

From the Archives of the AFIP

Mucinous Cystic Neoplasms of the Pancreas: Radiologic-Pathologic Correlation¹

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LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

- Describe the clinical and pathologic features that characterize mucinous cystic neoplasms of the pancreas.
- Recognize the imaging findings and the important differential diagnostic features that are typical of mucinous cystic neoplasms.
- Emphasize the appropriate therapy and disease prognostic implications for patients affected with mucinous cystic neoplasms.

Mucinous cystic neoplasms of the pancreas are rare primary tumors. They have pathologic and clinical similarities to biliary cystadenomas of the liver and mucinous cystic tumors of the ovary. Mucinous cystic neoplasms of the pancreas typically affect middle-aged women and arise in the tail of the pancreas. Gross pathologic and imaging features usually are those of a large, multilocular cystic mass. There is, however, a spectrum of radiologic findings that overlaps with those of other entities including pancreatic pseudocyst, other primary epithelial and nonepithelial tumors of the pancreas, and metastases. In most cases, ultrasound and computed tomography are the mainstays for radiologic evaluation, with magnetic resonance imaging having a complementary role. All mucinous cystic neoplasms should be considered as mucinous cystadenocarcinomas of low-grade malignant potential. Complete surgical excision alone results in an excellent clinical outcome and disease-free survival, irrespective of histologic or radiologic parameters in over 90% of cases studied.

■ INTRODUCTION

Mucinous cystic neoplasms of the pancreas are low-grade tumors that occur predominantly in middle-aged women, arise within the tail of the pancreas, and manifest as large multiloculated cystic masses. They represent approximately 10% of pancreatic cysts and 1% of pancreatic neoplasms (1-5). Other names used for this lesion include mucinous cystadenoma and mucinous cystadenocarcinoma. However, mucinous cystic neoplasm is a distinctly different clinical and pathologic entity from ductal adenocarcinoma and its variants (including mucinous adenocarcinoma of the pancreas) and

Abbreviations: AFIP = Armed Forces Institute of Pathology, CEA = carcinoembryonic antigen

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intraductal papillary tumors (mucin-hyper-secreting adenocarcinoma of the pancreas). Thus, attempts to group these lesions together should be dismissed to avoid confusion (6-15).

The pathogenesis of mucinous cystic neoplasm of the pancreas is uncertain. However, this lesion shares both clinical and pathologic characteristics with biliary and ovarian mucinous tumors (eg, the typical patient is a middle-aged woman) (16,17). It has been speculated that the embryologic origins of these lesions may be related to germ cell migration in early fetal life during the first 8 weeks of gestation (16). Each of these tumors is characterized by a columnar, mucin-producing epithelium that lines a cystic cavity, subtended by a densely cellular mesenchymal stroma, reminiscent of ovarian stroma. The entire tumor is usually encapsulated by an outer layer of fibrous connective tissue. The presence of ovarianlike stroma in biliary cystadenomas and cystadenocarcinomas has been shown to be an independent predictor of a favorable prognosis (16). However, the presence of the stromal component, in some form, is required for the diagnosis of mucinous cystic neoplasm of the pancreas, and all usually have a favorable prognosis. At the Armed Forces Institute of Pathology (AFIP), all cases of mucinous cystic neoplasms are interpreted as mucinous cystadenocarcinomas of low-grade malignant potential, irrespective of histologic appearance, to reinforce the need for complete surgical resection and close clinical follow-up.

This article is based on a review of the literature and over 50 cases of mucinous cystic neoplasms with cross-sectional imaging studies accessioned to the Radiologic Pathology Archives of the AFIP from January 1979 to January 1996. Representative examples of these cases are illustrated in this article, which will examine the clinical, pathologic, and radiologic features of mucinous cystic neoplasms. Cross-sectional imaging is an invaluable tool in characterizing these lesions, with ultrasound (US)

and computed tomography (CT) being the most commonly used modalities. Magnetic resonance (MR) imaging usually has a complementary role. The differential diagnosis, treatment, and prognosis of mucinous cystic neoplasms are also discussed.

■ CLINICAL FEATURES

Mucinous cystic neoplasms are found almost exclusively in women (greater than 95% of cases) in their 4th to 6th decades of life (2,4,5, 18-26). However, there is a spectrum, and cases can be seen in women ranging in age from 20 through 95 years old. In our series at the AFIP, we saw no cases in men, although they have been reported.

These tumors can manifest clinically with abdominal pain or a mass lesion, or the patient may complain of anorexia. Alternatively, these tumors may be incidentally discovered during work-up of unrelated symptoms or diseases. Symptoms probably result from mass effect on contiguous structures such as the stomach, intestines, or abdominal wall. The larger the lesion, the more likely it is to be symptomatic. In rare cases, the tumor may manifest with local invasion or distant metastases, especially within the liver. The rare lesion that occurs within the head of the pancreas can cause jaundice by obstructing the common bile duct. In addition, a very small number of patients present with findings suggestive of a pancreatic pseudocyst, such as a history of pancreatitis, trauma, or alcoholism, which is clinically helpful in distinguishing this lesion from a pancreatic pseudocyst (2,8,21,22). Rare systemic manifestations, caused by tumor production of gastrin and vasoactive intestinal polypeptide, have been reported, including Zollinger-Ellison syndrome and watery diarrhea, hypokalemia, and achlorhydria (20,23). In our review, we found one patient who presented with Zollinger-Ellison syndrome but was subsequently discovered to have a synchronous gastrinoma.

■ PATHOLOGIC FEATURES

Mucinous cystic neoplasms are large, ranging from 2 to 36 cm, with an average size of 10 cm. Over 90% of mucinous cystic neoplasms

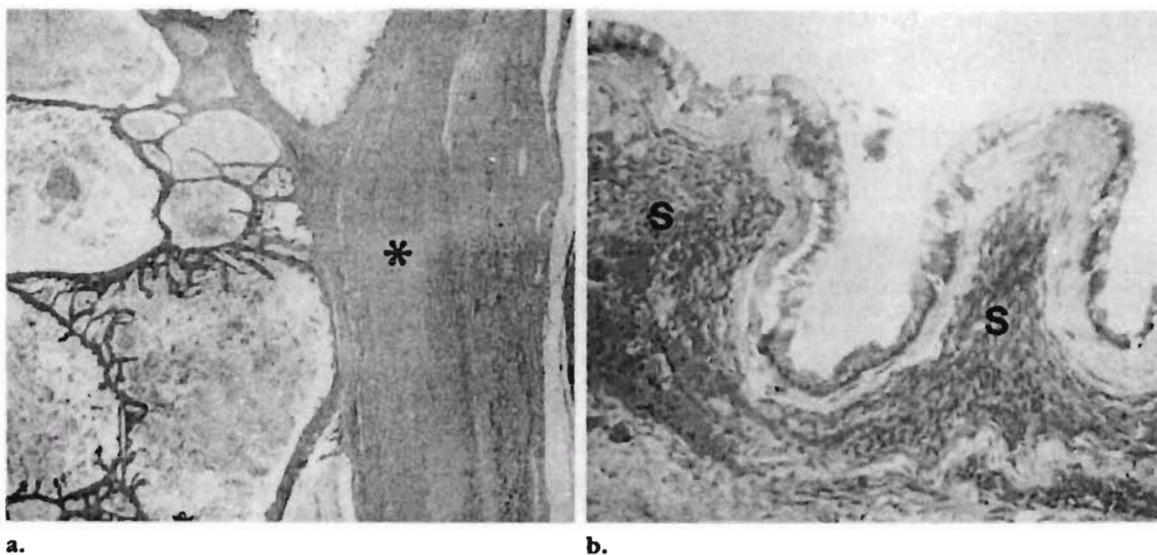


Figure 1. Histologic appearance of mucinous cystic neoplasm. (a) High-power photomicrograph (original magnification $\times 160$; hematoxylin-eosin stain) demonstrates a well-developed capsule (*) and thin internal septations. Small papillary projections are noted along the epithelium. (b) High-power photomicrograph (original magnification $\times 280$; hematoxylin-eosin stain) shows an epithelial layer of benign cells overlying the dense mesenchymal stroma. This ovarian-type stroma (S) characterizes the lesions.

