Lymphoplasmacytic Sclerosing Pancreatitis (Autoimmune Pancreatitis): Evaluation with Multidetector CT

Satomi Kawamoto, MD • Stanley S. Siegelman, MD • Ralph H. Hruban, MD • Elliot K. Fishman, MD

Lymphoplasmacytic sclerosing pancreatitis is a form of chronic pancreatitis characterized by a mixed inflammatory infiltrate that centers on the pancreatic ducts. It is a cause of benign pancreatic disease that can clinically mimic pancreatic cancer. Preoperative detection of lymphoplasmacytic sclerosing pancreatitis is important because patients usually respond to steroid therapy. Patients with lymphoplasmacytic sclerosing pancreatitis are often referred for computed tomography (CT) when they are suspected of having a pancreatic or biliary neoplasm; therefore, it is important to search for potential findings suggestive of lymphoplasmacytic sclerosing pancreatitis when typical findings of a pancreatic or biliary neoplasm are not found. Typical CT findings include diffuse or focal enlargement of the pancreas without dilatation of the main pancreatic duct. Focal enlargement is most commonly seen in the head of the pancreas, and the involved pancreas on contrast material–enhanced CT images may be isoattenuating relative to the rest of the pancreas, or hypoattenuating, especially during the early postcontrast phase. Thickening and contrast enhancement of the wall of the common bile duct and gallbladder may reflect inflammatory infiltrate and fibrosis associated with lymphoplasmacytic sclerosing pancreatitis. There are several features seen at CT that may help to differentiate lymphoplasmacytic sclerosing pancreatitis from pancreatic cancer, such as diffuse enlargement of the pancreas with minimal peripancreatic stranding in patients with obstructive jaundice, an absence of significant main pancreatic duct dilatation, and an absence of significant main pancreatic duct dilatation. When these findings are encountered, clinical, other imaging, and serologic data should be evaluated.

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Abbreviation: ERCP = endoscopic retrograde cholangiopancreatography

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1From the Russell H. Morgan Department of Radiology and Radiological Science (S.K., S.S.S., E.K.F.) and Department of Pathology, Sol Goldman Pancreatic Cancer Research Center (R.H.H.), Johns Hopkins Medical Institutions, JHOC 3235A, 601 N Caroline St, Baltimore, MD 21287. Recipient of a Certificate of Merit award for an education exhibit at the 2005 RSNA Annual Meeting. Received November 7, 2006; revision requested April 18, 2007, and received June 21; accepted June 28. All authors have no financial relationships to disclose. Address correspondence to S.K. (e-mail: skawamo1@jhmi.edu).

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Introduction

In recent years, the concept of autoimmune pancreatitis has been established to refer to a special type of chronic pancreatitis with unique clinical and histologic manifestations. Autoimmune pancreatitis can be defined as a chronic inflammatory process of the pancreas that is caused by an autoimmune mechanism (1,2). The morphologic hallmarks are periductal infiltration by lymphocytes and plasma cells and granulocytic epithelial lesions with consequent destruction of the duct epithelium and venulitis (3). Therefore, autoimmune pancreatitis has been morphologically described as lymphoplasmacytic sclerosing pancreatitis, non-alcoholic duct-destructive chronic pancreatitis (4,5), or chronic sclerosing pancreatitis (6). The terms autoimmune pancreatitis and sclerosing pancreatitis have been used interchangeably (7).

The gross morphologic alterations produced by lymphoplasmacytic sclerosing pancreatitis may simulate malignancy, with characteristics such as masslike enlargement of the pancreatic head and/or irregular narrowing of the pancreatic duct and stricture of the common bile duct. However, lymphoplasmacytic sclerosing pancreatitis responds to oral steroid therapy, with reversible improvement of pancreatic morphology and function (1,8–10). Thus, when findings typical of a pancreatic or biliary neoplasm are not seen on computed tomographic (CT) images, it is important to search for characteristics suggestive of lymphoplasmacytic sclerosing pancreatitis and allow accurate diagnosis and appropriate therapy to occur.

