Acute pancreatitis is one of the most common conditions for which emergent imaging is indicated. Alcohol consumption and cholelithiasis are the most common causes of acute pancreatitis in adults, whereas the majority of cases in children are idiopathic or secondary to trauma. A wide variety of structural and biochemical abnormalities may also cause pancreatitis. Although in some cases it is difficult to identify the specific cause of the disease radiologically, certain uncommon types of acute or chronic pancreatitis may have unique imaging features that can help the radiologist make an accurate diagnosis. These unusual types include autoimmune pancreatitis, groove pancreatitis, tropical pancreatitis, hereditary pancreatitis, and pancreatitis in ectopic or heterotopic pancreatic tissue. Pancreatitis may occasionally be seen in association with cystic fibrosis or pancreas divisum, or secondary to worm infestation of the pancreaticobiliary tree (eg, by *Ascaris lumbricoides*). In addition, primary pancreatic and duodenal masses may occasionally manifest as acute or recurrent acute pancreatitis. Knowledge of the classic imaging findings of these entities allows prompt recognition of the relevant pathologic condition, thereby preventing misdiagnosis and subsequent inappropriate or delayed management.

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**Introduction**

Pancreatitis is the most common pancreatic disease in children and adults and one of the most common causes of morbidity and mortality worldwide. In the United States, more than 300,000 patients are hospitalized annually with acute pancreatitis (1). Over one-half of cases of acute pancreatitis in adults are related to cholelithiasis or alcohol consumption, whereas trauma, viral infections, and systemic disease account for the majority of cases in children. The incidence of chronic pancreatitis is between three and nine cases per 100,000 persons per year, accounting for more than 120,000 outpatient visits and 50,000 hospitalizations annually (2, 3). Alcohol consumption accounts for the majority (80%) of cases of chronic pancreatitis in adults in developed countries, whereas malnutrition is the most common cause worldwide (4). Chronic pancreatitis is a disabling illness in children that carries a high morbidity rate due to exocrine and endocrine pancreatic insufficiency, resulting in stunted growth. Idiopathic pancreatitis is considered to be the most common cause of chronic pancreatitis in children (up to 30% of cases). In truth, however, hereditary and tropical pancreatitis are responsible for the majority of cases of chronic childhood pancreatitis in the United States and worldwide, respectively, many of which may fall under the “idiopathic” category if unrecognized (5). Biliary tract abnormalities account for the largest proportion (47%) of cases of childhood pancreatitis in Japan, with congenital dilatation of the common bile duct (CBD) being the most common abnormality (90% of cases) (6). To prevent complications, active and vigorous investigation is required in cases of so-called idiopathic pancreatitis, especially in children and in patients with recurrent pancreatitis.

Traditionally, computed tomography (CT) has been used to help confirm the diagnosis, assess disease severity, detect complications, and provide a “road map” for interventional procedures. CT also plays a pivotal role in evaluating the impact of various medical and surgical treatments. In most cases, acute and chronic pancreatitis do not have specific imaging characteristics on the basis of which an etiologic factor can be identified. However, a handful of causes do have specific clinical and imaging features that may be helpful in identifying the cause and thus may have a significant impact on management (Table). In cases with no underlying metabolic or systemic predisposing factors for acute or recurrent pancreatitis, special attention to these ancillary findings and critical reevaluation of these uncommon but potential causes are required.

The advent of endoscopic ultrasonography (US) and MR cholangiopancreatography has significantly enhanced the diagnostic power of CT. In addition, ERCP has played an important role in revealing many occult causes of pancreatitis.

In this article, we discuss and illustrate the clinical and radiologic features of (a) various uncommon types of acute and chronic pancreatitis (autoimmune pancreatitis, groove pancreatitis, tropical pancreatitis, hereditary pancreatitis, pancreatitis in ectopic or heterotopic pancreatic tissue, Ascaris-induced pancreatitis, pancreatitis in cystic fibrosis); (b) congenital pancreatic anomalies associated with pancreatitis (pancreas divisum, annular pancreas); and (c) some unusual causes of pancreatitis (pancreatic adenocarcinoma, duodenal villous adenoma).

