Nonductal Pancreatic Cancers

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KEYWORDS

• Pancreas • Solid pseudopapillary • Acinar • Pancreatoblastoma

Key points

- Nonductal pancreatic neoplasms share common features of cellular tumors with little intervening stroma and show abnormal beta-catenin expression.
- Pancreatoblastomas are the most common tumors with acinar differentiation in childhood, whereas acinar cell carcinomas are the most frequent tumors in adults.
- For differential diagnoses of nonductal pancreatic neoplasms, identifying characteristic histologic features, such as pseudopapillae, acinar architecture with single prominent nucleoli, or squamoid nests, is important.
- Acinar cell (trypsin/chymotrypsin/B-Cell Leukemia/Lymphoma 10 [BCL10]) or neuroendocrine (synaptophysin/chromogranin) markers and E-cadherin are helpful for differential diagnosis of nonductal pancreatic neoplasms and pancreatic neuroendocrine tumors.

ABSTRACT

Nonductal pancreatic neoplasms, including solid pseudopapillary neoplasms, acinar cell carcinomas, and pancreatoblastomas, are uncommon. These entities share overlapping gross, microscopic, and immunohistochemical features, such as well-demarcated solid neoplasms, monotonous cellular tumor cells with little intervening stroma, and abnormal beta-catenin expression. Each tumor also has unique clinicopathologic characteristics with diverse clinical behavior. To differentiate nonductal pancreatic neoplasms, identification of histologic findings, such as pseudopapillae, acinar cell features, and squamoid corpuscles, is important. Immunostainings for acinar cell or neuroendocrine markers are helpful for differential diagnosis. This article describes the clinicopathologic and immunohistochemical features of nonductal pancreatic cancers.

OVERVIEW

The pancreas is mainly composed of 3 types of epithelial cells: enzyme-producing acinar cells (85%), hormone-producing endocrine cells (3% to 5%), and ductal cells (up to 3%).[1] In general, the most common epithelial neoplasm arises from the most common normal epithelial component in an organ. However, in the pancreas, the most prevalent tumors are not acinar cell carcinomas (ACCs) but ductal adenocarcinomas.[2] Because pancreatic neuroendocrine tumors (PanNETs) are discussed elsewhere in this issue (see Salaria SN, Shi C: Pancreatic Neuroendocrine Tumors, in this issue), they are not discussed here. Excluding PanNETs, nonductal pancreatic tumors comprise less than 5% of pancreatic neoplasms.[2] This article discusses existing knowledge of these uncommon nonductal pancreatic neoplasms, including solid pseudopapillary neoplasms (SPNs), ACCs, and pancreatoblastomas (PBs).

Disclosure: This study was supported by a grant (1320200) from the National R&D Program for Cancer Control, Ministry of Health & Welfare, Republic of Korea.

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Surgical Pathology ■ (2016) ■–■
http://dx.doi.org/10.1016/j.path.2016.05.005
1875-9181/16/$ – see front matter

SOLID PSEUDOPAPILLARY NEOPLASM

SPNs are low-grade carcinomas characterized by a solid growth pattern and pseudopapillary formation by poorly cohesive monomorphic epithelial tumors cells with occasional degenerative cystic changes.[3] SPNs are rare, accounting for 1% to 3% of all exocrine pancreatic neoplasms and only 5% of cystic neoplasms.[4] These tumors occur predominantly in young women, with a male to female ratio of 1:9 and a mean patient age of 29 years.[5–7] Forty percent of SPNs are incidentally found without specific symptoms.[5,6] Clinical features for symptomatic patients are usually nonspecific and include abdominal pain and a palpable mass.[5]

GROSS FEATURES

SPNs are well-demarcated or partially encapsulated large single masses with an average size of 8 to 10 cm.[4,8] The tumors occur more frequently in the body or tail of the pancreas than in the head.[5–7] SPNs have variable appearances, including purely solid, mixed solid and cystic, and pure cystic (**Fig. 1**). Most cases are mixed solid and cystic tumors.[3] Small tumors tend to be purely solid, but as tumor size increases they are more likely to be cystic because of the development of degenerative cystic changes.[3] Gross features of completely cystic degenerative SPNs are similar to those of pseudocysts.[8] The cut surface shows variable colors, such as yellow, tan, red-tan, brown, and gray, based on the presence and proportions of accompanying hemorrhagic necrosis and degeneration.[4,8] Calcification is present in approximately 33% of SPN cases.[9,10]