The purpose of this article is to discuss and illustrate the spectrum of appearances of lymphoplasmacytic sclerosing pancreatitis on CT images. Pathologic and clinical features of lymphoplasmacytic sclerosing pancreatitis, CT techniques, and findings with other imaging modalities are also presented. CT features that may be potentially useful in differentiating lymphoplasmacytic sclerosing pancreatitis from pancreatic cancer are discussed and illustrated.

Most of the patients that are discussed in this article had no serologic parameters measured, and diagnosis was based on pathologic analysis. Therefore, in this article, we use the term lymphoplasmacytic sclerosing pancreatitis.

Pathologic Features of Lymphoplasmacytic Sclerosing Pancreatitis

At gross examination, the involved pancreas is firm or hard and may be enlarged or “mass forming.” These features may lead to surgical resection because of the suspicion that the lesion is carcinomatous (7). The inflammatory process may involve either the entire pancreas, or it may be limited to only a portion of the pancreas. When the inflammatory process involves only one portion of the pancreas, that segment is most often the pancreatic head. In a minority of cases, however, the inflammatory process is concentrated in the body or in the tail of the pancreas (7). It is not known how frequently and to what extent the entire pancreas is affected in lymphoplasmacytic sclerosing pancreatitis (3).

Microscopic findings of autoimmune pancreatitis are consistent with lymphoplasmacytic sclerosing pancreatitis. In pathologic specimens, dense lymphoplasmacytic infiltrate centered around the medium- and large-sized interlobular pancreatic ducts is seen (11) (Fig 1). Although the inflammatory infiltrate consists mainly of lymphocytes and plasma cells, it also contains some macrophages and occasionally neutrophilic and eosinophilic granulocytes (11). Immunocytochemical typing of the lymphocytes reveals that most of them are CD8- and CD4-positive T lymphocytes, with B lymphocytes present to a lesser degree (3). The lumen of inflamed pancreatic ducts is encompassed by infiltrate and is narrowed by infolding of the epithelium (3). A distinctive venulitis is also seen.

When the pancreas is only slightly affected, the inflammation centers almost entirely on the ducts; in severely affected pancreata, the inflammatory process involves the acinar parenchyma in addition to the ducts and leads to diffuse sclerosis (3). The acinar cells are replaced by inflammatory
cells and fibrosis, and the lobular architecture of the pancreas is almost lost (3). The peripancreatic and peribiliary lymph nodes are enlarged and show follicular hyperplasia (3).

It is also well known that patients with lymphoplasmacytic sclerosing pancreatitis display distal common bile duct strictures and inflammation, features that overlap with those of primary sclerosing cholangitis. Abraham et al (12) reported that among 20 patients with lymphoplasmacytic sclerosing pancreatitis treated with pancreaticoduodenectomy, inflammatory infiltrates were seen in the extrapancreatic common bile duct in 60%, in the intrapancreatic common bile duct in 84.2% (Fig 2), and in the gallbladder in 60%. However, inflammatory and sclerosing changes of the intrahepatic bile duct, which are typical findings in primary sclerosing cholangitis, are uncommon in lymphoplasmacytic sclerosing pancreatitis (3). Moreover, unlike typical primary sclerosing cholangitis, biliary lesions in lymphoplasmacytic sclerosing pancreatitis usually improve with administration of steroids. These findings suggest that the mechanism of the development of biliary lesions in lymphoplasmacytic sclerosing pancreatitis may differ from that of typical primary sclerosing cholangitis (3,13).