**Autoimmune Pancreatitis**

Autoimmune pancreatitis (also known as lymphoplasmacytic sclerosing pancreatitis, chronic sclerosing pancreatitis, pseudotumorous pancreatitis, or nonalcoholic duct-destructive chronic pancreatitis) is a type of chronic pancreatitis that is characterized by an autoimmune inflammatory process, with lymphoplasmacytic infiltration associated with fibrosis of the pancreas. First described by Yoshida et al in 1995 (7), autoimmune pancreatitis accounts for 1.8%–11% of all cases of chronic pancreatitis (8, 9). The unique clinical features of this entity include the absence of classic acute attacks of pancreatitis, elevated immune markers (immunoglobulin G4), dramatic response to steroid therapy, and diagnostic difficulty in differentiating it from pancreatic cancer. Patient age at presentation varies from 14 to 77 years, with most patients being over 50 years of age (10, 11). Men are affected twice as frequently as women (12). Signs and symptoms at presentation include jaundice (63% of cases), abdominal pain (35%), weight loss (35%), and diabetes mellitus (42%−76%) (9, 10). Presentation with acute pancreatitis or severe abdominal pain is unusual. The condition may be associated with various extrapancreatic manifestations of an autoimmune nature. These associated manifestations (if seen) may further strengthen the autoimmune basis of the condition and may help in making the correct diagnosis in the appropriate clinical setting. Common extrapancreatic manifestations...
<table>
<thead>
<tr>
<th>Condition</th>
<th>Clinical Features</th>
<th>Radiologic Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Autoimmune pancreatitis</td>
<td>Absence of the classic acute attacks of pancreatitis, elevated immune markers, dramatic response to steroid therapy</td>
<td>Diffusely enlarged gland with loss of lobular architecture, a “sausage” shape, and a peripheral “rind” of hypoa attenuation; nondilated or diffusely narrowed pancreatic duct, extrapancreatic autoimmune manifestations</td>
</tr>
<tr>
<td>Groove pancreatitis</td>
<td>Duodenal and biliary obstruction; symptoms overlap with those of pancreatic cancer, frequently leading to misdiagnosis</td>
<td>Soft tissue within the pancreaticoduodenal groove with or without delayed enhancement, small cystic lesions along the medial duodenal wall</td>
</tr>
<tr>
<td>Tropical pancreatitis</td>
<td>Young age at onset, associated with malnutrition, regional predisposition in tropical countries, rapidly progressive course with an increased risk of adenocarcinoma</td>
<td>Multiple large calculi within a dilated pancreatic duct (&gt;80% of cases); discrete, dense calculi up to 5 cm in size (vs small speckled intraductal calculi in alcohol-related chronic pancreatitis)</td>
</tr>
<tr>
<td>Hereditary pancreatitis</td>
<td>Young age at onset, at least two acute attacks of pancreatitis with no underlying cause, family history of pancreatitis in a first- or second-degree relative</td>
<td>Acute: nonspecific; chronic: significant pancreatic atrophy, pancreatic calcifications, and calculi</td>
</tr>
<tr>
<td>Pancreatitis in ectopic or heterotopic pancreatic tissue</td>
<td>Pancreatitis rarely diagnosed clinically and radiologically; may manifest as abdominal pain (77% of cases), abdominal fullness (30%), or malena (24%)</td>
<td>Centrally umbilicated nodule in the gastric mucosa at contrast material–enhanced upper gastrointestinal study; CT: oval or round masses in the gastric wall that follow the enhancement pattern of pancreatic tissue (80% in the prepyloric region along the greater curvature), nonspecific inflammatory changes relative to the gastric or intestinal wall</td>
</tr>
<tr>
<td>Ascaris-induced pancreatitis</td>
<td>Most common parasitic infection worldwide; <em>Ascaris lumbricoides</em> is the most commonly implicated parasite in pancreatitis; most commonly seen in middle-aged adult women (male-to-female ratio, 1:3); may result in biliary colic, cholangitis, acute cholecystitis, hepatic abscess, acute pancreatitis, or septicemia; usually mild but can be fatal</td>
<td>Roundworm appears as an echogenic tubular structure with a central hypoechoic tubular area representing the worm’s digestive tract; live worms may be seen within the bile duct; magnetic resonance (MR) imaging: linear hypointense filling defect within a dilated biliary radical associated with findings of acute pancreatitis, worms rarely seen within the main pancreatic duct (MPD)</td>
</tr>
<tr>
<td>Pancreatitis in cystic fibrosis</td>
<td>Exocrine pancreatic insufficiency more common, full-blown acute pancreatitis rare (1.2% of cases), pancreatitis may rarely be the first manifestation of cystic fibrosis</td>
<td>Fatty replacement of the pancreatic parenchyma with or without glandular atrophy (56%–93% of cases at CT), calcification (7%), and cyst formation (pancreatic cystosis); peripancreatic inflammation and fluid collections in the typical acute setting</td>
</tr>
<tr>
<td>Pancreas divisum associated with pancreatitis</td>
<td>Seen in young or middle-aged adults with recurrent acute pancreatitis or chronic relapsing pancreatitis with no other underlying cause</td>
<td>MRCP and ERCP are optimal for diagnosis; MRCP can adequately depict lack of communication between the dorsal and ventral ducts, independent drainage sites, and a dominant dorsal duct</td>
</tr>
<tr>
<td>Uncommon obstructive causes of pancreatitis</td>
<td>Pancreatic (carcinoma, lymphoma, metastases) or duodenal (adenoma, lipoma) neoplasms rarely manifest with acute pancreatitis; high level of suspicion in middle-aged or elderly patients with chronic signs and symptoms (eg, weight loss, abdominal pain)</td>
<td>Abrupt change in pancreatic duct caliber with focal pancreatitis beyond the site of obstruction, disproportion between the size of the body and head of the pancreas, regional lymphadenopathy, vascular invasion, MRCP and contrast-enhanced MR imaging are sensitive in depicting the lesion</td>
</tr>
</tbody>
</table>

Note.—ERCP = endoscopic retrograde cholangiopancreatography, MRCP = MR cholangiopancreatography.
include biliary diseases such as sclerosing cholangitis and primary biliary cirrhosis (68%–88% of cases); inflammatory bowel disease (most commonly, ulcerative colitis (17%); Sjögren syndrome; sialadenitis (12%–16%); renal involvement (3.4%–35%); and retroperitoneal fibrosis (3%–8%) (13,14).

At pathologic analysis, focal or diffuse inflammation of the gland with a predominant lymphoplasmacytic component is seen around the interlobular pancreatic ducts (15). Diffuse sclerosis and fibrosis at a later stage may result in a firm “mass-forming” pancreas that, along with a confusing clinical picture involving weight loss and sometimes elevated carinoembryonic antigen and cancer antigen 19-9 levels, can often lead to an incorrect diagnosis of pancreatic cancer (16). This misdiagnosis can in turn lead to inappropriate and sometimes harmful management.

CT is the investigative modality of choice. Diffuse enlargement of the gland with loss of lobular architecture (“featureless” pancreas) and homogeneously iso- or hypoattenuating parenchyma with a nondilated or diffusely narrowed pancreatic duct are characteristic features (Fig 1). However, the imaging appearance of autoimmune pancreatitis can vary widely depending on the degree of fibrosis and inflammatory infiltrate. Associated findings such as a peripheral rim of hypoenhancement that may demonstrate delayed enhancement, minimal peripancreatic stranding, involution of the pancreatic tail, and regional lymphadenopathy may also be seen. Calcifications, pseudocyst formation, and vascular encasement are rare (17). The focal form of the disease may manifest as a pancreatic mass, most commonly in the head of the pancreas, simulating pancreatic carcinoma.

At ERCP, the hallmark finding is focal, segmental, or diffuse narrowing of the pancreatic duct with nonvisualization of side branches. Associated biliary tract abnormalities such as a smooth stricture of the distal extrapancreatic CBD with dilatation of the proximal portion may be seen in up to 90% of patients (18). Strictures of more proximal bile ducts resembling primary sclerosing cholangitis may also be seen. Sclerosing cholangitis associated with autoimmune pancreatitis has been classified into various subtypes on the basis of the cholangiographic appearance (19). Differentiation of sclerosing cholangitis associated with autoimmune pancreatitis from primary sclerosing cholangitis is important: The latter demonstrates a poor response to steroid treatment, whereas the former responds dramatically.

MR imaging may reveal diffuse enlargement of the gland and variable T2 signal with homogeneous or heterogeneous enhancement. A peripheral rim of hypoenhancement and peripancreatic inflammatory changes may also be seen (Fig 1b–1f). MR cholangiopancreatography may reveal diffuse narrowing of the pancreatic duct, strictures of the pancreaticobiliary tree, and other associated biliary abnormalities.

Other extrapancreatic manifestations associated with autoimmune pancreatitis include retroperitoneal fibrosis; involvement of the renal parenchyma, perirenal tissue, renal sinus, or renal pelvic wall; pulmonary involvement with parenchymal nodules; infiltrates or mediastinal adenopathy; and, rarely, autoimmune prostatitis. Renal manifestations are regarded as an important clue in diagnosing autoimmune pancreatitis and in differentiating it from other types of pancreatitis and pancreatic malignancy (14). The renal parenchyma may show small peripheral cortical nodules, round lesions, well-defined wedge-shaped lesions, or diffuse patchy involvement.

