

REVIEW

Cystic pancreatic neoplasms

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Abstract

Cystic pancreatic neoplasms are uncommon, but are being seen more frequently due to the widespread use of cross-sectional imaging. In this article, we will address the clinical and imaging features of the more commonly seen neoplasms. Points of differentiation between these neoplasms, the use of cyst fluid analysis and an approach to the incidentally discovered cystic mass will be addressed.

Keywords: *Pancreas; CT; cysts; MR; neoplasms; pancreatic ducts.*

Introduction

Cystic pancreatic neoplasms are rare, accounting for less than 1% of pancreatic neoplasms. The differential diagnosis for a cystic pancreatic mass includes the following:

- pseudocyst
- serous neoplasms cystadenoma/adenocarcinoma
- mucinous neoplasms
- solid pseudo-papillary epithelial neoplasm
- Von-Hippel Lindau disease
- cystic islet cell neoplasms
- cystic metastases
- lymphangioma
- giant cell neoplasm.

However, Sarr and his colleagues^[1] have classified these rare neoplasms into a more simplified and practical working classification, which is as follows:

- serous cystic neoplasms
- mucinous cystic neoplasms
- intraductal papillary mucin-producing neoplasms
- other less common neoplasms.

The following discussion will be restricted to the more common neoplasms.

Serous cystic neoplasms

Previously referred to as a microcystic neoplasm, this is a benign neoplasm and is commonly seen in women, usually in the fifth or sixth decades of life. The most common type of serous neoplasm, the microlacunar type, has a characteristic imaging appearance on CT and MRI^[2–4]. It is usually seen as a lobulated mass with small cysts defined by septations and a central scar (Figs 1 and 2). The central scar may have areas of calcification, although this is uncommon. This results in a honeycomb appearance with the central calcifications producing a sunburst pattern. While this appearance is almost pathognomonic for the diagnosis of a serous cystic neoplasm, it is unfortunately not seen

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in all serous cystic neoplasms^[2,3]. The cystic nature of the mass and the septae is better appreciated on ultrasonography. However, if the cysts are very small, these neoplasms can appear echogenic and resemble other solid pancreatic neoplasms. On angiography these neoplasms can be hypervascular. Pathologically, the neoplasm has a honeycomb appearance with the septae and the central scar. Microscopically, the cysts are filled with glycogen and the cystic spaces are lined with flat epithelial cells. In view of the glycogen content within the cysts, these neoplasms are also referred to as the glycogen-rich neoplasms.



Figure 1 Contrast-enhanced CT shows a lobulated multiseptate mass in the pancreatic head. The small cystic spaces defined by the septae are typical for a serous tumor.

Mucinous cystic neoplasm (MCT)

Also called macrocystic neoplasms, these neoplasms are also seen most commonly in women in the fifth and sixth decades of life. Approximately 80% of these tumors tend to involve the body and tail of the pancreas. The mucinous neoplasms have larger thick-walled cysts (Fig. 3). The cysts are usually larger, greater than 2 cm in size, and fewer in number than serous neoplasms. They can be unilocular or multilocular in nature^[4-8]. When the neoplasm is unilocular it cannot be easily distinguished from a pseudocyst. In mucinous neoplasms, thick-walled cysts with nodularity and excrescences are noted on pathology. The cysts are filled with thick mucinous material and, on microscopy, are lined with goblet cells, which produce mucin. Epithelium ranging from normal to cellular atypia to frank adenocarcinoma may line the wall of one cyst. For this reason, cyst wall biopsy is not helpful in deciding whether or not the cyst is malignant. The biologic behaviour of these neoplasms is dependent on presence or absence of invasion. Non-invasive neoplasms, when completely resected, have an excellent prognosis; invasive neoplasms have a poor prognosis, similar to that seen in ductal adenocarcinoma.