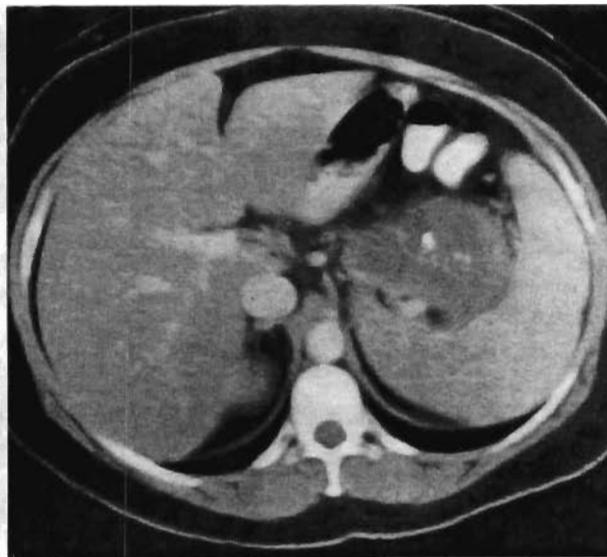
are found in the body or tail of the pancreas (2,4,5,18-26). The tumors are usually smooth and round; however, a bosselated or lobulated surface may be seen.

Cut sections of a typical mucinous cystic neoplasm reveal a multilocular cystic lesion with thin septations measuring less than 2 mm in greatest thickness. The cystic cavities vary in size and measure up to many centimeters in greatest dimension. The internal surface of the cyst may contain thin papillary projections into the lumen. The cystic cavity may be filled with thick, mucoid material or clear, green, or blood-tinged fluid. Less commonly, a mucinous cystic neoplasm may contain round, gray-white, mural nodules. In rare cases, a single dominant cystic cavity or a unilocular cystic mass may be seen. The lesions do not normally communicate with the pancreatic duct unless a fistula has developed (in contrast to pancreatic pseudocysts, which communicate with the pancreatic duct in up to 70% of cases) (19,24,25).

Histologically, the epithelial lining of mucinous cystic neoplasms is composed of tall, mucin-producing, columnar cells arranged in a

single orderly layer that abruptly transforms into a stratified, papillary epithelium with cytologic atypia or areas of frank carcinoma (Fig 1). These changes are focal and abrupt and frequently occupy a small area. If many sections are carefully examined, cytologic or architectural atypia can usually be seen. All tumors have a characteristic ovarian-type dense stroma, with spindle cells arranged in a haphazard configuration. Fibrous hyalinization can also be identified in many tumors.

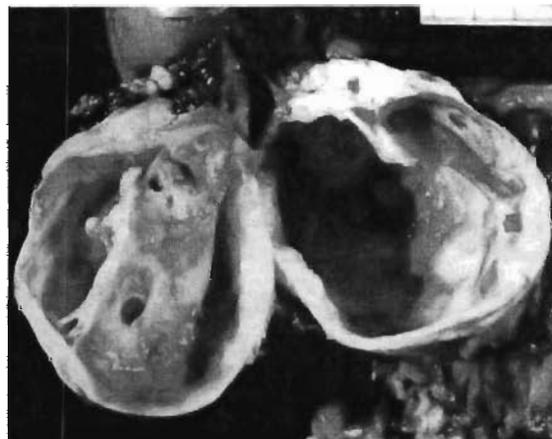
Calcification or osseous metaplasia, as well as other degenerative changes, can be seen in the wall and septa of a mucinous cystic neoplasm. Whether the benign-appearing epithelium transforms into malignant epithelium in a teleologic development of the tumor or whether the malignant epithelium "matures" into a benign-appearing epithelium is unknown. For these reasons, the pathologists at the AFIP consider all mucinous cystic neoplasms to be malignant neoplasms of low-grade malignant potential (26).



a.

b.

Figure 2. Mucinous cystic neoplasm in a 33-year-old woman with intermittent left upper quadrant pain for 1.5 years. (a) US scan demonstrates a multi-loculated cystic mass with echogenic internal septa within the tail of the pancreas. The mass measures approximately 9 cm in average transverse dimension. Echogenic foci with shadowing that correspond to calcifications are noted along the septa (arrow). Note the low-level internal echoes. (b) Axial contrast material-enhanced CT scan clearly demonstrates the calcifications along the internal septa, as well as the cystic nature of the mass. (c) Photograph of the gross specimen reveals the well-circumscribed mass with internal septa that incompletely separate the loculated cystic components of the lesion. Scale is in centimeters.



c.

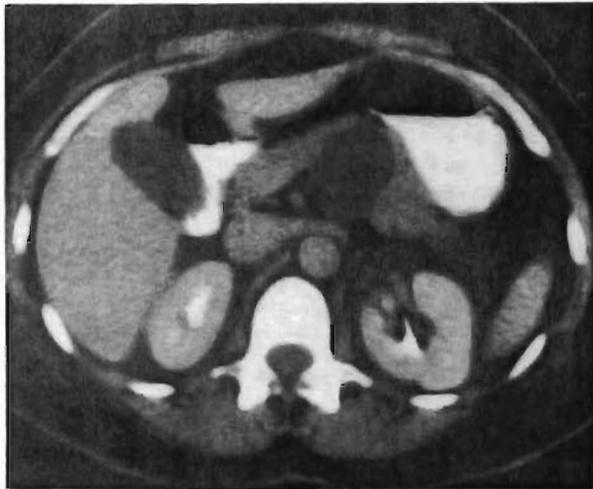
At this time, there is no epithelial immunohistochemical reaction (carcinoembryonic antigen [CEA], dupan-2, carbohydrate antigen 19-9 [CA 19-9], keratin, epithelial membrane antigen [EMA], anticytokeratin [CAM5.2], or B72.3 [an epithelial marker]) that allows reliable differentiation between mucinous cystic neoplasms and other pancreatic lesions, nor can an accurate distinction between mucinous cystic neoplasm and frank carcinoma be made by using immunoreaction. Therefore, these analyses are not helpful except in rare cases (26).

CEA was present in all the AFIP cases that were tested ($n = 7$), similar to findings reported the literature (20,26-30). Carbohydrate

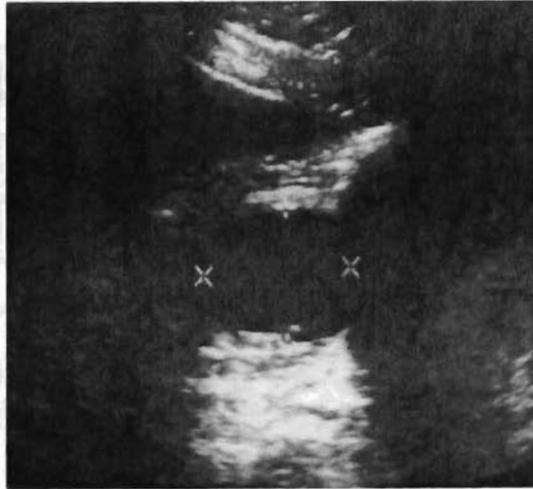
antigen 19-9 is present in both mucinous cystic neoplasm and ductal adenocarcinoma (8,20,21, 26). A variety of peptide hormones such as somatostatin, serotonin, gastrin, and vasoactive intestinal polypeptide have also been reported in mucinous cystic neoplasms (21). Such secretions may explain the rare patient who presents with systemic signs of Zollinger-Ellison syndrome and watery diarrhea, hypokalemia, and achlorhydria.