**Clinical Presentation of Lymphoplasmacytic Sclerosing Pancreatitis**

Lymphoplasmacytic sclerosing pancreatitis is a rare disorder, and the exact prevalence is unknown. Ito et al (8) reported that among 161 patients with chronic pancreatitis evaluated with endoscopic retrograde pancreatography, three patients (1.86%) had lymphoplasmacytic sclerosing pancreatitis. Okazaki et al (13) reported that among 620 patients with chronic pancreatitis, 30 of them (5%) had lymphoplasmacytic sclerosing pancreatitis. In a recent Italian multicenter study on the epidemiology of chronic pancreatitis that involved 21 centers and enrolled 282 patients suffering from chronic pancreatitis over 2 years, autoimmunity was recognized as an associated factor in 23 patients (6%) (7). The reported mean age of 41 patients with lymphoplasmacytic sclerosing pancreatitis was 62.2 years (range, 32–76 years) (14). Another study reported that the mean age of 53 patients with lymphoplasmacytic sclerosing pancreatitis was 56 years (range, 14–77 years) (15).

Clinically, lymphoplasmacytic sclerosing pancreatitis commonly manifests as obstructive jaundice with no or only mild abdominal pain, weight loss, and recent-onset diabetes in elderly patients. Acute attacks seen in severe or acute pancreatitis do not usually occur (13). Obstructive jaundice is often caused by stenosis of the intrapancreatic common bile duct (7) and is seen in 63%–75% of patients with lymphoplasmacytic sclerosing pancreatitis (13–15). Diabetes mellitus is also often associated with lymphoplasmacytic sclerosing pancreatitis and has a reported frequency of less than 20%–68% (7,13). Because these signs overlap with those of pancreatic cancer, lymphoplasmacytic sclerosing pancreatitis has frequently been misdiagnosed as pancreatic cancer. Hardacre et al (16) reported that between pancreatic cancer and lymphoplasmacytic sclerosing pancreatitis, there were no statistically significant differences in the rates of abdominal pain, weight loss, jaundice, preoperative carcinoembryonic antigen, or CA19-9 levels. Analysis of a large series involving more than 1200 patients who underwent pancreaticoduodenectomy with a presumed preoperative diagnosis of pancreatic cancer, periampullary neoplasm, or cholangiocarcinoma revealed that 2.2%–2.4% of patients had pathologic features consistent with lymphoplasmacytic sclerosing pancreatitis (16,17).

Elevation of serum gamma globulin or immunoglobulin G (IgG) concentration and the presence of some autoantibodies are often observed. Serum IgG4 concentrations were reported to be significantly and specifically high in patients with lymphoplasmacytic sclerosing pancreatitis and are closely associated with disease activity (18). Hamano et al (18) reported that the accuracy, sensitivity, and specificity of elevated serum concentrations of IgG4 in the diagnosis of this disease, with a cutoff value of 135 mg/dL, were 97%, 95%,
and 97% respectively. However, other groups reported lower sensitivity (19–21). Ghazale et al (21) reported that serum concentrations of IgG4 were elevated in 10% of pancreatic cancer patients (13 of 135), but that only 1% had serum IgG4 levels of >280 mg/dL, compared with 53% of patients with lymphoplasmacytic sclerosing pancreatitis.

Occasionally, patients with lymphoplasmacytic sclerosing pancreatitis have extrapancreatic lesions that are seen in other autoimmune diseases, including Sjögren syndrome, sclerosing cholangitis, primary biliary cirrhosis, interstitial nephritis, sialoadenitis, enlarged mediastinal or cervical lymph nodes, ulcerative colitis, and retroperitoneal fibrosis (22). The incidence of associated extrapancreatic autoimmune lesions is reported to range from 19% to more than 50% (7,15,22). These extrapancreatic autoimmune diseases may be recognized at the time of diagnosis or they may develop later (7). The presence of an associated autoimmune disease may help in the diagnosis of lymphoplasmacytic sclerosing pancreatitis (7).