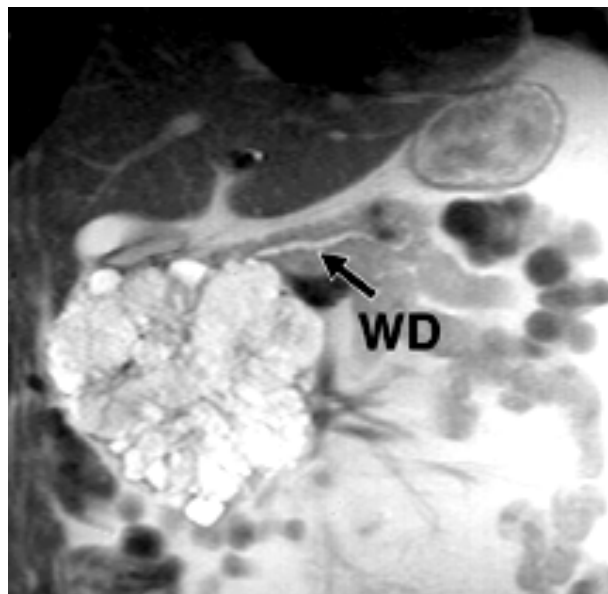


Figure 2 Single-shot fast spin-echo coronal MR image demonstrates a lobulated multiseptate mass in the uncinus process. The small cystic spaces and septae, both of which are characteristic of a serous tumor, are well demonstrated. Arrow demonstrates the duct of Wirsung (WD). [Courtesy of Dr Mark R Paley MD, Hammersmith Hospital, London, UK].

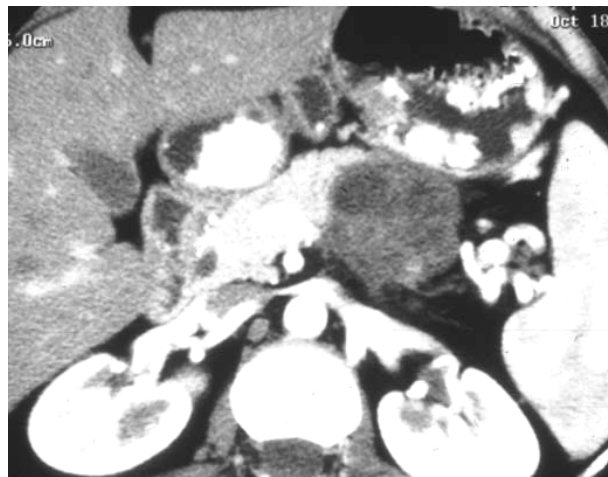


Figure 3 Contrast-enhanced CT shows a cystic mass in the mid-body of the pancreas. The mass has areas of cystic degeneration but has solid nodules. The location and appearance are suggestive of mucinous neoplasm, which this proved to be on subsequent surgical resection.

Imaging differences between serous and mucinous cystic neoplasms

Johnson *et al.* reported a classification scheme which is of some use in distinguishing between serous and mucinous neoplasms^[2]. Useful criteria include the number of cysts

