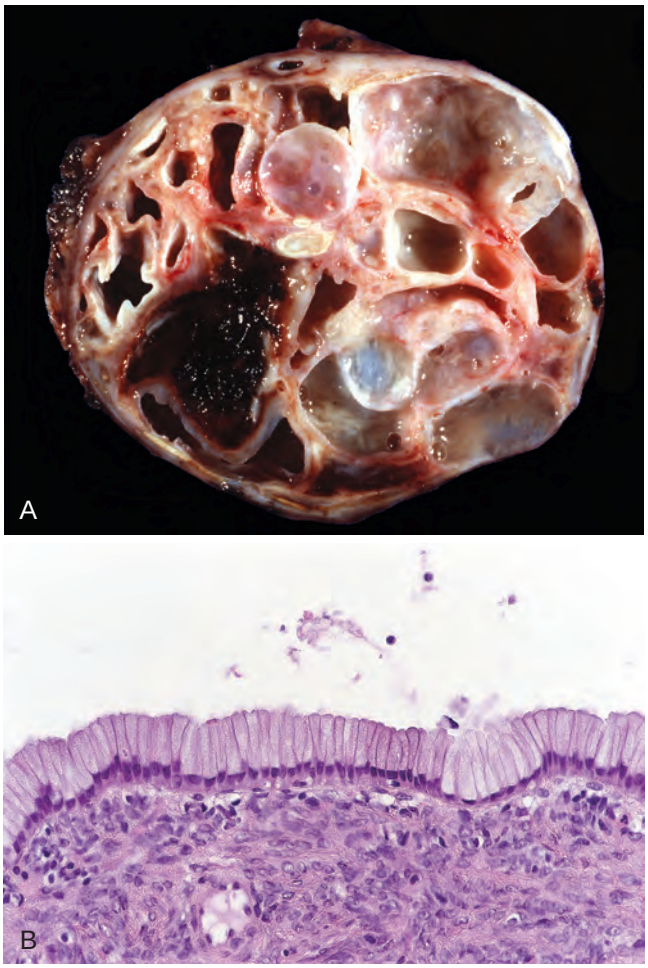

Figure 19-9 Pancreatic mucinous cystic neoplasm with low-grade dysplasia. **A,** Cross-section through a mucinous multiloculated cyst in the tail of the pancreas. The cysts are large and filled with tenacious mucin. **B,** The cysts are lined by columnar mucinous epithelium, and a dense "ovarian" stroma is noted.

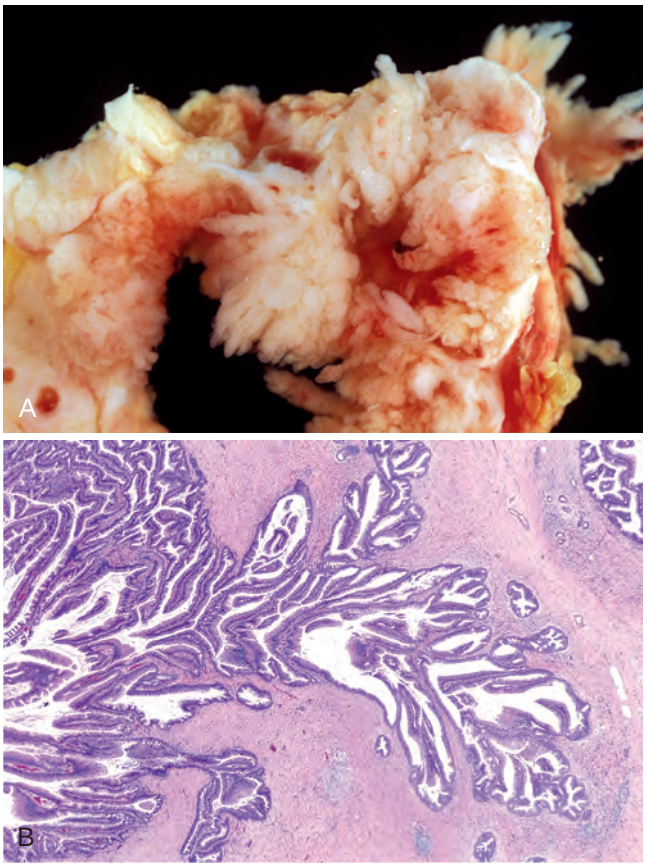

Figure 19-10 Intraductal papillary mucinous neoplasm. **A,** Cross-section through the head of the pancreas showing a prominent papillary neoplasm distending the main pancreatic duct. **B,** The neoplasm involves the main pancreatic duct *(left)* and extends down into the smaller ducts and ductules *(right)*.

can be precursors to invasive carcinomas. These neoplasms usually arise in the tail of the pancreas and present as painless, slow-growing masses. The cystic cavities are larger than those in serous cystic neoplasms. They are filled with thick, tenacious mucin and lined by a columnar mucin-producing epithelium associated with a dense stroma similar to ovarian stroma (Fig. 19-9). Up to one third of surgically resected mucinous cystic neoplasms harbor an associated invasive adenocarcinoma. While surgical resection is curative for noninvasive mucinous cystic neoplasms, half of patients with an invasive carcinoma arising in a mucinous cystic neoplasm will die of their disease. Early detection and treatment before an invasive cancer develops is therefore critical. The *KRAS* oncogene and the *TP53* and *RNF43* tumor suppressor genes are frequently mutated in these neoplasms.

Intraductal papillary mucinous neoplasms (IPMNs) are mucin-producing neoplasms that involve the larger ducts of the pancreas. In contrast to mucinous cystic neoplasms, IPMNs arise more frequently in men than in women, and they involve the head of the pancreas more often than the tail. Ten to twenty percent are multifocal. Two features are useful in distinguishing IPMNs from mucinous cystic neoplasms: (1) absence of the dense "ovarian" stroma seen in mucinous cystic neoplasms and (2) involvement of a pancreatic duct (Fig. 19-10). Just as with mucinous cystic neoplasms, IPMNs can progress to an invasive cancer. Early detection and treatment of IPMNs before they progress to an invasive cancer is therefore critical. Frequent mutations in the *GNAS* and *KRAS* oncogenes and the *TP53*, *SMAD4*, and *RNF43* tumor suppressor genes have been reported in these neoplasms.

The unusual *solid-pseudopapillary neoplasm* is seen mainly in young women. These large, well-circumscribed malignant neoplasms have solid and cystic components filled with hemorrhagic debris. The neoplastic cells grow in solid sheets or, as the name suggests, as pseudopapillary projections, and often appear to be poorly cohesive. These neoplasms often cause abdominal discomfort because of their large size. Of note, this neoplasm is virtually always associated with hyperactivation of the Wnt signaling pathway due to acquired activating mutations of the *CTNNB1* (β-catenin) oncogene. Surgical resection is the treatment of choice. Although some solid-pseudopapillary neoplasms are locally aggressive, most patients are cured following complete surgical resection of the neoplasm.