**Figure 1.** Autoimmune pancreatitis in a 38-year-old man. (a) Contrast-enhanced CT scan through the upper abdomen reveals diffuse enlargement of the pancreas with a peripheral rim of hypoenhancement (black arrow), involvment of the pancreatic tail, and narrowing of the splenic vein (white arrow). Note also the associated multiple bilateral hypoenhancing renal lesions (arrowheads), which represent foci of tubulointerstitial nephritis. (b, c) Axial non-fat-suppressed (b) and fat-suppressed (c) T2-weighted MR images (repetition time msec/echo time msec = 1030/80, 5-mm section thickness) reveal diffuse enlargement of the pancreas, which has a featureless appearance with loss of lobular architecture and narrowing of the MPD (arrow in b). Pseudocyst formation, a rare manifestation, is also seen (arrow in c). (d–f) Axial precontrast (d) and postcontrast arterial phase (e) and venous phase (f) fat-suppressed T1-weighted MR images (3.35/1.5, 3-mm section thickness) reveal hyperintensity within the pancreatic tail and heterogeneous postcontrast enhancement. The low-signal-intensity peripheral rim (arrow) is a characteristic finding. (g, h) ERCP images reveal smooth narrowing of the distal CBD (arrow in g) with diffuse “irregular” narrowing of the MPD (arrowheads in h). Laboratory evaluation revealed a significantly elevated IgG4 level of over 5 g/L (normal range, 0.06–1.21 g/L), a finding consistent with autoimmune pancreatitis. Steroid therapy was initiated. (i) ERCP image obtained 6 months after the completion of steroid therapy shows resolution of the biliary stricture and normalization of the contour and caliber of the MPD (arrows). (j) Contrast-enhanced CT scan obtained 12 months after the completion of steroid therapy shows a reduction in the size of the pancreas and resolution of inflammatory changes (arrow). The renal lesions are also less conspicuous (arrowheads).
At histopathologic analysis, these multifocal renal masses are composed of lymphoplasmacytic infiltrates and are usually bilateral.

The most important diagnostic option in the differential diagnosis for focal mass-forming autoimmune pancreatitis is pancreatic cancer. Vascular encasement or abrupt narrowing of the pancreatic duct with significant dilatation of the proximal pancreatic duct and marked atrophy of the parenchyma proximal to the mass should favor a diagnosis of pancreatic carcinoma. Various diagnostic criteria have been proposed for the diagnosis of autoimmune pancreatitis. Abnormally elevated levels of IgG4 or serum gamma globulin or the presence of autoantibodies such as ALA (antilactoferrin antibody), ACA II (anti-carbonic anhydrase II antibody), ASMA (anti-smooth muscle antibody), and ANA (antinuclear antibody) are required to confirm the diagnosis when characteristic imaging findings are seen (20). Occasionally, inflammation associated with pancreatic cancer may partially respond to corticosteroid therapy. Thus, histopathologic confirmation with pancreatic biopsy may be required when the laboratory values are indeterminate or when there is incomplete resolution of the mass in response to steroid therapy (21). Positron emission tomography with 2-[fluorine 18]fluoro-2-deoxy-D-glucose also has a pivotal supplemental role in differentiating between autoimmune pancreatitis and pancreatic cancer. A longitudinal heterogeneous pattern with multiple areas of uptake within the gland and extrapancreatic uptake in the lachrymal or salivary gland, biliary system, retroperitoneal space, or prostate gland favors a diagnosis of autoimmune pancreatitis. In contrast, uptake in pancreatic cancer is nodular, homogeneous, and more localized within the gland, without any extrapancreatic uptake (21). The diffuse form of autoimmune pancreatitis also needs to be differentiated from lymphoma and mild acute pancreatitis. Differentiation may be difficult on the basis of imaging findings alone. The presence of concurrent multi-systemic disease with massive adenopathy would favor a diagnosis of lymphoma. Similarly, acute peripancreatic inflammatory signs would favor a diagnosis of acute pancreatitis (22).

Corticosteroid therapy is considered to be the standard treatment for autoimmune pancreatitis, although spontaneous resolution of this entity has also been reported on occasion (23). Follow-up imaging demonstrates significant reduction in the bulk of prominent pancreatic parenchyma and normalization of duct caliber as early as 4–5 weeks after steroid therapy (22).

**Figure 2.** Groove pancreatitis with cystic dystrophy of the duodenal wall. Drawing illustrates the disease process in groove pancreatitis. Inflammation is predominantly centered in the pancreaticoduodenal groove, with multiple cystic lesions within the medial wall of the duodenum (D).

**Groove Pancreatitis**

Groove pancreatitis (Figs 2, 3) is a rare form of chronic pancreatitis that may mimic pancreatic carcinoma. The term *pancreaticoduodenal groove* refers to the potential space between the head of the pancreas, the duodenum, and the CBD. Two forms of groove pancreatitis have been described: the “segmental” form, which involves the pancreatic head with development of scar tissue within the groove; and the “pure” form, which affects the groove only, sparing the pancreatic head (24). The clinical manifestation is related primarily to duodenal and biliary obstruction. Although biliary strictures are reported in up to 50% of cases of groove pancreatitis, symptoms due to biliary obstruction seldom precede those due to duodenal obstruction (24).