■ RADIOLOGIC FEATURES

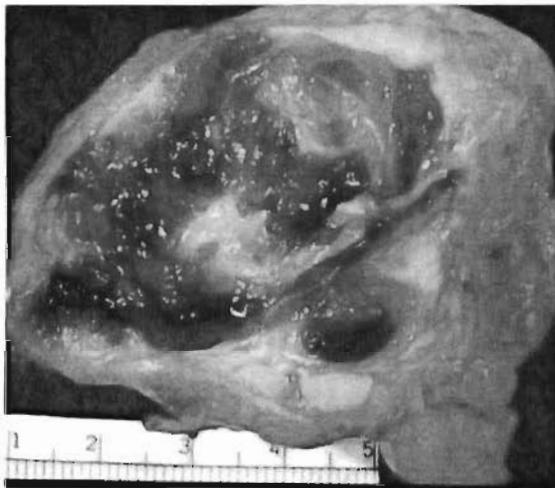
Cross-sectional imaging is the most accurate radiologic method of detecting and characterizing these lesions. Radiography may demon-



a.



b.



c.

Figure 3. Mucinous cystic neoplasm mimicking a unilocular pseudocyst in a 43-year-old woman who presented with abdominal pain and vomiting. (a) Contrast-enhanced CT scan demonstrates a 4-cm low-attenuation cystic mass within the body of the pancreas. (b) US scan helps confirm the unilocular cystic nature of the lesion. (c) Photograph of a cross section of the gross specimen demonstrates the unilocular nature of the lesion, which is irregular along the internal surfaces. This appearance is indistinguishable from that of a pancreatic pseudocyst in the appropriate clinical setting. Scale is in centimeters.

strate discontinuous peripheral calcifications in approximately 16% of lesions, but otherwise radiographic findings do not contribute to the diagnosis (4). There is excellent correlation between the findings at US, CT, and MR imaging (in certain instances, results from these studies may complement each other) and the gross pathologic appearances. All cross-sectional modalities are useful in detecting and characterizing the lesion as well as in evaluating for metastatic disease (1,22,31-39).

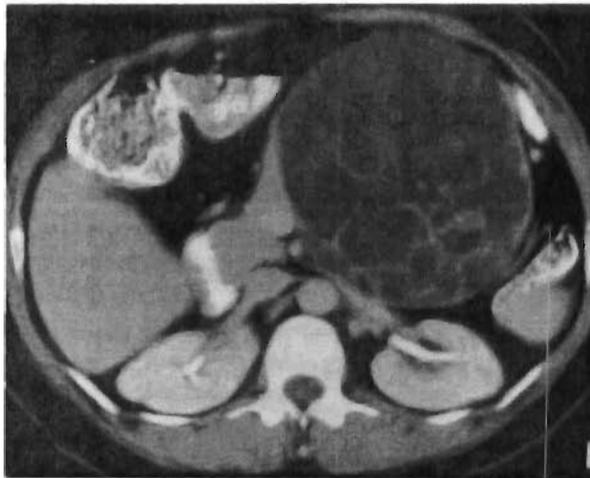
Cross-sectional imaging is ineffective for differentiating between mucinous cystic neoplasms with and without malignant epithelium, except in cases with invasion of adjacent organs, vascular invasion, or metastatic disease. This parallels the findings seen in cases of biliary cystadenomas and cystadenocarcinomas,

in which cross-sectional imaging does not allow differentiation between these lesions (16).

In the appropriate clinical setting, the prospective diagnosis of a mucinous cystic neoplasm is often possible on the basis of cross-sectional imaging features. However, this differentiation cannot always be made, as the appearance of mucinous cystic neoplasm overlaps with that of other benign and malignant cystic masses. Conversely, atypical features of mucinous cystic neoplasms may suggest an alternative diagnosis.

■ US CHARACTERISTICS

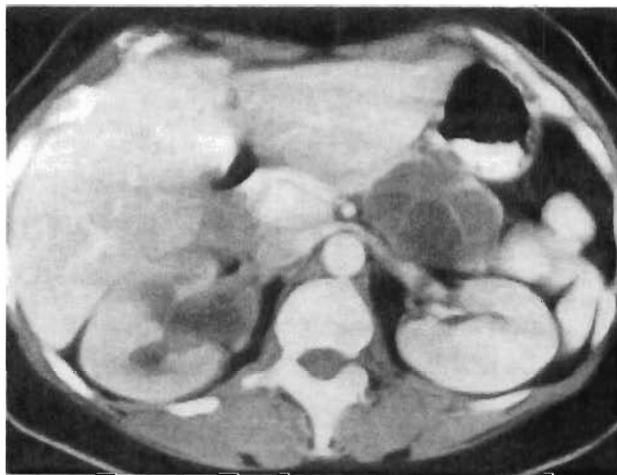
On US scans, mucinous cystic neoplasms appear as multilocular or, less commonly, unilocular lesions with good through-transmission and posterior wall enhancement (Figs 2, 3). Internal septations are usually visualized and more conspicuously demonstrated at US than at CT (1,4,22,31,32,37,39). Nodularity and papillary projections may be demonstrated along



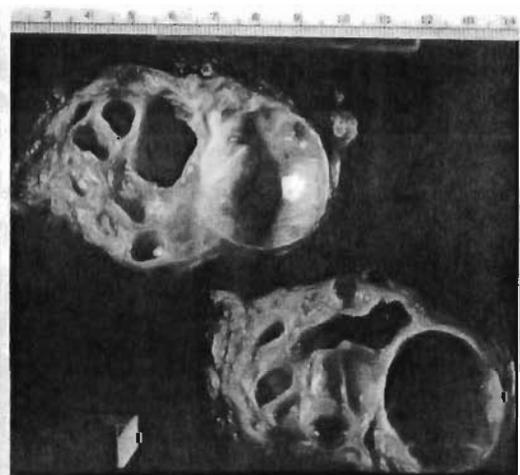
4a.



4b.



5a.



5b.

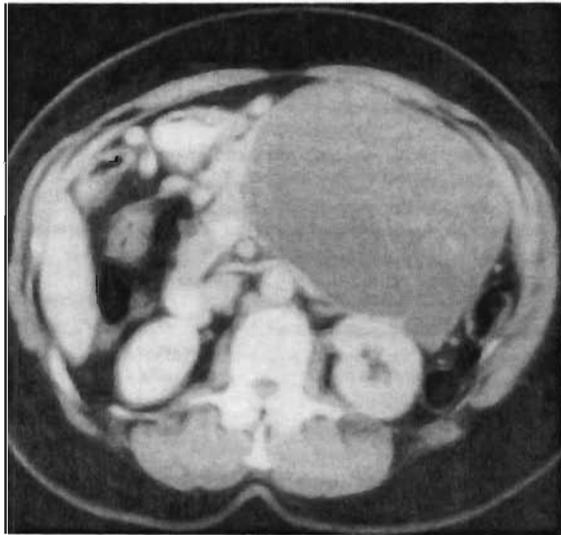
Figures 4, 5. (4) Mucinous cystic neoplasm in a 41-year-old woman with a palpable abdominal mass for over 3 months. (a) Contrast-enhanced CT scan demonstrates a large, well-circumscribed, 18-cm mass within the tail of the pancreas, with enhancement of the thin internal septa and the peripheral wall. These findings are typical of a mucinous cystic neoplasm. (b) Photograph of the bivalved gross specimen demonstrates the well-circumscribed mass with extensive internal septations. Note the differences in color of the fluid within the locules, ranging from clear white mucinous material to hemorrhagic material. (5) Mucinous cystic tumor incidentally discovered in a 45-year-old woman during evaluation for cervical carcinoma and right hydronephrosis. (a) Axial contrast-enhanced CT scan demonstrates a multiloculated 5-cm mass in the tail of the pancreas. Thin internal septa separate the loculated collections, and a more dominant soft-tissue component is seen along the posterolateral margin. (b) Photograph of the gross specimen demonstrates variably sized cystic spaces with intermixed yellow soft-tissue components along the inferior margin. Scale is in centimeters.

the internal wall of the cysts. Metastatic lesions may manifest as simple cysts within the liver.