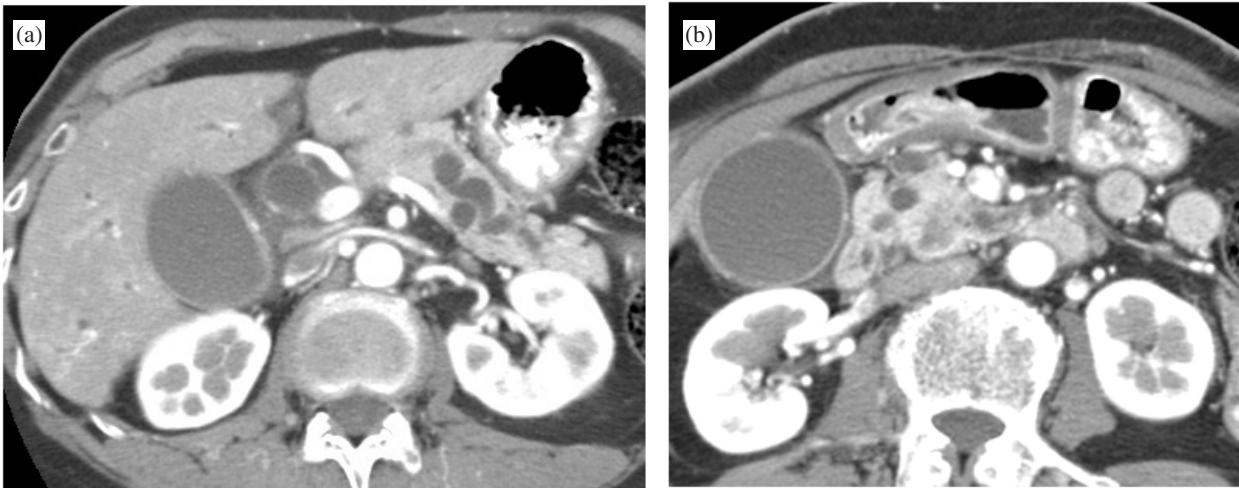


Figure 4 Contrast-enhanced CT demonstrates marked pancreatic duct dilatation throughout the main pancreatic duct, with cystic spaces in the uncinate process. On endoscopy a dilated bulging papilla with abundant mucin exuding from it was found. These findings are characteristic of an intraductal mucin-producing tumor of the main duct type.

(less than six in MCT neoplasm, greater than six in serous cystic neoplasm) and size of the majority of cysts (greater than 2 cm in MCT neoplasm and less than 2 cm in serous cystic neoplasm). Using these criteria, an accuracy rate of 70–80% was achieved in distinguishing the benign serous from the malignant mucinous neoplasms.

However, a more recent study of 100 cystic neoplasms (excluding pseudocysts) by Procacci *et al.* found that accurate characterisation was possible in only 60% of neoplasms. In general, serous neoplasms were more accurately diagnosed than mucinous neoplasms. 15–20% of neoplasms were incorrectly diagnosed due to the overlap of imaging features^[9]. Curry *et al.* also reported low accuracy rates when readers were asked to classify cystic neoplasms into serous or mucinous categories^[10].

Intraductal papillary mucinous or mucin-producing neoplasm (IPMT)

Considered to be a relatively new entity, IPMT represents a spectrum of a neoplastic process, which has been previously referred to by a variety of terminology. Controversy remains as to the exact nature of the neoplasm, its biology and also its management and treatment. However, there are some distinct cross-sectional imaging characteristics that correlate well with the documented endoscopic retrograde cholangiopancreatography (ERCP) findings. These include a markedly dilated pancreatic duct, excessive mucin secretion and bulging papilla.

Depending on whether the main pancreatic duct or a side branch is involved, these neoplasms have been divided into:

- main duct type
- side branch or branch duct type.

The imaging appearances of the two types can vary [11–17].

Main duct type

The main duct type of this neoplasm can involve either a segment of the main duct or the duct diffusely. The pancreatic duct is dilated and is filled with mucin. Filling defects due to mucin deposits or neoplasm can be seen within the dilated main pancreatic duct as well as within large tubular cystic spaces (Fig. 4). Thin-section CT and magnetic resonance cholangiopancreatography (MRCP) can demonstrate the communication of these dilated cystic spaces with the main pancreatic duct^[14]. ERCP has been the gold standard for making this diagnosis, when it demonstrates a bulging papilla with an outpouring of large amounts of mucinous material from it. As the imaging findings often resemble that of pancreatitis, these patients are often misdiagnosed and treated as having chronic pancreatitis. On ERCP and MRCP, a dilated main pancreatic duct with beading of the side branches can be seen (Fig. 5(a)) and at times filling defects within these dilated ducts due to mucin or tumor nodules can be seen (Fig. 5(b))^[15,16].

Side branch type

The side branch or branch duct type of this neoplasm is usually confined to the head and uncinate process. On CT and MR a cystic mass is seen which can resemble serous and mucinous neoplasms (Fig. 6). On ERCP and MRCP,