Lymphoplasmacytic sclerosing pancreatitis is difficult to diagnose. In 2002, the Japan Pancreas Society proposed the following diagnostic criteria for autoimmune pancreatitis: (a) pancreatic imaging studies show diffuse enlargement of the pancreas and diffuse narrowing of the main pancreatic duct with an irregular wall (more than one-third the length of the entire pancreas), (b) laboratory data demonstrate abnormally elevated levels of serum gamma globulin and/or IgG or the presence of autoantibodies, and (c) fibrotic change with dense lymphoplasmacytic infiltration is noted in the pancreas at histopathologic examination. A diagnosis of lymphoplasmacytic sclerosing pancreatitis can be established if all of the criteria are present or if criterion a is present with either criterion b or criterion c (23). Recently, these criteria have been modified by the Japan Pancreas Society to allow diagnosis of autoimmune pancreatitis to include focal pancreatic mass and focal pancreatic duct stricture (24). Recently, other groups have also proposed diagnostic criteria for autoimmune pancreatitis (19,25).

**CT Technique**

CT examinations were performed with multidetector CT scanners, including a Siemens Volume Zoom scanner (4 × 1 mm collimation), a Siemens Sensation 16 scanner (16 × 0.75 mm collimation), and a Siemens Sensation 64 scanner (64 × 0.6 mm collimation) (Siemens Medical...
The data were reconstructed to obtain 1.25-mm section thickness at 1-mm intervals (0.25-mm overlap) with the Siemens Volume Zoom Scanner and 0.75-mm section thickness at 0.5-mm intervals (0.25-mm overlap) with the Siemens Sensation 16 and 64 scanners. After fasting for at least 2–3 hours, each patient ingested 750–1000 mL of water over a 15–20 minute period before scanning was begun. Arterial and venous phase images were acquired at 25 seconds and 50–60 seconds from the start of intravenous administration of contrast material. We injected 120 mL of iohexol (Omnipaque 350; Amersham Health, Princeton, NJ) through the peripheral venous line at a rate of 3 mL/sec. Other scanning parameters included 120 kV and 150–200 mAs.

All image data were reconstructed with the body soft tissue algorithm and sent to the workstation (Leonardo, Siemens Medical Solutions). InSpace software (Siemens Medical Solutions) was used for data analysis, which was the volume imaging application for interactive viewing of volume data available on the Leonardo workstation.

CT Findings of Lympho-plasmacytic Sclerosing Pancreatitis

Enlargement of the Pancreas

Typically, CT images show diffuse enlargement of the pancreas (Fig 3). Focal enlargement may also be seen, particularly in the pancreatic head (Fig 4), but also in the body or tail (5) (Fig 5). The presence

Figure 4. (a, b) Venous phase axial images show mild enlargement of the pancreatic head, with a stent present in the common bile duct. Nondilated main pancreatic duct is seen in the body and tail (arrowheads in a). Focal hypodense lesions are seen in both kidneys (arrows in b). Although biopsy of the lesions was not performed, follow-up CT showed that these lesions became less obvious. (c, d) Contrast-enhanced chest CT scans (d, lung window setting) show a left hilar mass with bulky mediastinal adenopathy. Results of biopsy performed after mediastinoscopy and limited anterior thoracotomy revealed anthracotic lymph nodes with fibrosis in the mediastinal lymph nodes, fibrous tissue of chronic inflammation in the mediastinal soft tissue, and mild chronic inflammation and pleural fibrosis in the left upper lobe.
of multiple masses has also been reported (26). In prior studies with CT, diffuse enlargement of the pancreas was more commonly encountered than was focal enlargement, and the incidence of diffuse enlargement ranged from 56% to 100% (9,27–31). Sahani et al (27) reported that among 25 patients with lymphoplasmacytic sclerosing pancreatitis who underwent helical CT examina-

Figure 5. Venous phase oblique axial (a) and coronal (b) reformatted CT images show focal enlargement of the pancreatic tail with a distinct area of decreased attenuation (arrows). Minimal stranding is seen around the enlarged pancreatic tail.