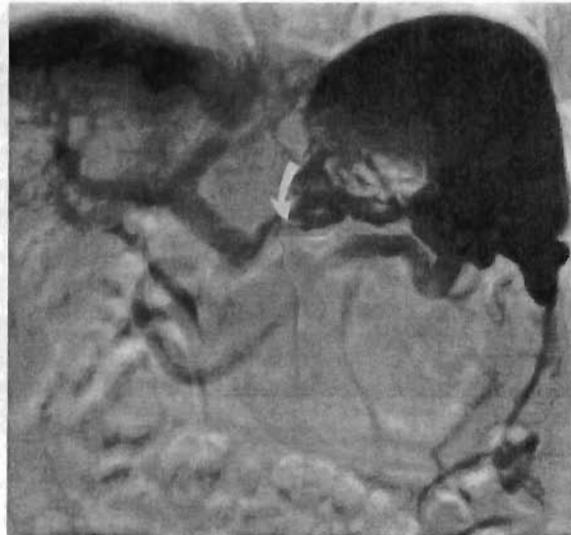
The size and number of pancreatic cysts may be used to distinguish mucinous cystic neoplasms from serous cystadenomas (an invariably benign lesion). Johnson et al (1) demonstrated that when there were six or fewer

cysts measuring greater than 2 cm in diameter, 93% of the mucinous cystic neoplasms were correctly characterized at sonography and 95% at CT.

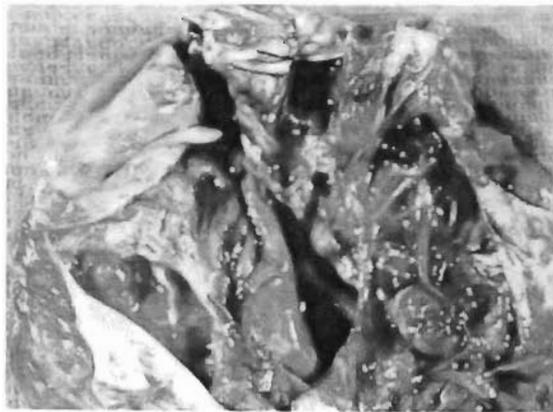
Unilocular cystic masses with or without prominent wall irregularity or mural nodules may also be seen. Such lesions are difficult to distinguish from pancreatic pseudocysts (Fig 3). Less commonly, the cystic cavities may contain echogenic debris or hemorrhage. Echogenic foci due to calcification may be seen in the wall or septa (Fig 2).



a.



b.



c.

Figure 6. Mucinous cystic neoplasm with secondary splenic vein thrombosis incidentally discovered in a 48-year-old woman during physical examination. (a) Contrast-enhanced CT scan demonstrates a multiseptated 14-cm mass at the tail of the pancreas. (b) Angiogram shows large collateral veins extending from the splenic hilum around the periphery of the mass. Thrombus is noted within the splenic vein (arrow). At surgery, there was bland thrombus and no evidence of local invasion or metastases. (c) Photograph of the gross specimen shows the complex multiloculated internal architecture of the lesion.

Endoscopic US, which is performed with a high-frequency (up to 10 MHz) transducer, may provide improved resolution of all these features. Color Doppler US may prove useful in further characterizing these lesions, but its utility has not yet been studied.

■ CT CHARACTERISTICS

On unenhanced CT scans, a mucinous cystic neoplasm appears as a round to slightly lobulated mass that is well encapsulated with smooth external margins. Internal septations may or may not be visualized (1,4,22,34,36-38). Different levels of attenuation may be noted within the cystic cavities depending on their contents, from mucoid to hemorrhagic fluid. Usually, however, the internal contents have attenuation values similar to that of water. Calcifications within the capsule or septa are exquisitely demonstrated by CT in about 10% of cases (Fig 2) (1,4,22,34,36-38). Dynamic

contrast-enhanced CT may demonstrate enhancement of the cyst walls and increased conspicuity of the internal curvilinear septa (Figs 4, 5).

Local invasion of surrounding structures, when present, is accompanied by obliteration of fat planes and distortion and disruption of adjacent organ margins (Fig 6). Metastatic lesions, although uncommon at initial presentation, may appear as solitary or multiple simple cysts within the liver.

■ MR IMAGING CHARACTERISTICS

On unenhanced T1-weighted MR images, mucinous cystic neoplasm demonstrates variable signal intensity, depending on the fluid content. Fluidlike material has low signal intensity on T1-weighted images, whereas proteinaceous or hemorrhagic material may demonstrate high signal intensity on T1-weighted images. On T2-weighted images, these lesions have high signal intensity, similar to that of cerebrospinal fluid (18,27,34,35,40,41), and