Pathologic analysis reveals scar tissue in the pancreaticoduodenal groove, with sparing of the pancreatic parenchyma in the pure form and variable involvement of the pancreatic head in the segmental form. The pathogenesis of groove pancreatitis is still unclear. Several factors such as peptic ulcer disease, gastric resection, true duodenal wall cysts, pancreatic heterotopia, and disturbance of flow in the MPD are related to the development of groove pancreatitis (25). It is unclear whether groove pancreatitis and cystic dystrophy of the duodenum are distinct entities or part of the same spectrum. Hence, a broad category labeled “paraduodenal pancreatitis” has been proposed to include groove pancreatitis, cystic dystrophy of the duodenal wall, and paraduodenal wall cysts, all of which occur in and
Figure 3. Groove pancreatitis with cystic dystrophy of the duodenal wall. (a) Transverse US image through the pancreas (P) demonstrates a sheetlike hypoechoic area in the pancreaticoduodenal groove with areas of cystic change (arrowhead). (b, c) Venous phase CT scans show a hypoattenuating area in the pancreaticoduodenal groove (arrow in b) with inflammatory stranding within the surrounding fat and in the right anterior pararenal-paraduodenal space (arrows in c). P = pancreas. (d, e) Axial (d) and coronal (e) non-fat-suppressed T2-weighted MR images (1030/80, 5-mm section thickness) show hyperintense cystic lesions (arrowhead) within the thickened duodenal wall with minimally hyperintense soft tissue in the groove (arrow). D = second part of duodenum, P = pancreatic head. (f) Coronal delayed contrast-enhanced fat-suppressed T1-weighted MR image (3.4/1.5, 3.75-mm section thickness) obtained 5 minutes after the injection of gadolinium-based contrast material shows partially enhanced cystic lesions (arrow) in the pancreaticoduodenal groove. Histopathologic analysis of multiple surgical biopsy specimens showed pancreatic heterotopia, Brunner gland hyperplasia, and pancreatitis, findings that are consistent with the diagnosis of groove pancreatitis with cystic dystrophy of the duodenal wall.
around the minor papilla and have several features in common such as the presence of dilated ducts and cysts in the duodenal wall, Brunner gland hyperplasia, and hamartomatous pancreatic tissue located in the groove (26). These entities may resemble pancreatic cancer both clinically and radiologically.

At CT, the classic finding is soft tissue within the pancreaticoduodenal groove (Fig 3b, 3c); this tissue may demonstrate delayed enhancement. Small cystic lesions may be seen along the medial wall of the duodenum. MR imaging is better in characterizing these findings and may demonstrate a sheetlike mass in the pancreaticoduodenal groove that is usually hypointense relative to the pancreas on T1-weighted images, is usually iso- to slightly hyperintense relative to the pancreas on T2-weighted images, and may show delayed enhancement following the administration of gadolinium-based contrast material (Fig 3d–3f). The segmental form of groove pancreatitis involves the pancreatic head in addition to the scar tissue in the groove and produces a masslike appearance of the head, a finding that corresponds histologically to chronic pancreatitis (25). Associated duodenal wall thickening may also be seen. The presence of cystic lesions within the thickened wall of the second portion of the duodenum would favor a diagnosis of cystic dystrophy of the duodenal wall, which, as mentioned earlier, may represent a form of paraduodenal pancreatitis.

The differential diagnosis for groove pancreatitis depends on whether the pure form or the segmental form is being considered. The most important diagnostic option in the differential diagnosis for the segmental form is pancreatic adenocarcinoma. The imaging characteristics of these two entities may overlap considerably. In particular, owing to its significant fibrous component, the scirrhous variant of pancreatic adenocarcinoma may demonstrate delayed enhancement similar to that shown by groove pancreatitis. The presence of vascular invasion is considered to be the most useful sign in differentiating pancreatic carcinoma from groove pancreatitis. MR cholangiopancreatography may reveal a smooth stricture of the distal intrapancreatic portion of the bile duct in patients with groove pancreatitis, as opposed to an abrupt irregular ductal stricture in patients with pancreatic carcinoma.

A variety of pancreatic lesions may mimic the pure form of groove pancreatitis. These lesions include groove carcinoma, duodenal or periampullary carcinoma, pancreatic groove neuroendocrine tumors, and acute pancreatitis with inflammatory changes in the groove. Differentiation from pancreatic groove carcinoma is almost impossible on the basis of clinical and imaging features alone, although the presence of cystic lesions within the mass or a thickened duodenal wall would favor a diagnosis of groove pancreatitis (27,28).

Acute pancreatitis with fluid collections and inflammation in the pancreaticoduodenal groove may be relatively easy to identify, since the associated changes evolve rapidly on serial images and demonstrate high signal intensity on T2-weighted images (29). Rarely, neuroendocrine tumors such as gastrinomas may occur within the groove. These tumors can be differentiated from groove pancreatitis by virtue of their hypervascularity on postcontrast images and their hyperintensity on T2-weighted images.

**Tropical Pancreatitis**

Tropical pancreatitis is a variant of chronic pancreatitis that is characterized by a constellation of findings including young age at onset, association with malnutrition, regional predisposition in tropical countries, rapidly progressive course with severe pancreatitis, the presence of large intraductal calculi, and an increased risk of adenocarcinoma. Tropical pancreatitis was first described in 1959 in Indonesia (30). Since that time, a number of cases have been reported in Asia, Africa, and South America, with a relatively large number of cases occurring in the southern state of Kerala in India.

A number of factors are implicated in the etiopathogenesis of tropical pancreatitis, including protein energy malnutrition, pancreatic ductal anomalies, food toxicities such as chronic cyanide toxicity from cassava, and possible genetic predisposition (SPINK 1 N34S mutation and CFTR mutation) (31,32). Protein energy malnutrition is thought to suppress pancreatic function, making the gland more susceptible to exogenous toxins (33). However, malnutrition is argued by some to be the effect of the disease rather than the cause. This juvenile nonalcoholic chronic relapsing form of pancreatitis typically manifests as abdominal pain (80%–90% of cases), weight loss, glucose intolerance, or frank diabetes mellitus. Patient age at disease onset is variable, ranging from infancy to adulthood (mean age, 12.5 years) (34). A male predilection (male-to-female ratio, 1.6–5:1) has been observed. Nearly two-thirds of affected patients develop fibrocystic pancreatic diabetes within a decade of onset (33). The characteristic features of this entity are absence of association with alcohol consumption, biliary tract disease, and absence of other biochemical or structural predisposing factors for pancreatitis.
At imaging, the hallmark finding is the presence of multiple large pancreatic calculi within a dilated pancreatic duct; these calculi may be detected at conventional radiography, US, or CT (Fig 4). Ductal dilatation and calculi are known to occur in more than 80% of patients, with parenchymal atrophy occurring in nearly 50% (35). The intraductal calculi in tropical pancreatitis are discrete, dense, and up to 5 cm in size. They can
Figure 5. Hereditary pancreatitis in an 18-year-old man with recurrent episodes of idiopathic acute pancreatitis. Axial (a) and coronal (b) unenhanced CT scans reveal multiple focal areas of punctate calcifications throughout the pancreatic parenchyma (arrows in a, arrowheads in b), findings that are nonspecific and suggest chronic calcific pancreatitis. However, the young age of the patient, the absence of any known predisposing factors, and the family history of pancreatitis favor a diagnosis of hereditary pancreatitis.

As in tropical pancreatitis, acute attacks usually begin in childhood (5–10 years of age), with age at onset varying from infancy to the 5th or 6th decade of life (38). By the teenage years, these recurrent attacks result in chronic pancreatitis with parenchymal and ductal calcifications, duct destruction, and secondary exocrine and endocrine insufficiency. However, no obvious predisposing factors for chronic pancreatitis are identified.