MICROSCOPIC FEATURES

At scanning-power magnification, SPNs show variable growth patterns in combination with solid, pseudopapillary, and pseudocystic structures (**Fig. 2**A). Although the tumors are well demarcated from normal pancreatic parenchyma and are even found partially encapsulated on gross examinations, tumor cells have been found to have infiltrative borders with surrounding normal pancreatic parenchyma microscopically (**Fig. 2**B).[11] In solid

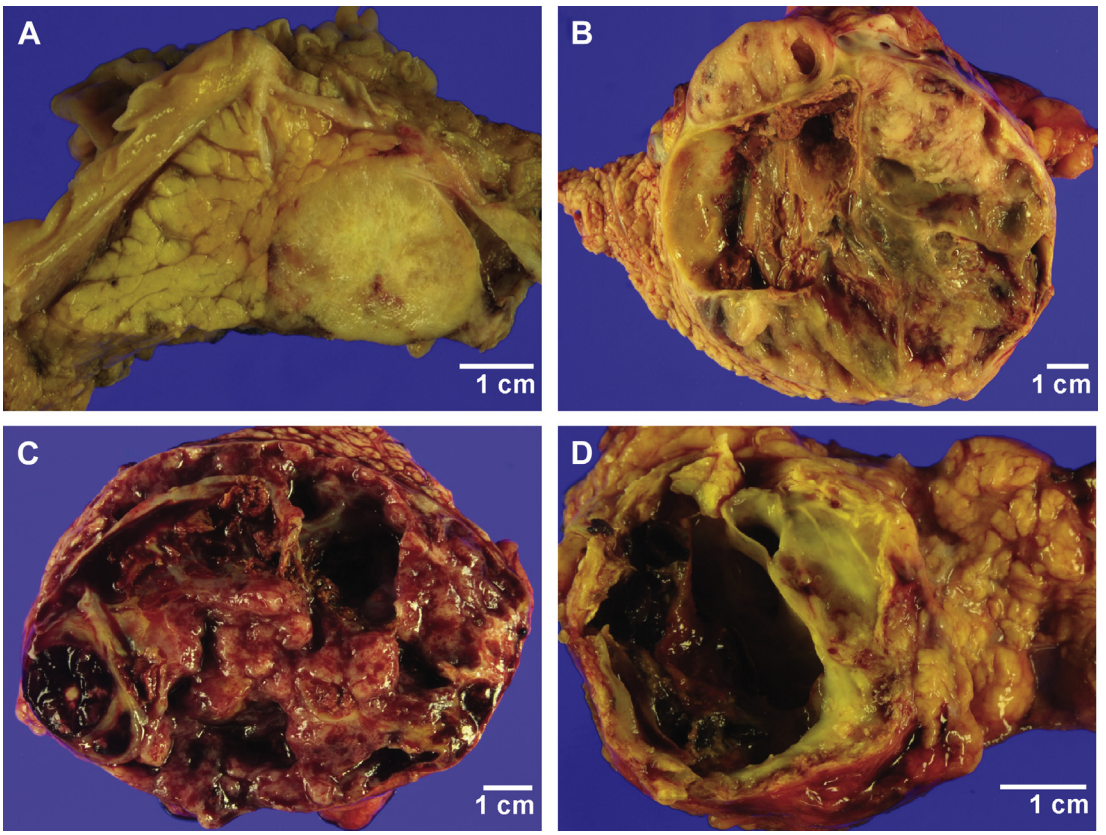

Fig. 1. Solid-pseudopapillary neoplasm showing variable gross appearances of (*A*) pure solid, (*B*, *C*) mixed solid and cystic, and (*D*) pure cystic tumor.