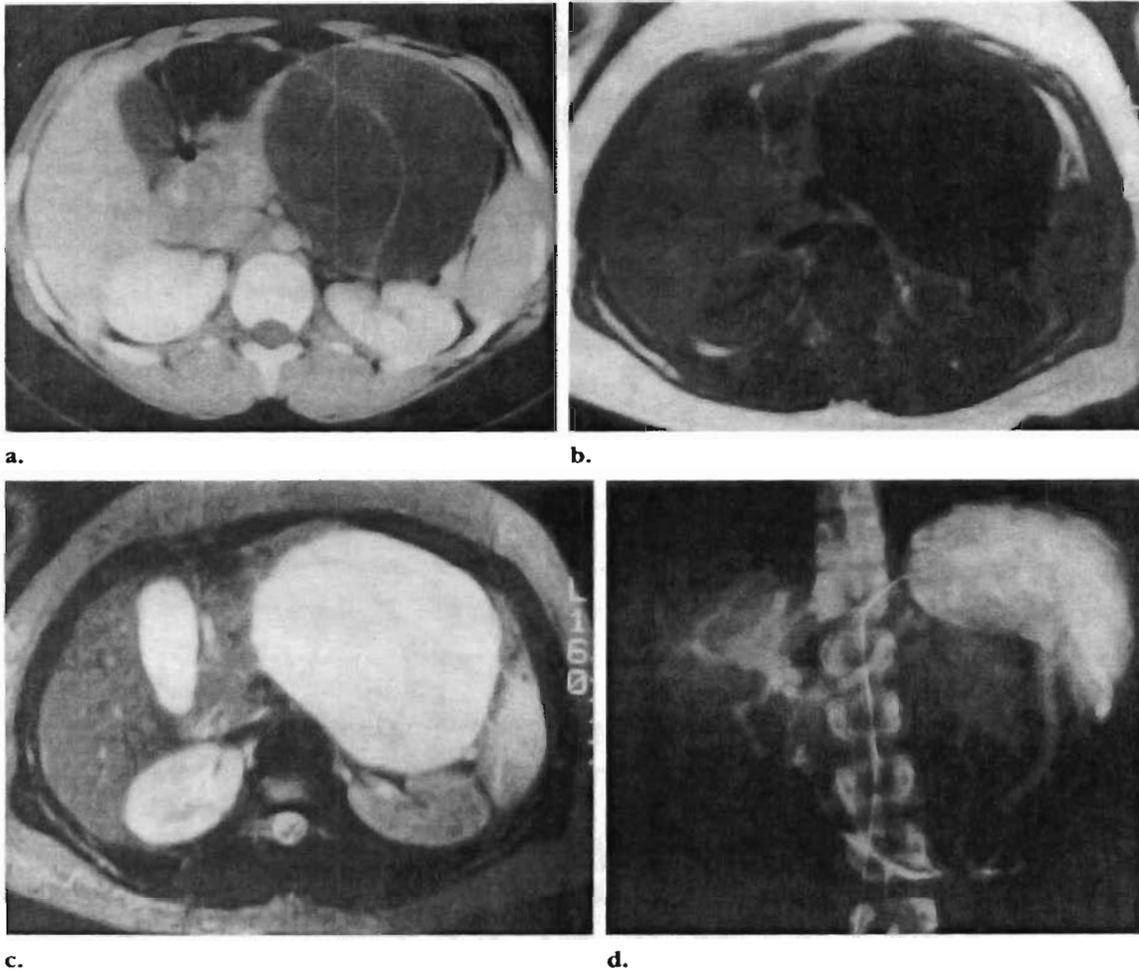


Figure 7. Mucinous cystic neoplasm in a 20-year-old woman who complained of intermittent epigastric pain radiating through her back. **(a)** Contrast-enhanced CT scan demonstrates a multiloculated cystic mass within the tail of the pancreas measuring approximately 17 cm in diameter. **(b)** Axial T1-weighted MR image (repetition time msec/echo time msec = 267/20) demonstrates a lobulated, low-signal-intensity mass. The internal septa are not visualized. **(c)** T2-weighted MR image (2,400/90) demonstrates the overall high signal intensity of the lesion and increased conspicuity of the internal septations. **(d)** Late-phase splenic arteriogram demonstrates displaced parapancreatic and splenic veins but no significant tumor blush or evidence of vascular invasion.

the internal septations are more conspicuous as low-signal-intensity, curvilinear structures (Figs 7, 8).

The role of a gadolinium-based contrast agent has not yet been elucidated, but it may allow more conspicuous demonstration of the

cyst wall and septa on T1-weighted images (Fig 8). Although MR imaging will likely prove to be helpful in the detection of liver metastases, this has not yet been confirmed, and the distinction between metastases and liver cysts on MR images may be as problematic as it is on US and CT scans. In addition, MR imaging does not demonstrate calcifications as well as CT.

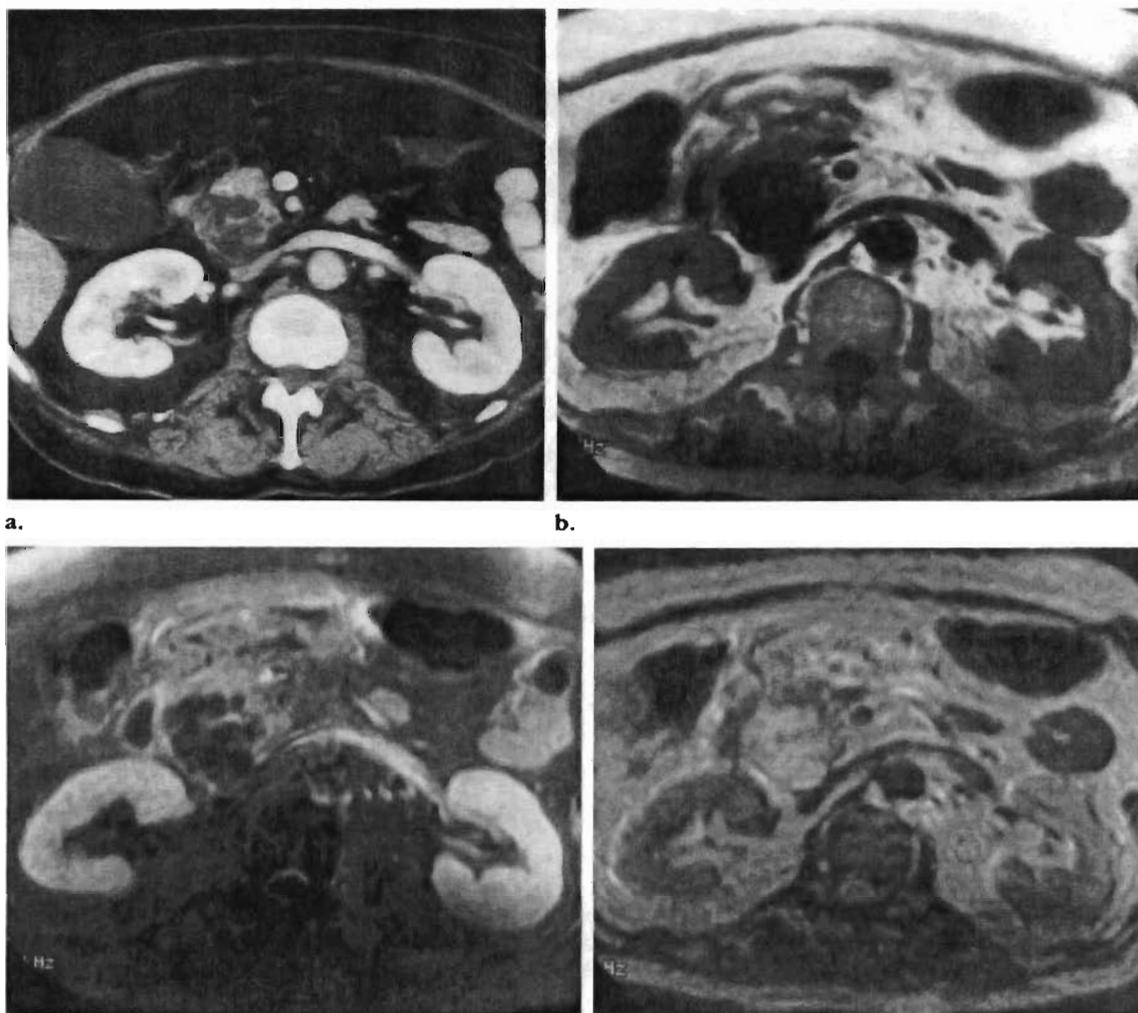


Figure 8. Mucinous cystic neoplasm of the head of the pancreas (an atypical location) in a 63-year-old woman. **(a)** Contrast-enhanced CT scan demonstrates a multiloculated 3.5-cm cystic mass within the head of the pancreas with a focal area of peripheral calcification. **(b)** T1-weighted MR image (700/10) demonstrates the multiloculated mass with areas of low signal intensity that correspond to fluid. Internal septations are less conspicuous than on the CT scan. **(c)** Contrast-enhanced T1-weighted image (700/10 with fat suppression) demonstrates the internal septations to better advantage and highlights the low-signal-intensity cystic areas that constitute the lesion. **(d)** Axial T2-weighted MR image (3,000/102) shows only high signal intensity within the cystic collections. Internal septa are suggested and partially obscured by motion artifact.

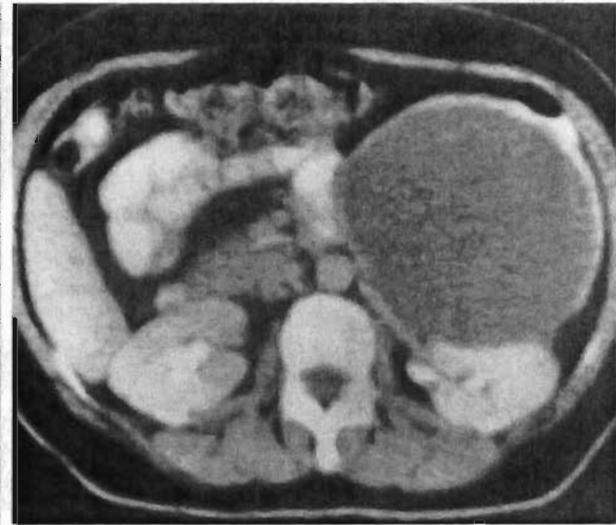
● **Angiographic Characteristics**

Mucinous cystic neoplasms are predominantly avascular masses, which corresponds with their large cystic components. The cyst wall and solid components of the tumor, however, may demonstrate small areas of vascular blush or neovascularity (18,27). There is often dis-

placement of surrounding arteries and veins. In rare cases, mucinous cystic neoplasms may cause splenic vein thrombosis, either by mass effect or invasion (Fig 6).