The imaging manifestations of hereditary pancreatitis often resemble those of tropical pancreatitis. Imaging manifestations during an acute attack may be nonspecific; therefore, in this setting, hereditary pancreatitis may not be diagnosed with confidence. However, the chronic variety is characterized by significant pancreatic atrophy, parenchymal and ductal calcifications, and calculi (Fig 5). Hereditary pancreatitis should be suspected in any patient who has suffered at least two attacks of acute pancreatitis for which there is no explanation (absence of anatomic anomalies, ampullary or MPD strictures, trauma, viral infection, gallstones, alcohol consumption, drug use, or hyperlipidemia). The diagnosis may also be considered in patients with unexplained (idiopathic) chronic pancreatitis, patients with a family history of pancreatitis in a first- or second-degree relative, and children with an unexplained episode of pancreatitis that required hospitalization (39).
Genetic diagnosis of hereditary pancreatitis is vitally important for patients and their extended families. Mutations of various genes like PRSS1 and CFTR have been demonstrated. However, no mutations are seen at genetic analysis in up to 30% of families with hereditary pancreatitis (40). The role of imaging is to exclude underlying structural causes of pancreatitis. Establishing a diagnosis of hereditary pancreatitis early in the patient’s life is important due to its tendency to recur and the increased risk of pancreatic cancer. Unlike chronic pancreatitis due to other causes, in which the risk of pancreatic adenocarcinoma is increased 15–25-fold, hereditary pancreatitis carries a 50- to 70-fold increased risk of pancreatic cancer within 7–30 years of disease onset (41,42). The cumulative lifetime risk is estimated to be 40% by the age of 70 years, and is even higher in smokers and alcoholic patients. Therefore, screening should start at 30 years of age (43) and should usually be performed with endoscopic US. Heterogeneous parenchyma and distinct hypoechoic nodules and masses at endoscopic US are considered to be positive findings that should raise suspicion for pancreatic intraductal neoplasia in a high-risk patient (44). However, these imaging findings overlap with those of chronic pancreatitis and therefore require confirmation with ERCP. If ERCP findings are normal, regular endoscopic US surveillance at 6–12-month intervals is warranted (44).

**Pancreatitis in Ectopic or Heterotopic Pancreatic Tissue**

Heterotopic pancreas (pancreatic choristoma) is defined as the presence of abnormally located pancreatic tissue with its own ductal system, with no vascular, neural, or anatomic contact with the normal pancreas. It is the most common heterotopia within the gastrointestinal system, with a prevalence of 0.55%–14% in autopsy series (45). The entity is usually asymptomatic but can manifest in adulthood with various complications (46).

The proposed pathogenesis is metaplasia of multipotent endodermal cells. Displacement and transplantation of pancreatic cells to adjacent structures during embryonic development has also been postulated. The most common locations for heterotopic pancreatic tissue include the duodenum (28% of cases), stomach (26%), and jejunum (16%) (46). Pancreatic heterotopia in the stomach has a prevalence on the order of 1%–2% and commonly occurs in a submucosal location (75% of cases), but can also be present within the muscularis propria and serosa (47). Less common sites of origin include the following: Meckel diverticulum, ileum, ileal and jejunal diverticula, gallbladder, fallopian tubes, umbilicus, mediastinum, esophagus, spleen, and omentum (48). The ectopic tissue appears as a pale yellow lobulated nodule or mass at gross examination, with the centrally umbilicated nodule representing the opening for the pancreatic duct. Varying proportions of acinar elements, ducts, and smooth muscle elements are seen at histologic analysis.

The majority of patients are asymptomatic at clinical examination, with the lesions being incidentally detected at surgery or autopsy. Common clinical manifestations include abdominal pain similar to that from peptic ulcer disease (77% of cases), abdominal fullness (30%), and melena (24%). Other complications include biliary and intestinal obstruction, massive gastrointestinal bleeding, pancreatitis, and malignant transformation (49,50). Pancreatitis is a complication that is usually detected at microscopy and is rarely diagnosed clinically or radiologically. Pancreaticolithiasis has also been reported in ectopic pancreas in the stomach and transverse colon (51) and can manifest over a wide age range (6–77 years) (52).

A barium swallow upper gastrointestinal study demonstrates nonspecific fold thickening with the characteristic appearance of a centrally umbilicated nodule in the gastric mucosa within the gastric heterotopic pancreatic rest. These lesions rarely exceed 3–4 cm in size. A polyp, a nodule, or fold thickening due to other causes cannot be reliably differentiated from an ectopic pancreas. CT may reveal nonspecific extrapancreatic inflammatory changes in relation to the gastric or intestinal wall with mural thickening. CT may also demonstrate oval or round masses in the gastric wall with an enhancement pattern similar to that of pancreatic tissue (Fig 6). In the stomach, the heterotopic pancreatic tissue characteristically occurs in the prepyloric region along the greater curvature (up to 90% of cases) (53). Esophageal pancreatic heterotopia is associated with a wide variety of
heterotopic pancreatic tissue such as diverticulitis or enteritis. A characteristic location within the stomach and the presence of central umbilication should raise suspicion for heterotopic pancreatic tissue. If the patient is asymptomatic and an accurate diagnosis can be made at imaging and endoscopy, management is conservative. However, if the patient is symptomatic or differentiation from other benign or malignant processes is not possible, resection of the involved segment is usually performed, such resection being curative.

Ascaris-induced Pancreatitis

Ascariasis is the most common parasitic infection worldwide, affecting an estimated 1 billion people. The disease is endemic to Southeast Asia,
Africa, China, and Latin America (57,58). The roundworm *Ascaris lumbricoides*, which usually inhabits the small intestine, may occasionally migrate through the ampulla of Vater into the pancreaticobiliary tree. This migration may occur through an abnormally open ampullary orifice due to either preexisting biliary tract disease or endoscopic sphincterotomy. Adult worms are 15–50 cm in length and 3–6 mm thick. The presence of the worm in the biliary tree and pancreatic duct may result in biliary colic, cholangitis, acute cholecystitis, hepatic abscess, acute pancreatitis, and even septicemia.

Biliary-pancreatic ascariasis most commonly affects adult women (male-to-female ratio, 1:3), with a mean age at presentation of 35–42 years (59). *A. lumbricoides* is the most common parasitic cause of pancreatitis, which is usually due to the presence of the worm in the bile duct. On rare occasions, the worm may enter the MPD, resulting in severe pancreatitis. *Ascaris*-induced pancreatitis is usually mild but can be fatal in rare instances (60).