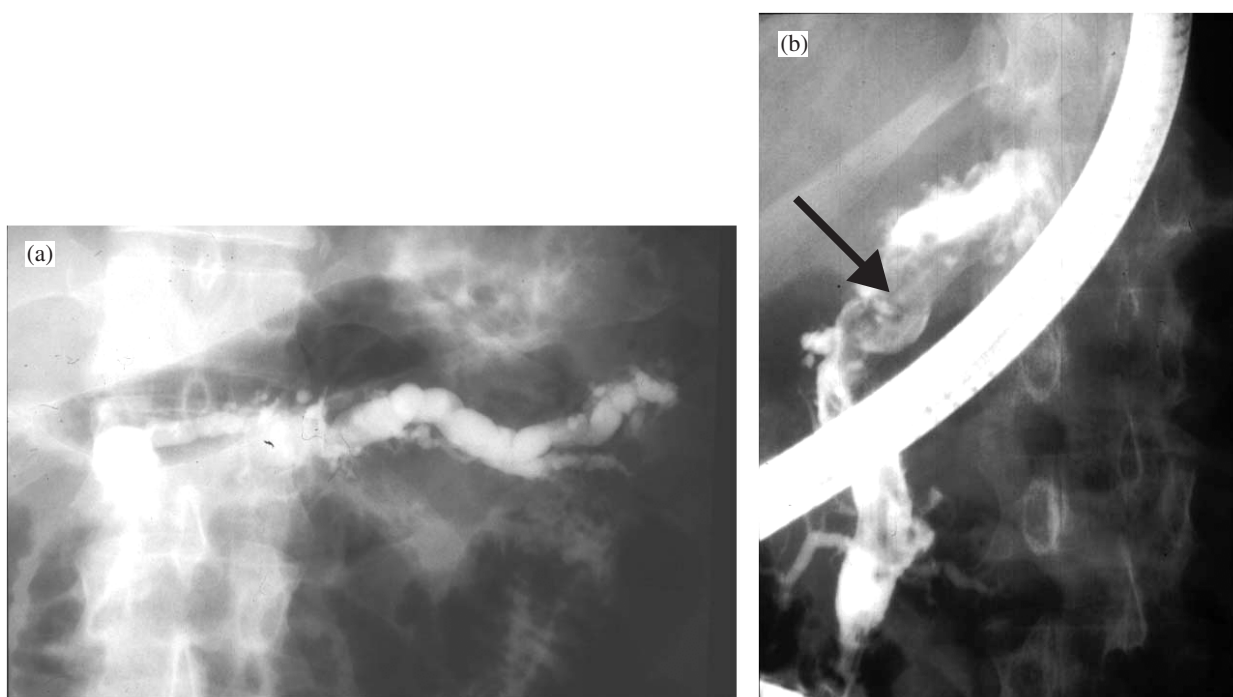


Figure 5 ERCP can demonstrate features which resemble those of pancreatitis, such as a dilated main duct with beaded appearance to the side branches (a), or demonstrate filling defects in these dilated ducts which may be due to mucin or tumor nodules (arrow) (b).

a characteristic appearance of cystic masses is seen, resembling a 'cluster of grapes', which communicates with the pancreatic duct^[11-17].

The main duct type of IPMT is thought to be more malignant and requires surgical treatment in the form of a total or subtotal pancreatectomy. In contrast, side branch neoplasms are thought to be less aggressive and can be either observed or resected with a Whipple procedure. The prognosis for the side branch type neoplasms is excellent in contrast to that for the main duct type, which if invasive can be similar to that for ductal adenocarcinoma.

Solid pseudo-papillary epithelial neoplasms

Solid and cystic papillary epithelial neoplasms are unusual neoplasms commonly seen in women in the 20-40-year age range. These neoplasms have an excellent prognosis and are considered a low-grade malignant neoplasm. They can be entirely cystic or solid but can have both solid and cystic components (Fig. 7). Hemorrhage and fluid levels are reportedly common. These neoplasms can metastasise to the liver and upper abdominal nodes^[18,19]. As they are considered to be of low malignant potential and have a good prognosis, neoplasm resection is at times undertaken even in the presence of metastatic disease.