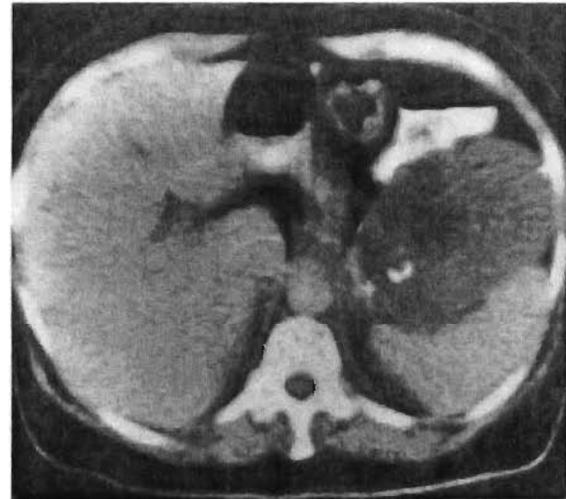


a.



b.

Figure 9. Mucinous cystic neoplasm with an atypical unilocular appearance that had been treated incorrectly as a pseudocyst in a patient with no history of pancreatitis. (a) US scan obtained before treatment demonstrates a well-circumscribed, unilocular mass arising in the tail of the pancreas. (b) Contrast-enhanced CT scan obtained before treatment demonstrates the unilocular mass within the tail of the pancreas. There are no associated inflammatory changes in the surrounding peripancreatic fat or evidence of pancreatic calcifications. (c) Unenhanced CT scan obtained after treatment demonstrates the heterogeneous mixed solid and cystic mass with calcification in the region of the tail of the pancreas. This finding represents recurrent disease after marsupialization of the lesion. Peritoneal metastases were noted at surgery.



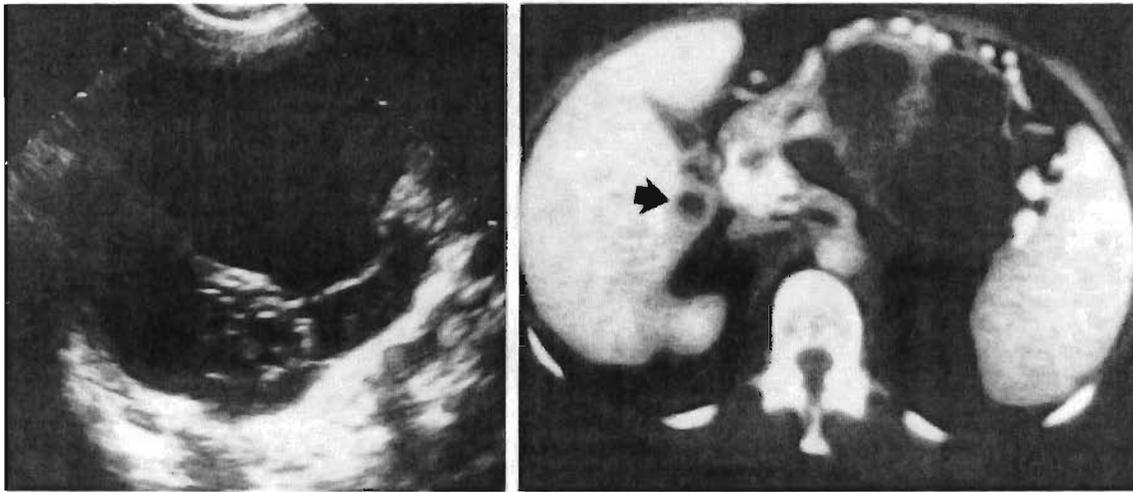
c.

● Radiologic Differential Diagnosis

There is a wide spectrum of radiologic appearances for mucinous cystic neoplasms of the pancreas. The differential diagnosis includes all cystic or partially cystic masses of the pancreas, including inflammatory conditions, benign neoplasms (epithelial and mesenchymal), and malignant neoplasms.

The most important distinction is between mucinous cystic neoplasm and pseudocysts (19,24,25) (Figs 3, 9, 10). Because pseudocysts represent up to 85% of all cystic pancreatic lesions (18,26,31), they are commonly encountered entities that need to be distinguished from rare unilocular mucinous cystic neoplasms. The distinction is important to prevent

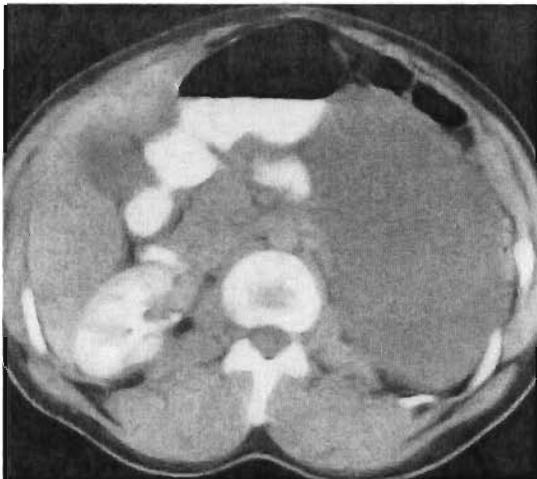
attempts at treating mucinous cystic neoplasms as pseudocysts with drainage or marsupialization. Such procedures are inappropriate for mucinous cystic neoplasms because they invariably lead to recurrence or metastatic disease within the pancreas, liver, or peritoneal cavity (Figs 9, 10). Imaging features that favor a pancreatic pseudocyst include inflammatory changes within the peripancreatic fat, pancreatic calcifications associated with chronic pancreatitis, and the temporal evolution of the lesion at serial examinations. Patients with mucinous cystic neoplasms usually lack a clinical history of pancreatitis or alcoholism, which is often found in patients with pseudocysts. De-



a.

b.

Figure 10. Mucinous cystic neoplasm in a 59-year-old woman with epigastric pain, who was treated with cyst drainage in April 1988 and marsupialization in June 1989. She presented 4 years later with a recurrent multicystic mass of the pancreatic tail with splenic vein invasion, varices, and a cystic lesion within the liver that could represent either a metastasis or simple cyst. (a) US scan demonstrates a complex multiloculated cystic mass within the tail of the pancreas. (b) Contrast-enhanced CT scan demonstrates the posterior mass effect of the cystic mass on the gastric wall. In addition, there is a cystic lesion within the right hepatic lobe (arrow).



a.

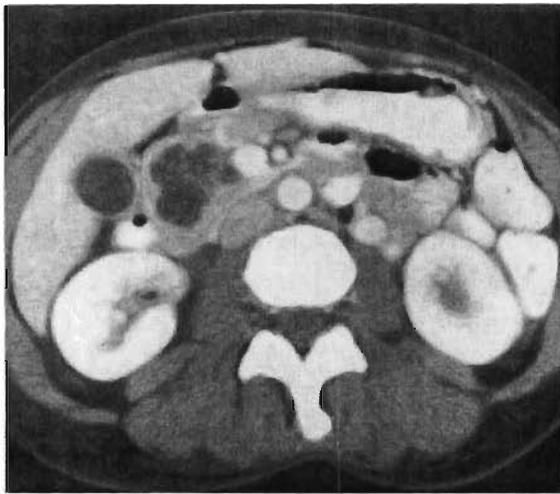


b.

Figure 11. Lymphangioma in a 47-year-old woman with severe abdominal pain, nausea, and leukocytosis. (a) Contrast-enhanced CT scan shows a large, fluid-attenuation, septated mass in the pancreatic body and tail. Each individual cyst is quite large and the borders of the lesion are well-defined. (b) Transverse US scan reveals the multiseptated cystic nature of the mass.

spite these distinguishing features, these lesions may be radiologically indistinguishable. Analysis of the cyst fluid, however, may be helpful for differentiating between these entities (20,21,26-28). Levels of amylase are elevated in cases of pancreatic pseudocysts, and CEA is elevated in cases of mucinous cystic neoplasms.