At US, the roundworm appears as an echogenic tubular structure without distal acoustic shadowing within the bile duct or pancreatic duct, with a central hypoechoic tubular area representing the worm’s digestive tract. Live worms may occasionally be observed moving within the bile ducts. Associated findings of biliary or pancreatic duct dilatation, pancreatic and peripancreatic inflammatory changes, and cholecystitis may be evident. Rarely, hepatic abscess formation may occur. MR cholangiopancreatography reveals a linear hypointense filling defect within a dilated biliary radical, a finding that, when associated with acute pancreatitis, is diagnostic for *Ascaris*-induced pancreatitis (Fig 7). With these imaging appearances, the diagnosis is fairly straightforward.
There are two different schools of thought regarding the management of biliary ascariasis (59). The first recommends initial conservative management with endoscopic treatment in patients who do not respond to conservative therapy and who demonstrate persistent worms after 3 weeks of antihelminthic therapy (57). The second proposes ERCP with therapeutic clearance of the ducts shortly after presentation (58). Chronic biliary ascariasis has also been implicated in the development of oriental cholangihepatitis, an entity that is more commonly seen with clonorchiasis and opisthorchiasis.

**Pancreatitis in Cystic Fibrosis**

Cystic fibrosis is a common autosomal recessive inherited disease secondary to mutation of a gene-encoding chloride channel, resulting in impaired chloride transport through the epithelial cells of exocrine glands. It occurs in one in 2500 children (60). Patients commonly present with recurrent pulmonary infections and pulmonary insufficiency, which are the major causes of morbidity and mortality. However, gastrointestinal manifestations often precede the pulmonary manifestations, with exocrine pancreatic insufficiency being one of the most common (85%–90% of patients).

Patients present with signs and symptoms of exocrine pancreatic insufficiency such as failure to thrive, steatorrhea, fat intolerance, abdominal pain, bloating, and flatulence (61). Cystic fibrosis is the most common cause of exocrine pancreatic insufficiency in young patients. Endocrine insufficiency with glucose intolerance is seen in up to 50% of cases, with frank diabetes mellitus occurring in 13% (62). In spite of ongoing inflammation in the pancreas and histologic evidence thereof, full-blown acute pancreatitis is rare (1.2% of cases) (63). In addition, pancreatitis may on rare occasions be the first manifestation of cystic fibrosis, although the final outcome—gland destruction with fatty replacement—is in fact due to numerous episodes of recurrent pancreatitis (63).

At pathologic analysis, widespread loss of acinar cells with fatty replacement of the pancreas and interstitial fibrosis may be seen. Obstruction of the ducts caused by inspissated secretions is the primary event, resulting in an initial mild inflammatory reaction followed by progressive fibrosis, fatty change, calcification, and cyst formation.

Conventional radiography may reveal pancreatic calcifications that are indistinguishable from those in other forms of chronic pancreatitis. US may show a small pancreas with increased echogenicity secondary to fatty replacement of the glandular parenchyma. Focal hypoechoic areas, calcification, and cyst formation may also be seen. Fatty replacement with or without pancreatic glandular atrophy is the most common and characteristic finding at CT (up to 93% of cases) (64). The degree of pancreatic fatty infiltration correlates with the severity of exocrine dysfunction (64). Other CT findings include calcification (7% of cases); cyst formation; and abnormalities of the pancreatic duct, including strictures, beading, dilatation, and obstruction. The peripancreatic inflammation and fluid collections seen in typical acute pancreatitis are rare, likely because some pancreatic exocrine function is required for typical acute pancreatitis to manifest. Pancreatic cystosis is a rare manifestation of cystic fibrosis in which the entire pancreas is replaced by multiple cysts of varying size.

Findings at MR imaging are similar to those at CT, with variable T1 hyper- and hypointensity depending on the proportion of fatty infiltration and fibrosis, respectively (65,66). MR cholangiopancreatography is more sensitive in the detection of ductal changes. Evaluation of the pancreas with MR imaging and MR cholangiopancreatography has the inherent advantage of lack of ionizing radiation, especially in children and adolescents. The differential diagnosis for fatty replacement with atrophy of the pancreas is extensive and includes obesity, severe malnutrition, aging, Cushing syndrome, steroid therapy, Shwachman-Diamond syndrome, hemochroma-
Figure 8. Acute pancreatitis in a 20-year-old man with cystic fibrosis who presented with severe abdominal pain and borderline elevated serum amylase level. (a, b) Contrast-enhanced venous phase CT scans show fatty replacement of the pancreas with minimal residual normal parenchyma in the tail region anteriorly (arrowhead in a). Arrow indicates peripancreatic inflammatory changes and fluid, findings that are consistent with a diagnosis of acute pancreatitis. Note also the foci of calcification within the pancreatic head (arrowhead in b). (c, d) Contrast-enhanced venous phase CT scans obtained after resolution of the acute pancreatitis (d obtained at the level of the pancreatic head) reveal changes of fatty replacement and calcification in the pancreatic head (arrow), findings that are characteristic of cystic fibrosis.

tosis, and viral infections (64). However, a constellation of clinical and radiologic manifestations with foci of fat, calcification, and inflammatory changes around the pancreas helps establish the diagnosis of acute pancreatitis in cystic fibrosis. Extrapancreatic manifestations of cystic fibrosis may also aid in differentiating pancreatic cystosis from other causes of pancreatic cysts such as autosomal dominant polycystic kidney disease with pancreatic involvement, von Hippel–Lindau syndrome, cystic pancreatic neoplasms, and pancreatic lymphangioma (66).
Pancreas Divisum

Pancreas divisum is the most common congenital anomaly of the pancreatic ductal system. This condition results from the failure of fusion of the ventral and dorsal pancreatic anlagen. Pancreas divisum occurs in 4%-10% of the population and has been reported in 12%-50% of cases of so-called idiopathic acute pancreatitis in children (67). Women are affected slightly more frequently than men (68). Although controversy does exist regarding the cause-and-effect relationship between the divisum anomaly and pancreatitis, there is a definite association between the two entities (69). Therefore, pancreas divisum should be suspected in young or middle-aged adults who present with recurrent acute pancreatitis or chronic relapsing pancreatitis with no other obvious cause such as gallstones or alcohol consumption.

Pathophysiologically, a relative obstruction to the flow of pancreatic juice is generated because the majority of the gland empties through the dorsal duct (duct of Santorini) into the minor papilla, which is small. The ventral duct (duct of Wirsung), which opens into the major papilla, drains only the ventral pancreatic anlage, which forms the head and uncinate process (Fig 9). Symptoms of pancreatitis can sometimes develop even in patients with a small communication between the ventral and dorsal ducts, which drain separately into the duodenum, representing an anatomic variant (dorsal dominant duct syndrome).