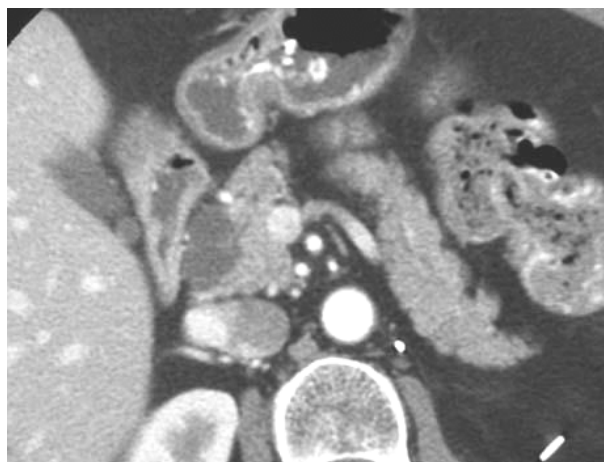


Figure 6 Contrast-enhanced CT demonstrates a cystic mass in the uncinus process of the pancreas. This on surgery proved to be an intraductal mucin-producing tumor of the side branch or branch duct type. These are usually confined to the uncinus process but can be only readily diagnosed on ERCP (Fig. 7). On imaging, the appearances can resemble that of a serous or mucinous tumor.

Aspiration biopsy and cyst fluid analysis of cystic neoplasms

There are data suggesting that differentiation between the benign and malignant cystic neoplasms may be

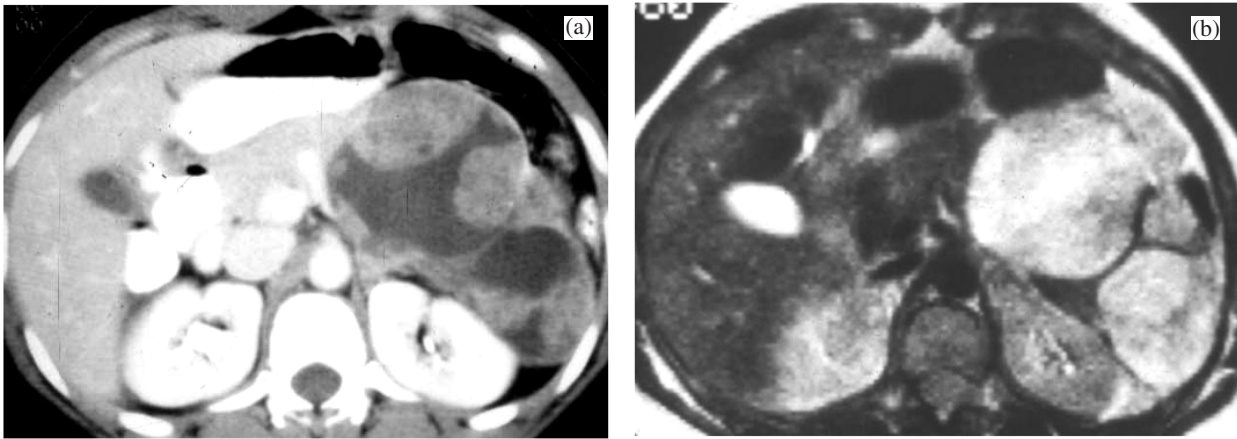


Figure 7 Contrast-enhanced CT (a) in a 25-year-old woman demonstrates a large multilobulated cystic mass with soft tissue nodules and excrescences involving the body and tail of the pancreas. This imaging appearance is re-demonstrated on a moderately T2-weighted MR image (b). On surgery this proved to be a solid pseudopapillary neoplasm.

achieved with high sensitivity, based on analysis of the cyst fluid for carcino-embryonic antigen (CEA) and other neoplastic markers such as CA-129 as well as amylase levels^[20,21]. The CEA and other neoplasm marker levels are high in the malignant cystic neoplasms and low in the pseudocysts and in the serous cystic neoplasms.

Histological analysis of fine-needle aspiration (FNA) biopsy is fraught with problems, as the lining epithelium of the serous and cystic neoplasms can be incomplete and/or heterogeneous; thus a biopsy at one site may not be a true representation of the neoplasm type.

Incidental 'cystic lesions' in asymptomatic patients

Due to the widespread use of imaging, small 1–3 cm cystic masses in the pancreas are being more commonly seen in asymptomatic individuals. As true epithelial cysts of the pancreas are extremely rare, these are a diagnostic problem. Some believe that they may represent side duct intraductal neoplasms, and recommend follow-up to ensure stability. An ERCP or MRCP may also prove the true nature of the lesion by demonstrating a communication with the pancreatic duct, which is characteristic of an IPMT. If these lesions are being managed conservatively and followed with serial imaging, surgical resection should be performed if there is any change in size or internal features^[22].

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