Pancreatic Cystic Neoplasms: An Update



Gyanprakash A. Ketwaroo, мр, мsc^a, Koenraad J. Mortele, мр^b, Mandeep S. Sawhney, мр, мs^c,*

KEYWORDS

• Pancreas • Cystic neoplasm • Mucinous • Cyst

KEY POINTS

- Cystic neoplasms of the pancreas have diverse presentations, varying malignant potential, and with the uncertain natural history of some of these lesions, an evidence-based approach to management is limited.
- There is significant potential for improving the differential diagnosis of cystic neoplasms of the pancreas based on the detection of genetic mutations within cyst fluid.
- There are now several guidelines for the management of cystic neoplasms of the pancreas, each with its own limitations.

INTRODUCTION

Pancreatic cystic neoplasms were historically considered a rare subset of pancreatic tumors. However, the incidence of these lesions is rising, in part from detection through the increasing use of high-resolution cross-sectional imaging techniques. The reported prevalence of pancreatic cystic lesions on imaging studies ranges from 2% to 16%, and increases with advancing age. These cystic neoplasms of the pancreas are diverse and can be benign or frankly malignant. Given the rising incidence of cystic pancreatic neoplasms and the demonstrated malignant potential of certain subtypes, accurate diagnosis and multidisciplinary management is paramount.

Initial diagnosis of cystic pancreatic lesions is generally based on imaging characteristics identified on computed tomography (CT) and/or MRI. Endoscopic ultrasound (EUS) provides further imaging characterization, often with increased resolution, and also enables fluid aspiration and analysis to additionally aid differentiation. Cyst fluid

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^a Division of Gastroenterology, Baylor College of Medicine, Houston, TX, USA; ^b Division of Abdominal Imaging, Department of Radiology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA; ^c Division of Gastroenterology, Beth Israel Deaconess Medical Center, Harvard Medical School, Boston, MA, USA

^{*} Corresponding author. 330 Brookline Avenue, Rabb-Rose 101, Boston, MA 02215. *E-mail address:* msawhney@bidmc.harvard.edu

analysis commonly involves biochemical and cytologic characterization, and in certain cases, assessment for genetic mutations. After diagnosis, the general approach to these lesions includes surgical intervention and/or surveillance imaging. Taking into account diverse presentations, varying malignant potential, and the uncertain natural history of some of these lesions, an evidence-based approach is limited. Consensus guidelines by experts attempt to bridge this gap, and research is ongoing. This article discusses recent updates in the diagnosis and management of cystic neoplasms of the pancreas.

TYPES OF PANCREATIC CYSTIC NEOPLASMS Serous Cystadenoma

Serous cystadenomas constitute 1% to 2% of exocrine pancreatic tumors with 80% found in women older than 60 years and are therefore sometimes referred to as the "grandmother" tumors. ⁴ This lesion is considered benign and is typically found incidentally. Occasionally, larger tumors can cause mass effect on surrounding structures, leading to symptoms, such as nausea or abdominal discomfort. ^{4,5}

Serous cystadenomas are comprised of multiple cysts usually measuring less than 2 cm in size and separated by thin septations that are lined by epithelial cells (Figs. 1–3). The appearance is often described as a "cluster of grapes." On cyst fluid analysis, hemosiderin-laden macrophages are seen histologically in 43% of cases. Characteristically, the cyst fluid has low levels of amylase (<250 IU/L), carcinoembryonic antigen (CEA; <5 ng/mL), and serum carbohydrate-associated antigen 19.9 (CA-19.9; <37 U/mL).

Recent research into serous cystadenomas has focused on their pathogenesis. Clinical case series have suggested an association with von Hippel-Lindau disease, which may implicate a mutation in this gene.⁹

Given the benign nature of the serous cystadenoma, no further follow-up is needed for small cysts once a diagnosis has been made. Larger cysts may demonstrate an increased rate of growth (approximately 2 cm per year), and should be followed



Fig. 1. A 5-cm cyst, composed mostly of microcysts with a honeycombed appearance, is shown on EUS. A few larger 4- to 8-mm cysts are also seen. This appearance is characteristic for a serous cystadenoma.

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