Intraductal neoplasms of the pancreas.

Abstract:
There are three types of pancreatic neoplasms that predominantly have an intraductal growth pattern: the common, usually cystic, intraductal papillary mucinous neoplasms (IPMNs); the rare, usually solid intraductal tubulopapillary neoplasms (ITPNs); and the rare intraductal tubular pyloric gland-type adenoma. In addition to these three tumor types, pancreatic neoplasms with a usually solid growth pattern such as acinar cell carcinomas, neuroendocrine tumors, and undifferentiated carcinomas may present, though very rarely, as predominantly intraductally growing neoplasms. IPMNs can be subclassified into main duct and branch duct tumors; into low- and high-grade dysplasia groups; and into tumors with intestinal, pancreatobiliary, oncocytic, or gastric cellular differentiation. The intestinal-, pancreatobiliary-, and oncocytic-type IPMNs occur predominantly in the main duct of the head of the pancreas and more commonly progress to invasive adenocarcinomas. The gastric-type IPMNs are frequently multifocal, occur predominately in the branch ducts of the uncinate process, and have a low risk of progressing to invasive carcinoma. The prognosis for patients with an IPMN depends largely on the subtype and the presence and the stage of an invasive carcinoma. ITPNs are nodular tumors, often in the pancreatic head, and composed of densely packed tubular glands. Molecular genetics reveal KRAS, GNAS, and RNF43 as the most
frequently mutated genes in IPMNs, while ITPNs show wild-type KRAS. Recent progress in genetic sequencing of pancreatic neoplasms and the identification of specific genetic mutations also holds promise for the future development of novel gene-based diagnostic tests in intraductal neoplasms of the pancreas that might even be used in preoperative conditions.