

Diagnosis and Management of Pancreatic Cystic Neoplasms



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KEYWORDS

- Pancreatic cyst • Intraductal papillary mucinous neoplasm (IPMN)
- Mucinous cystic neoplasm (MCN) • Serous cystadenoma (SCA)
- Endoscopic ultrasound (EUS)

KEY POINTS

- Pancreatic cystic neoplasms are becoming increasingly prevalent due to increased awareness and increased utilization of high-resolution cross-sectional imaging.
- Different types of pancreatic cystic neoplasms vary in risk of current and future malignancy. Accurate diagnosis is, therefore, key to selecting optimal management, which involves either surgical resection or clinical and radiologic surveillance.
- Intraductal papillary mucinous neoplasms (IPMNs) present as 1 of 2 variants, main duct (MD-IPMNs) or branch duct (BD-IPMNs), with differing malignant potential and, hence, management. MD-IPMNs have a higher risk of malignancy and should be referred for surgical resection. BD-IPMNs are more likely to be benign and, particularly in older patients without high-risk features, such as mural nodules, can be observed.
- Mucinous cystic neoplasms (MCNs) represent another type of mucinous tumor with malignant potential. MCNs most often present in middle-aged women, in the body or tail of the pancreas, and should be referred for surgical resection.
- Serous cystadenomas (SCAs) are nonmucinous, benign lesions that often occur in older women and can be observed unless symptomatic or growing.
- Cystic pancreatic endocrine neoplasms (CPENs) are rare, potentially malignant tumors, with characteristic arterially enhancing walls on CT. CPENs should be referred for surgical resection.
- Solid pseudopapillary neoplasms (SPNs) are rare, potentially malignant tumors almost exclusively seen in young women. SPNs should be referred for surgical resection.

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INTRODUCTION

Cystic lesions of the pancreas pose a growing diagnostic and management challenge. Incidental pancreatic cysts are now detected in 2.6% to 13.5% of adults undergoing CT or MRI, with an even higher prevalence, up to 10% to 40%, in those above the age of 80.^{1–3} A vast majority of such cystic lesions do not contain invasive cancer, but many subtypes—IPMNs, MCNs, CPENs, and SPNs—carry a risk of current or future malignancy and, therefore, may require surgical resection.⁴ The morbidity and mortality of pancreatic resection, however, mandate careful risk/benefit analysis of surgery versus surveillance for each individual patient and lesion. The current challenge for oncologists, gastroenterologists, and surgeons is 2-fold. The malignant behavior of pancreatic cysts must be predicted more accurately and then this information used to better stratify patients into surgical versus nonoperative management.

To provide clinical context to this question, this article reviews the most common types of pancreatic cystic neoplasms, detailing each disease's characteristic clinical and radiologic features and recommended management, followed by a generalized approach to diagnosis and treatment. International consensus guidelines⁵ are included where relevant. Attention is paid to controversial diagnostic and treatment questions requiring further investigation.

DISEASE ENTITIES

In light of increased use of high-resolution cross-sectional imaging and heightened awareness of pancreatic cystic neoplasms, pancreatic cysts are diagnosed with increasing frequency, at a smaller size (median diameter 1.6 cm vs 2.4 cm in the most recent vs prior decades), and, increasingly incidentally, now in more than two-thirds of cases.^{6,7} The most common types of resected pancreatic cystic neoplasms include IPMNs (27%–48%), MCNs (11%–23%), SCAs (13%–23%), CPENs (4%–7%), SPNs (2%–5%), and, infrequently, cystic degeneration of pancreatic ductal adenocarcinomas (PDACs) (1%–2%) (**Fig. 1**).^{6–8} Over the past several decades, MCNs and SCAs have decreased in frequency, whereas IPMN has become the most common pathologic diagnosis.^{6,8} Current knowledge about each disease entity's presentation, diagnosis, and management is discussed.

Intraductal Papillary Mucinous Neoplasm

IPMNs have markedly increased in incidence over the past 20 years and are now the most commonly diagnosed and resected type of pancreatic cystic neoplasm.^{4,6,8} From a histologic standpoint, IPMNs are cystic, intraductal, mucin-producing tumors whose dysplastic cells range from benign adenoma to invasive carcinoma (**Fig. 2**).⁴ IPMNs are suspected to represent a different disease entity than PDACs but are thought to progress through a similar pathway of benign to dysplastic to invasive disease.^{9,10} Two variants, MD-IPMNs and BD-IPMNs, are distinguished based on main pancreatic duct involvement, which, in the absence of histology, is defined as ductal dilatation on imaging.⁵ Mixed or combined IPMNs involving both the main pancreatic duct and side branches behave similarly to MD-IPMNs and are often grouped with MD-IPMNs for simplicity.^{11,12} Because MD-IPMNs carry a higher risk of malignancy at presentation and require immediate surgical resection, whereas the more commonly encountered BD-IPMNs are more likely benign lesions that can be managed nonoperatively, it is important to discern the variant of IPMN at the time of diagnosis to help guide management and determine prognosis.⁵

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