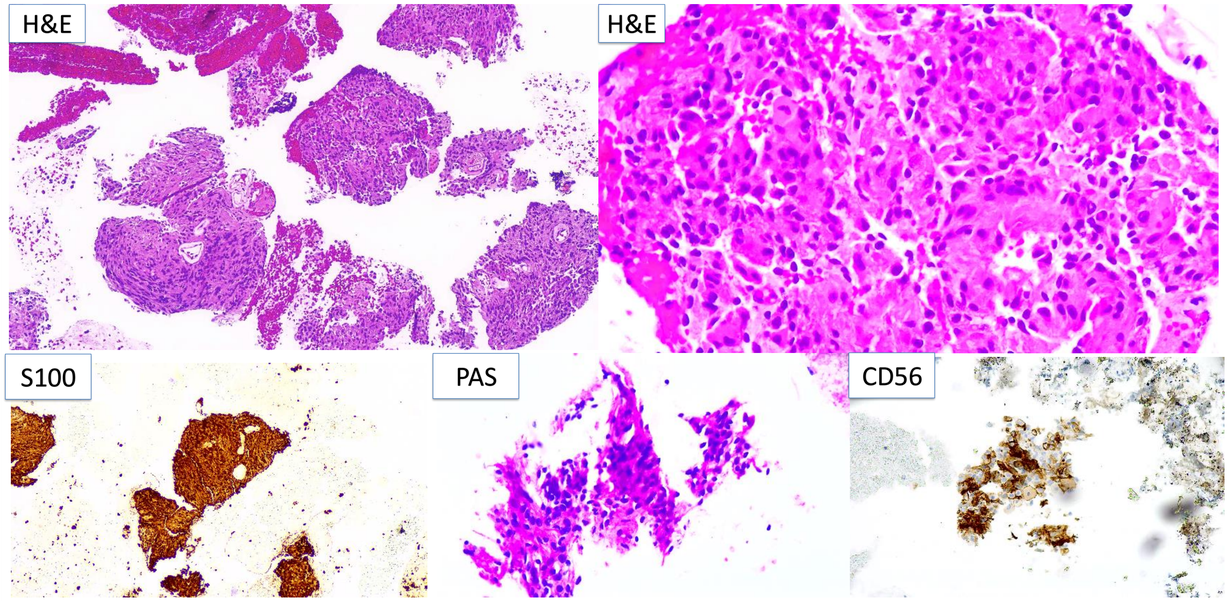

Figure 2. Granular cell tumor. Biopsy of the solid and cystic pancreatic lesion revealed polygonal to slightly spindled cells with coarse cytoplasmic granules and small, round nuclei (H&E, 100X and 400X). The lesional cells were positive for S100, PAS, and CD56 (S100: 100X, PAS: 400X, CD56: 200X).

Discussion

We report a case of GCT occurring in the pancreas of a 50-year-old female. The GCT was associated with abdominal pain and had a mixed solid and cystic appearance by endoscopic ultrasound, raising concern for pancreatic malignancy.

Granular cell tumors are known to occur at a variety of anatomic sites but are uncommon in viscera. Cases of GCT involving the pancreas may show dilatation of the main pancreatic duct and raise concern for pancreatic ductal adenocarcinoma or intraductal papillary mucinous neoplasm. Fine needle aspiration of such lesions could result in misdiagnosis due to reactive atypia of the adjacent ductal epithelium. Some cases are only recognized after resection, but successful diagnosis is possible using cytology material. Keys to cytologic diagnosis include recognition of the characteristic granular cytoplasm and judicious use of ancillary testing. In this case, S100 positivity and a lack of cytokeratin expression helped navigate the pathologist toward this unusual diagnosis.

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