The finding of a large, well-circumscribed, multiloculated cystic mass arising within the tail of the pancreas in a middle-aged woman is characteristic of mucinous cystic neoplasms. However, this appearance may be mimicked by other pancreatic lesions, including lymphangioma (42,43) (Fig 11), which often extends from or into the retroperitoneal soft tissues; hemangioma (44); variants of adenocarcinoma;

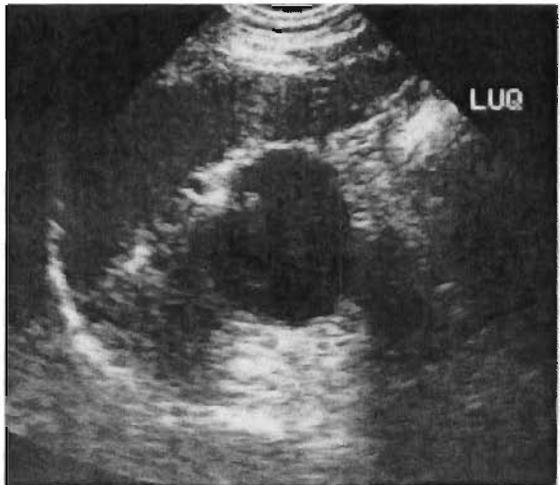


a.

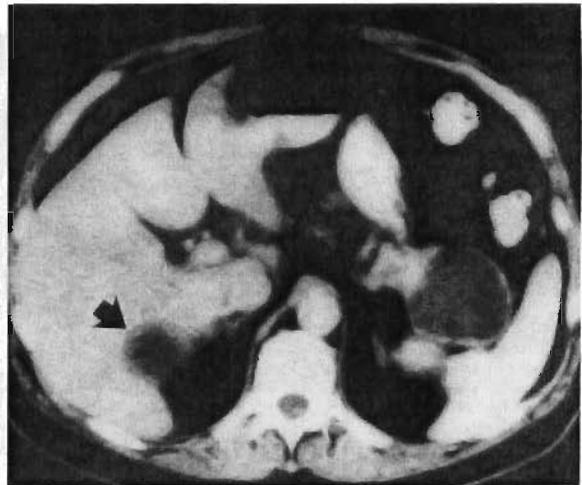


b.

Figure 12. Microcystic adenoma in a 47-year-old woman with vague epigastric and right upper abdominal pain, nausea, and vomiting. (a) Contrast-enhanced CT scan demonstrates a well-defined, multicystic mass in the pancreatic head. No pancreatic ductal dilatation is evident. (b) Transverse US scan helps confirm the multicystic composition of the mass. The size of the cysts is relatively large compared with that generally encountered in microcystic (serous) cystadenoma.



a.



b.

Figure 14. Adenosquamous carcinoma of the pancreas. (a) Longitudinal US scan through the tail of the pancreas demonstrates a well-circumscribed hypoechoic lesion with through transmission and a well-defined posterior wall. There are internal echoes. (b) Contrast-enhanced CT scan helps confirm the well-circumscribed nature of the lesion within the tail of the pancreas. The predominantly low attenuation corresponds to cystic degeneration and necrosis. Note the cystic metastasis within the right lobe of the liver (arrow).

solid and cystic papillary epithelioid neoplasms; cystic islet cell tumors; cystic metastases; atypical serous cystadenoma (1,45) (Fig 12); and sarcomas. Infectious parasitic diseases such as amebiasis or those caused by *Echinococcus multilocularis* rarely affect the

pancreas (46), but may demonstrate an imaging appearance similar to that of mucinous cystic neoplasms. In these cases, clinical history and serologic tests are helpful for distinguishing infections from neoplastic processes.

Malignant neoplasms that produce excessive amounts of mucin or that are prone to cystic degeneration may radiologically resemble the

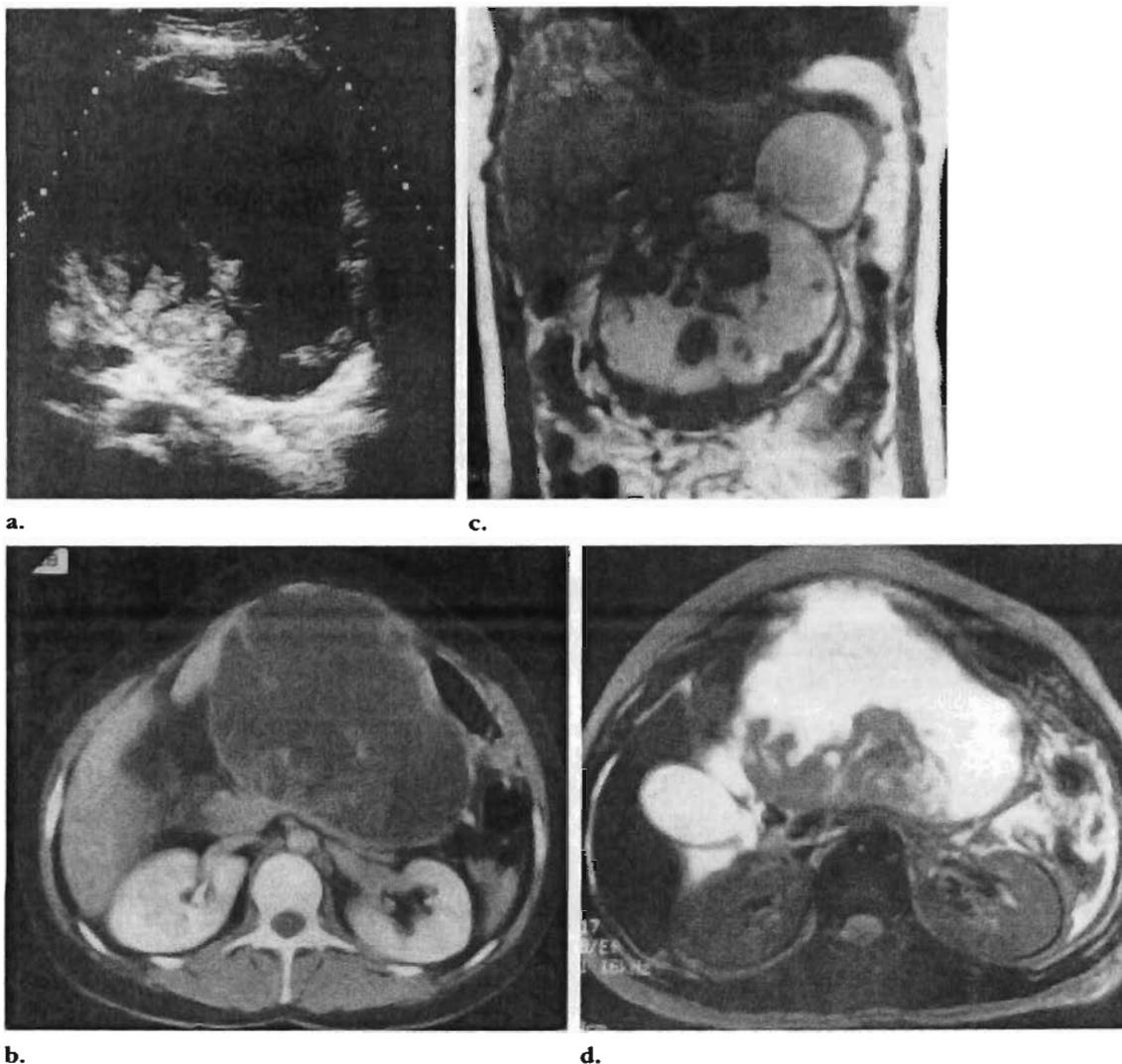


Figure 13. Adenocarcinoma of the pancreas, with mucin secretion. **(a)** Transverse US scan demonstrates a complex, predominantly cystic mass centered within the body of the pancreas. There are low-level echoes within the dependent portion of the mass and a lobulated, hyperechoic posterior soft-tissue component. **(b)** Axial contrast-enhanced CT scan demonstrates a well-circumscribed, lobulated low-attenuation mass with enhancing soft-tissue components, the largest of which is noted along the posterior wall, corresponding to the findings in **a**. **(c)** Coronal T1-weighted MR image (150/20) demonstrates a large multiloculated mass with heterogeneous signal intensity. Central areas of increased signal intensity correspond to the proteinaceous contents of the extracellular mucin. There are also low-signal-intensity solid areas with irregular lobulated margins noted peripherally along the inferior wall and centrally along the upper right margin. **(d)** Axial T2-weighted MR image (5,217/108) demonstrates high signal intensity within the mucinous deposits and heterogeneous lower signal intensity of the solid components.