US may reveal findings of pancreatitis such as a bulky hypoechoic pancreas with peripancreatic inflammation or pseudocyst formation. Endoscopic US is useful in demonstrating the course of the ventral and dorsal ducts. The “stack sign” is defined as visualization of the bile duct and pancreatic duct running parallel through the pancreatic head. Absence of the stack sign favors the diagnosis of pancreas divisum (70). The sensitivity, specificity, positive predictive value, and

Figure 9. Pancreas divisum. (a) Drawing illustrates complete pancreas divisum, with the dorsal duct (arrowhead) draining the majority of the gland into the minor papilla. The smaller ventral duct (arrow) drains the head and uncinate process of the pancreas into the major papilla; the CBD also drains into the major papilla. D = duodenum. (b) Drawing illustrates incomplete pancreas divisum, which is considered to be a normal variant. A short communication exists between the dorsal duct (arrowhead), which drains the majority of the gland into the minor papilla, and the smaller ventral duct (arrow), which drains into the major papilla along with the CBD. D = duodenum.
negative predictive value of endoscopic US in diagnosing pancreas divisum are 95%, 97%, 86%, and 99%, respectively (71).

CT is the first-line imaging modality for the diagnosis and staging of acute pancreatitis (Fig 10a). However, CT has traditionally been thought to be unreliable for depicting the pancreatic ductal anatomy. Nevertheless, the advent of multidetector CT allows acquisition of thinner sections and multiplanar reformation techniques, making possible the diagnosis of this ductal anomaly with reasonable accuracy. Multidetector CT has a sensitivity of 90%, a specificity of 98%, and a negative predictive value of up to 98% in depicting pancreas divisum (72). MR cholangiopancreatography with heavy T2 weighting has a very high sensitivity and specificity in demonstrating normal ductal anatomy and anomalies. In the appropriate clinical setting, this modality may obviate an invasive procedure like ERCP, which entails a small (5%) but definite risk of postprocedural pancreatitis. MR cholangiopancreatography usually reveals two separate noncommunicating ducts with separate drainage territories (Fig 10b, 10c). Associated findings
of acute pancreatitis such as a bulky pancreas, pancreatic necrosis, peripancreatic inflammatory changes, or pseudocysts may be seen. Changes of chronic pancreatitis with significant atrophy of the duct and dilatation of the dorsal duct may also be observed, especially if the ductal anomaly is unrecognized and untreated. The ventral duct is not identified in a substantial number of cases and in many cases may be markedly atrophic. Visualization of the MPD (dorsal duct) coursing anterior to the CBD before draining into the duodenum is also a valuable sign that should raise suspicion for pancreas divisum, especially when the ventral duct is not well visualized (Fig 11). Secretin stimulation may improve visualization of the ductal anomaly (73). The dorsal duct is usually prominent, with depiction of the relative functional duct obstruction.

Management of pancreas divisum is primarily aimed at relieving the functional obstruction of the minor papilla by means of endoscopic or surgical sphincterotomy of the papilla in symptomatic patients. Interestingly, only 5%–10% of patients with pancreas divisum ever present with symptoms related to pancreatitis (74).

**Annular Pancreas**

Annular pancreas is a rare congenital anomaly in which a ring of pancreatic tissue surrounds
the second part of the duodenum. It occurs in about one in 20,000 persons. There is a bimodal distribution, with one-half of cases manifesting during childhood (commonly with duodenal obstruction) and the other half manifesting in adults in the 4th to 5th decades of life (75). In adults, annular pancreas may be incidentally detected, or the patient may present with symptoms of peptic ulcer disease (24.8% of cases) or pancreatitis (13.3%) (76).

At imaging, the diagnosis is established with visualization of the annular pancreatic parenchymal tissue and the duct encircling the second part of the duodenum. MR imaging, especially with T1 weighting, is useful in depicting the anatomy. MR cholangiopancreatographic and ERCP findings of an aberrant pancreatic duct encircling the duodenum are corroboratory. The duct within the annular pancreas communicates with the MPD in most cases (85%) (77). Annular pancreas may be associated with a number of other congenital anomalies in children and with pancreas divisum and bile duct carcinoma in adults (78).

**Uncommon Obstructive Causes of Pancreatitis**

Both primary pancreatic tumors and duodenal lesions obstructing the ampulla can rarely manifest as either acute or recurrent acute pancreatitis. Common pancreatic masses that may result in pancreatitis include intraductal papillary mucinous tumor, adenocarcinoma, and metastases. Duodenal lesions that may result in pancreatitis include duplication cysts, lipoma, duodenal diverticula, and adenomas. Any cause of obstruction to the MPD, including herniation of the pancreas through the diaphragmatic hiatus hernia, can also result in pancreatitis. The presence of intraductal stones is unusual in obstruction-induced pancreatitis, and the changes of acute pancreatitis usually resolve with relief of the obstruction.

**Pancreatic Cancer Manifesting as Acute Pancreatitis**

Pancreatitis due to pancreatic tumors such as adenocarcinoma, lymphoma, and metastases has been reported but is rare. Although chronic pancreatitis can coexist with pancreatic cancer, clinically evident acute pancreatitis occurs in only 3.1%–13.8% of cases (79,80). Pancreatic cancer has been implicated in only 1%–2% of cases of acute pancreatitis. The majority of cases are accompanied by masses of the pancreatic head and are secondary to ductal obstruction (81). However, a “nonobstructive” theory also exists, which postulates that the tumor itself releases enzymes that activate pancreatic trypsinogen, resulting in acute pancreatitis. In such cases, the diagnosis is often delayed because the nature of the underlying mass does not raise suspicion. The average delay in diagnosis may be up to 18–24 months (81), due to the fact that the inflammatory changes caused by the acute pancreatitis dominate the clinical picture and mask the underlying tumor.

