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REVIEW

Diagnosis and Treatment of Cystic Pancreatic Tumors

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Cystic pancreatic tumors (CPTs) have more frequently been identified in the last decade because of increased use of cross-sectional abdominal imaging. Although serous CPTs follow an indolent course and do not necessarily require surgical resection or long-term follow-up, mucinous CPTs (mucinous cystic neoplasms and intraductal papillary mucinous neoplasms) have a greater risk for malignancy. Although most CPTs are initially detected with imaging modalities such as computed tomography or magnetic resonance imaging, these tests alone rarely permit an accurate clinical diagnosis. Endoscopic ultrasound and endoscopic ultrasound-guided, fine-needle aspiration allow real-time examination and biopsy analysis of CPTs, which increases diagnostic accuracy because cytopathology features and tumor markers in cyst fluid can be analyzed. Management of patients with mucinous CPTs by surgery or imaging surveillance is controversial, partially because of limited information about disease progression and the complexities of surgical resection. We review approaches to diagnosis and management of common CPTs.

Keywords: EUS-FNA; Pancreas; Tumor; Neoplasm.

Pancreatic cysts are increasingly encountered in clinical practice. Pseudocysts or acute fluid collections complicate acute or chronic pancreatitis and have no potential for malignant transformation. Recognition and management of these will not be addressed in this review. The remaining cysts, referred to in this review as cystic pancreatic tumors (CPTs), represent a distinct group of benign, premalignant or malignant tumors that require further evaluation and management. Because of their variable presentation and malignancy risk, as well as the limited information typically deduced from imaging studies alone, clinicians may face difficulties in diagnosing and treating these tumors.

The purpose of this review is to provide an overview of the diagnosis and management of various types of commonly encountered CPTs. Thus, a detailed literature review that identified key papers published in the last 14 years was conducted and referenced herein.

Epidemiology

Although the exact prevalence of CPTs is unknown, it was previously estimated at 1% of the general population based on previous large-scale observational imaging studies[1] and up to 24% based on autopsy studies.[2] However, recent magnetic resonance imaging (MRI)[3,4] and computed tomography (CT) studies[5] indicate a prevalence ranging between 2.4% and 14%. These studies also suggest that pancreatic cysts occur equally in males and females and that prevalence increases with age. It is unclear if this increased detection represents a true rise in incidence or is merely a reflection of the frequent use of high resolution imaging.

Pathological Classification and Malignancy Risk

Pathologically, pancreatic cystic lesions can be classified by the presence or absence of epithelium lining the cyst. Pseudocysts, which lack an epithelial lining, are typically associated with acute or chronic pancreatitis.[6] Neoplastic CPTs which constitute 15% of all pancreas cysts are lined by epithelium which may harbor a potential risk of progression to malignancy.[6,7] These CPTs include serous cystadenomas (SCA), intraductal papillary mucinous neoplasms (IPMN), mucinous cystic neoplasms (MCN), cystic neuroendocrine tumors, solid pseudopapillary tumors (SPT), and a few other rare tumors (Table 1).[8]

Morphologically, the conventional microcystic SCA is a well delineated lesion with multiple, small fluid-filled cavities (typically less than 5 mm in size) which are separated by thin septae and lined by cuboidal epithelial cells (Figure 1). A central scar (usually referred to as sunburst calcification but may represent only fibrosis) may be present in up to 1 quarter of images.[9] The less often encountered macrocystic variant (cyst size over 2 cm) of SCA cannot be reliably distinguished morphologically from MCNs. The presence of any intramural nodules, cyst wall thickening, floating debris, mucin or associated pancreatic duct dilation in a suspected SCA is unusual and could alternatively indicate a mucinous

Abbreviations used in this paper: CA, cancer antigen; CEA, carcinoembryonic antigen; CPT, cystic pancreatic tumor; CT, computed tomography; EUS, endoscopic ultrasound; FNA, fine needle aspiration; IPMN, intraductal papillary mucinous neoplasms; MCN, mucinous cystic neoplasm; MD, main pancreatic duct; MRCP, magnetic resonance cholangiopancreaticography; MRI, magnetic resonance imaging; SB, pancreatic duct side branch; SCA, serous cystadenoma; SPT, solid pseudopapillary tumors.

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Table 1. Clinical, Demographic, and Cyst Fluid Characteristics of Commonly Encountered Benign Cystic Pancreatic Tumors

			Cyst fluid characteristics and cytology					
Lesion	Demographics	Location	Fluid color	Viscosity	Cytology	CEA	CA 19–9	Amylase
Nonmucinous								
Serous cystadenoma	Seventh decade; F > M	Body/tail > head	Colorless with blood contaminant	Usually low	Usually acellular. Small glycogen staining cuboidal cells may be seen in the background	Undetectable to low[a]	Low[a]	Low
Pancreatic neuroendocrine tumors[b]	Third to sixth decade; M > F	Body/tail > head	Colorless	Usually low	Small homogenous small-cell population with round nuclei that stain positive for chromogranin and synaptophysin on cell block	Undetectable to low[a]	NA	Low
Solid pseudopapillary tumors[c]	Second and third decade; F > M	Body/tail > head	Colorless	Usually low	Branching papillae with myxoid stroma that reacts to vimentin on cell block	NA	NA	NA
Mucinous								
Intraductal papillary mucinous neoplasm	M = F	Main duct or side branch; head > body and tail	Colorless	Usually high	Acellular with background of mucin. Occasionally mucinous epithelial cells with papillary projections and variable atypia may be seen	Moderate increase[a]	Variable[a]	High
Mucinous cystic neoplasm	Fifth and sixth decade; F > M	Body/tail > head	Colorless	Usually high	May be acellular with background mucin. Mucinous epithelia cells may be seen	Moderate increase[a]	Variable[a]	Variable

F, female; M, male; NA, not applicable.
[a]Marker levels usually as described, but overlap may occur.
[b]Usually solid but can be partially or completely cystic in 10% of cases.
[c]Can be purely solid or a mixed solid and cystic.