less typical mucinous cystic neoplasms (which may appear more solid than cystic). Variants of ductal adenocarcinomas (mucinous colloid adenocarcinoma, papillary intraductal adenocarcinoma, adenosquamous carcinoma, and anaplastic adenocarcinoma) may all manifest radiologically as a partially or predominantly cystic mass (6-15) (Figs 13, 14). The difference between adenocarcinoma variants and mucinous cystic

neoplasms is important clinically, pathologically, and prognostically (6-15).

Adenocarcinomas arising within the pancreatic duct system contain malignant epithelial cells capable of producing copious amounts of mucin but do not have ovarian-type stroma. In

some of these cases, the excessive mucin secretion is contained within the pancreatic duct and may be seen flowing out from the ampulla of Vater at endoscopy, and there can be profound ectasia of the pancreatic duct at imaging (Fig 15). In these cases, treatment by means of total pancreatectomy may yield a clinical outcome that is better than that of unconfined adenocarcinoma but is not as favorable as that of mucinous cystic neoplasms. If malignant mucin-secreting epithelial cells are not confined to the pancreatic duct, pools of extracellular mucin may insinuate into the peripancreatic soft tissues and peritoneal cavity, resulting in metastatic implants. Such extensive disease portends the same dismal prognosis attributed to the more typical ductal adenocarcinomas.

Adenosquamous carcinoma contains a mixture of adenocarcinoma and squamous cell carcinoma. Similar to squamous cell carcinomas of the lung, the squamous component, especially when predominant, predisposes the pancreas to necrosis and cystic degeneration. Both adenocarcinoma and squamous cell carcinoma can be identified together or separately in metastatic foci (47) (Fig 14).

Anaplastic adenocarcinoma and giant cell (or pleomorphic) carcinoma of the pancreas may demonstrate a variable amount of cystic degeneration. However, these lesions are very aggressive and frequently demonstrate associated adenopathy and metastases at the time of diagnosis (47).

The solid and cystic papillary epithelial neoplasms of the pancreas almost always show a variable degree of hemorrhagic degeneration. Hemorrhage may progress to cystic changes within the lesions in approximately 20% of cases (48). This change is usually associated with an irregular inner margin and a thick wall (48).

Islet cell tumors can demonstrate variable amounts of cystic degeneration, which, in isolated cases, may be the predominant component. This type of islet cell tumor still demonstrates a hypervascular solid component at CT

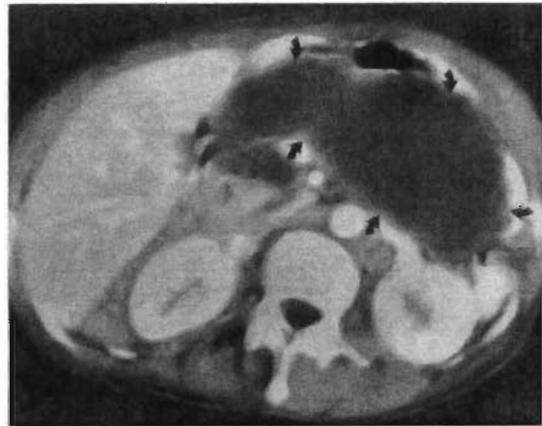


Figure 15. Mucin-hypersecreting adenocarcinoma of the pancreas. Contrast-enhanced CT demonstrates extensive ectasia of the pancreatic duct (arrows) and mild irregularity along the posterior wall that corresponds to the malignant epithelium and papillary projections that were seen at histologic analysis.

and MR imaging, and the walls surrounding the cystic cavities are thicker than those expected with mucinous cystic neoplasms. Calcification, vascular invasion, and metastasis to the liver (which can also have a cystic appearance) may also be seen in islet cell tumors (49,50).

Cystic metastases to the pancreas are not uncommon, occurring in 3%-12% of cases in autopsy series (47). These tumors are usually asymptomatic during life and include metastases of renal cell carcinoma, melanoma, lung tumors, breast carcinoma, hepatocellular carcinoma, and ovarian carcinoma (47,51). Of these, ovarian carcinoma would be the most likely to manifest as a predominantly cystic mass. A known history of primary malignant disease, combined with the presence of metastatic foci, are helpful clues in making the diagnosis.

Serous cystadenomas are distinguished from mucinous cystic neoplasms on the basis of their size and generally greater number of small cysts (1). Less than 5% of serous cystadenomas (1,45) have a few large cysts and may mimic mucinous cystic neoplasm. Serous cystadenomas are virtually all benign lesions, al-

though there are reported cases of a malignant variant (52). In our experience, the difficulty in differentiating these lesions radiologically almost always occurs with small (less than 5-cm) lesions (Rao P, Buetow PC, unpublished data, 1996) (Fig 12).

Sarcomas, although extremely unusual pancreatic neoplasms, may manifest as partially or predominantly cystic masses. They may appear as well-circumscribed masses or may demonstrate a more aggressive appearance (47,53).

■ TREATMENT AND PROGNOSIS

Because mucinous cystic neoplasms of the pancreas are considered as low-grade malignant neoplasms, the treatment of choice is complete surgical excision. In one series when the tumor was completely excised, over 90% of patients survived without evidence of recurrence or metastatic disease (mean follow-up, 9.4 years) (26). If, however, these tumors are incompletely excised, marsupialized, or drained, the prognosis is less favorable. The 5-year survival rate for all patients with malignant tumors was 68% in a report by Hodgkinson and colleagues (54,55), while our series demonstrated a 74.3% survival for all those patients that were followed up for 5 years (regardless of their surgical treatment) (26). In all cases, irrespective of histologic characteristics, the prognosis for mucinous cystic neoplasm is far better than that for ductal adenocarcinoma (which is associated with an overall 5-year survival rate of less than 1%) (2,4,19,26,28).

■ SUMMARY

Mucinous cystic neoplasms of the pancreas are low-grade malignant tumors that typically arise within the tail of the pancreas and manifest as large, multiloculated, cystic masses in middle-aged women. There are, however, variable radiologic manifestations, which include a unilocular mass and a predominantly solid lesion with focal cysts. In most cases, US and CT are the mainstays for radiologic evaluation, with MR imaging having a complementary role.

The radiologic appearance of this lesion overlaps with that of other cystic masses of the pancreas. The most problematic differentiation is between a unilocular mucinous cystic neoplasm and a pancreatic pseudocyst. Sometimes these entities may be differentiated on clinical grounds or by noting associated findings of pancreatitis; but, in certain cases, fluid aspiration followed by surgery may be necessary for definitive diagnosis. Such differentiation is important because complete surgical excision is the treatment of choice for mucinous cystic neoplasms. Once this tumor completely resected, long-term survival can be expected in the overwhelming majority of cases.

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