The mass may not be visualized radiologically at initial presentation, with acute pancreatitis dominated by significant inflammatory changes. Nonetheless, an abscess, a pseudocyst, and focal pancreatitis may all mimic a neoplasm and may be indistinguishable from one another on a single scan. At imaging, the findings that are useful in accurately identifying an underlying malignancy include significant dilatation of the pancreatic duct with acute pancreatitis, disproportion between the size of the pancreatic head and that of the pancreatic body (the head being substantially bulkier than the body and tail), peripancreatic and upper abdominal lymphadenopathy, distant metastases, and vascular encasement or invasion. Malignancy should also be sought in any adult with recurrent attacks of pancreatitis without a known underlying risk factor. At times, the lesion may be detected only on follow-up scans obtained after the inflammation has subsided (Fig 12). Percutaneous imaging-guided needle biopsy is required to confirm the diagnosis. The management of the mass is no different from that of pancreatic cancer, but the prognosis may be influenced by the delay in diagnosis.
Figure 12. Pancreatic adenocarcinoma in a 44-year-old man who presented with acute left flank pain and laboratory evidence of pancreatitis. (a, b) Unenhanced CT scans obtained at initial presentation as part of a low-dose CT study show peripancreatic inflammation localized around the pancreatic tail with thickened anterior pararenal fascia (arrow in b), findings that are consistent with acute pancreatitis. The body and proximal tail of the pancreas are bulkier than the rest of the tail (arrowhead in a), a finding that should have raised suspicion for an underlying mass. (c) Contrast-enhanced CT scan obtained 6 weeks later shows residual inflammation around the pancreatic tail. A focal ill-defined hypodensity at the junction of the body and tail of the pancreas (arrow) with a mildly dilated MPD distal to it, findings that are suggestive of an underlying neoplasm. Extensive laboratory work-up did not reveal an underlying cause for acute pancreatitis. MR imaging was performed to address the suspicion raised at contrast-enhanced CT. (d, e) Axial unenhanced (1030/80, 5-mm section thickness) (d) and contrast-enhanced (3.35/1.5, 3-mm section thickness) (e) T2-weighted MR images show a vague, heterogeneously hyperintense lesion at the junction of the body and tail of the pancreas (arrow) that remains unenhanced on the postcontrast image. Disproportionate dilatation of the MPD in the pancreatic tail with atrophy of the tail is also seen.
Duodenal Villous Adenoma with Pancreatitis

Masslike lesions of the ampulla of Vater are unusual, accounting for less than 10% of periampullary and pancreatic tumors. Villous adenoma, the most common of these lesions, is a rare, benign epithelial lesion representing 1% of all duodenal tumors (82). The average patient age at presentation is 56 years, with lesions manifesting in patients under 50 years of age usually being benign (83). Villous adenomas commonly occur around the ampulla of Vater, although they may be located anywhere within the duodenum. Common clinical manifestations include jaundice secondary to biliary obstruction (70% of cases), abdominal pain, bleeding resulting in anemia, malena, and, rarely, pancreatitis (84). Only a very few cases of recurrent acute pancreatitis secondary to a benign neoplasm of the ampulla of Vater have been reported. Both microscopic and macroscopic adenomas have been linked to recurrent pancreatitis.

Small tumors confined to the ampulla may not be detected with a barium swallow upper gastrointestinal study, CT, or even MR imaging. For larger tumors, a barium swallow upper gastrointestinal study may demonstrate a filling defect in the duodenum (Fig 13a), and the lesion can usually be localized as an intraluminal-mucosal disease process. The characteristic “soap bubble” appearance of these tumors at a barium swallow upper gastrointestinal study may be seen in only

Figure 13. Duodenal villous adenoma in a 45-year-old man who presented with acute abdominal pain. Clinical findings and laboratory test results were consistent with acute pancreatitis. (a) Conventional radiograph shows a subtle filling defect (arrowheads) projecting along the medial wall of the duodenum. (b, c) Contrast-enhanced CT scans show a polypoidal mass (arrowheads) along the medial wall of the second part of the duodenum obstructing the ampulla of Vater, with resultant acute pancreatitis and peripancreatic inflammatory changes (arrow in c). (d) Virtual endoscopic image of the duodenum shows the mass with nodular projections along the medial wall of the second part of the duodenum. Surgery was performed, and the results of histopathologic analysis confirmed duodenal villous adenoma with no foci of malignant change.
one-third of cases (85). Endoscopic US is very sensitive in detecting small tumors in the ampullary-periampullary region. CT with adequate distention of the duodenal lumen with negative oral contrast material such as air, water, or methylcellulose may adequately reveal a soft-tissue mass arising from the duodenal wall and extending into the duodenal lumen (Fig 13b, 13c). MR cholangiopancreatography may demonstrate a mass in the ampullary region with the characteristic “double duct sign” that is common to all periampullary lesions, a finding that is helpful in making the diagnosis.

ERCP with biopsy is required for differentiating adenoma from adenocarcinoma and other primary ampullary tumors, since further characterization is not usually feasible at cross-sectional imaging. In the absence of known predisposing factors for recurrent pancreatitis, an obstructive cause in the ampullary-periampullary region should be considered, especially in the setting of pancreatic ductal dilatation. Careful evaluation with MR cholangiopancreatography or dedicated CT with adequate duodenal distention and ERCP with biopsy should be performed. However, preoperative biopsies often have low sensitivity, and, given the fact that 35%–63% of lesions can harbor malignancy, multiple biopsies are required to definitively exclude underlying carcinoma (84). ERCP may reveal a normal-looking papilla in up to 37% of patients with a microscopic adenoma. Surgical removal with wide excision is usually required for these lesions. Only pathologic analysis of a surgical specimen can confidently help exclude an underlying carcinoma within a villous adenoma (Fig 13d).

Conclusions
A variety of uncommon types and causes of pancreatitis have characteristic imaging features. For some of these entities, imaging findings may serve as the only clue to diagnosis. Therefore, despite the rarity of these entities, familiarity with their clinical and radiologic manifestations helps the radiologist in formulating an accurate differential diagnosis and allows him or her to exclude ominous pancreatic malignancy, thereby facilitating appropriate management in a timely fashion.

References


In cases with no underlying metabolic or systemic predisposing factors for acute or recurrent pancreatitis, special attention to these ancillary findings and critical reevaluation of these uncommon but potential causes are required.

CT is the investigative modality of choice. Diffuse enlargement of the gland with loss of lobular architecture (“featureless” pancreas) and homogeneously iso- or hypoattenuating parenchyma with a nondilated or diffusely narrowed pancreatic duct are characteristic features.

Tropical pancreatitis is a variant of chronic pancreatitis that is characterized by a constellation of findings including young age at onset, association with malnutrition, regional predisposition in tropical countries, rapidly progressive course with severe pancreatitis, the presence of large intraductal calculi, and an increased risk of adenocarcinoma.

Visualization of the MPD (dorsal duct) coursing anterior to the CBD before draining into the duodenum is also a valuable sign that should raise suspicion for pancreas divisum, especially when the ventral duct is not well visualized.

At imaging, the findings that are useful in accurately identifying an underlying malignancy include significant dilatation of the pancreatic duct with acute pancreatitis, disproportion between the size of the pancreatic head and that of the pancreatic body (the head being substantially bulkier than the body and tail), peripancreatic and upper abdominal lymphadenopathy, distant metastases, and vascular encasement or invasion.