

SECTION EDITOR: ENID GILBERT-BARNES, MD

Pathological Case of the Month

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A 15-YEAR-OLD white girl presented with a history of acute, left upper quadrant abdominal pain that developed while she was performing gymnastics. She had no history of trauma or pancreatitis. An abdominal computed tomographic scan revealed a large mass in the tail of the pancreas. The differential diagnosis based on the computed tomographic scan included pancreatic pseudocyst and papillary cystadenoma. An endoscopic retrograde cholangiogram and pancreatogram showed a J-shaped stomach con-

sistent with extrinsic compression, an inferiorly displaced main pancreatic duct with a smooth, tapered narrowing, and no filling of the pancreatic tail. There was no evidence of communication of the pancreatic duct with the mass. The cholangiogram showed no abnormalities. Exploratory laparotomy showed a large mass involving the pancreatic tail and adherent to the splenic artery and vein. A distal pancreatectomy and splenectomy was performed; the gross specimen showed an 11-cm-diameter mass that markedly distended the pancreatic tail (**Figure 1**). On cut section, the mass had a multicystic, hemorrhagic, and necrotic appearance (**Figure 2**). The tissue was examined histopathologically (**Figure 3**).

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Figure 1.

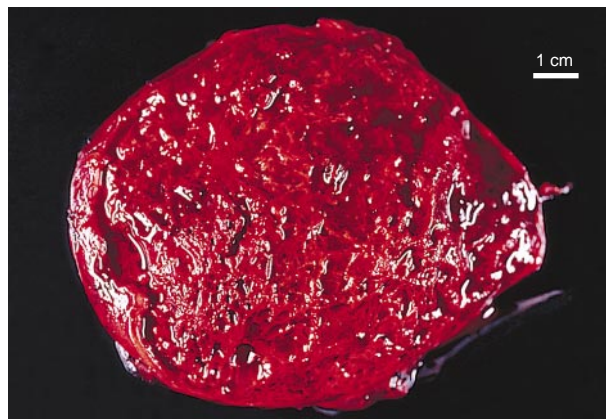


Figure 2.

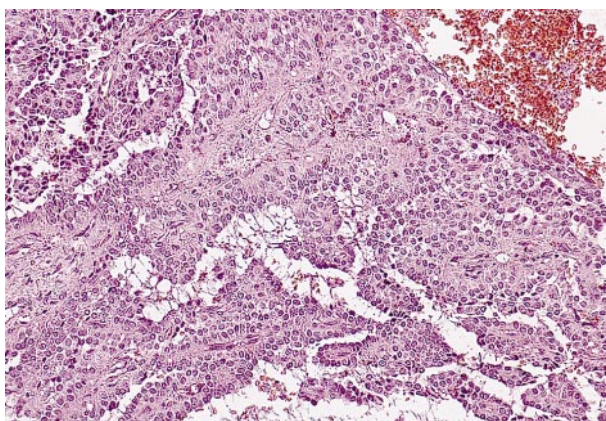


Figure 3.

Diagnosis and Discussion

Solid and Papillary Epithelial Neoplasm

Figure 1. Surgical specimen shows the tail of the pancreas markedly expanded by the tumor and adjacent to the spleen.

Figure 2. Cut section of the tumor has a hemorrhagic and cystic appearance that mimics a pseudocyst.

Figure 3. Microscopic view shows solid sheets of tumor cells admixed with pseudopapillary structures (hematoxylin-eosin, original magnification $\times 100$).

The patient presented with the uncommon finding of a pancreatic mass in an adolescent. Radiologically, the differential diagnoses included pancreatic pseudocyst and pancreatic cystadenoma. Grossly, the lesion consisted of a well-circumscribed mass with a thick fibrous capsule and a diffuse, soft, hemorrhagic center that resembled a pseudocyst; however, the patient lacked a history of trauma or pancreatitis.

Histopathologic examination showed a solid and papillary epithelial neoplasm of the pancreas. These are uncommon tumors, accounting for 1% to 2% of all exocrine pancreatic neoplasms.¹ They predominantly occur in adolescent and young adult women.^{2,3} Although seen in all races, some studies have shown a predisposition in blacks.⁴ Clinically, these tumors usually present as palpable masses, but abdominal pain has also been reported.^{3,4} The onset of pain occasionally correlates with a history of trauma.¹ These large, solitary tumors have no preferential location in the pancreas and rarely cause jaundice even when located in the head of the pancreas.^{1,5-7} Grossly, they often have large zones of hemorrhage and necrosis that mimic a pancreatic pseudocyst.^{1,8}

The tumor derives its name from the microscopic appearance of epithelial-like cells arranged in a variety of patterns. The periphery of the tumor is usually composed of solid, monomorphic areas that give way to pseudopapillary structures associated with more central hemorrhage and cystic degeneration. Individual tumor cells are uniform, polygonal to elongated cells that surround delicate fibrovascular cores. These tumor cells have eosinophilic to vacuolated cytoplasm, nuclei with fine chromatin, and, frequently, nuclear grooves.

A pancreatic neuroendocrine tumor was strongly considered in the microscopic differential diagnosis because of the overlap in architectural patterns and cytologic features. Although uncommon, 1% to 5% of insulinomas occur in the first 2 decades of life. Immunohistochemistry helped make the distinction. The tumor cells stained diffusely positive for vimentin, had patchy but intense positivity for α_1 -antitrypsin, and were focally positive for cytokeratin, which is the characteristic immunohistochemical profile of this tumor.³⁻⁶ The tumor was negative for neuroendocrine markers chromogranin, insulin, glucagon, somatostatin, and gastrin. Interestingly, solid and papillary epithelial neoplasms have shown immunohistochemical and ultrastructural evidence of both neuroendocrine and acinar/ductal differentiation that suggest origin from a multipotential stem cell.³

Most patients with this tumor do well after limited resection; however, cases of metastatic lesions have been reported.^{3,7} It has been suggested that angioinvasion or perineural invasion are indicators of malignant behavior.^{1,7} In this tumor, residual, compressed islets in the fibrous tissue resembled vascular invasion, but this was easily dismissed by the immunohistochemical stains. No perineural invasion was present. Criteria for predicting malignant behavior have not been definitively delineated, and these tumors should probably be regarded as borderline neoplasms even in the absence of aggressive histologic features or metastasis.¹

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Submissions

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