Figure 6. Venous phase axial (a) and coronal (b) reformatted CT images show a diffusely hypoattenuating pancreas that appears to be normal in size. The pancreas appears featureless, and the normal lobular appearance is effaced. A stent is seen in the common bile duct.
tion, 14 showed a diffusely enlarged pancreas, seven had smooth focal enlargement or a masslike appearance in the pancreatic head and/or the uncinate process, and four had a pancreas that appeared to be normal in size (Fig 6). Kamisawa et al (28) reported that among 17 patients with lymphoplasmacytic sclerosing pancreatitis, 10 had diffuse enlargement and seven had segmental enlargement of the pancreatic head. However, another study reported that diffuse enlargement was seen in only six of 22 patients (27%) with lymphoplasmacytic sclerosing pancreatitis (25). Atrophy of the pancreas is not usually seen (28), although the normal lobular appearance of the pancreas may be effaced and the gland may appear featureless in the involved region (32) (Figs 6, 7).

**Figure 7.** Venous phase axial (a) and coronal (b, c) reformatted CT images show diffuse enlargement of the entire pancreas. The pancreas appears featureless, and the normal lobular appearance is effaced. There is minimal peripancreatic stranding, and a stent is seen in the common bile duct.

Contrast Enhancement Pattern of the Pancreatic Parenchyma

The reported contrast enhancement pattern of the involved pancreas seen in lymphoplasmacytic sclerosing pancreatitis is variable. Previous studies reported that on arterial phase or early post-contrast phase images, the involved portion of the pancreas appears hypoattenuating compared with unaffected pancreatic parenchyma and occasionally has a distinct margin (9,33) (Fig 5). On venous phase images, the involved portion of the pancreas may remain hypoattenuating (33) or may become nearly isoattenuating compared with
unaffected pancreatic parenchyma (Figs 8, 9). However, a discrete area of differential contrast enhancement may not be observed on arterial or venous phase images in the areas of focal enlargement, and the pancreas may show homogeneous contrast enhancement (27,30,32,34). In previous CT studies, the area of focal enlargement may appear homogeneously isoattenuating relative to the pancreas in 25%–71% of patients, or it may appear hypoattenuating in 29%–75% of patients (27,34). When delayed phase images are obtained at several minutes after intravenous administration of contrast material, the involved portion is reported to become homogeneously isoattenuating or hyperattenuating compared with the surrounding pancreatic parenchyma (31,34,35). However, another study reported that the involved lesion remains hypoattenuating compared with unaffected pancreatic parenchyma on delayed phase images (33). These differences may be attributed to the different CT techniques and the degree of inflammatory infiltrate and fibrosis of the involved pancreas.

**Figure 8.** (a, b) Arterial phase axial (a) and coronal (b) reformatted CT images show enlargement of the pancreatic head with a distinct area of decreased attenuation within the enlarged pancreatic head (arrowheads). The body and tail of the pancreas are relatively atrophic. (c) Venous phase axial image shows that the hypoattenuating area seen in the pancreatic head on arterial phase images becomes nearly isoattenuating relative to adjacent pancreatic parenchyma. A stent (arrow in a and c) is seen in the common bile duct.
Narrowing of the Pancreatic Duct
Thin-section CT with multiplanar reformation helps to delineate the main pancreatic duct. In patients with lymphoplasmacytic sclerosing pancreatitis, the main pancreatic duct is diffusely or segmentally narrowed. On CT images, the main pancreatic duct may be seen as a small nondilated duct, or it may appear attenuated, particularly in the area of the pancreatic enlargement. Mild dilation of the main pancreatic duct may also be seen proximal to the enlarged portion of the pancreas (Fig 9). However, significant dilatation of the main pancreatic duct, a characteristic of pancreatic ductal adenocarcinoma, is not found in lymphoplasmacytic sclerosing pancreatitis.

Other Pancreatic/Peripancreatic Findings
Minimal peripancreatic stranding, which may simulate mild edematous acute pancreatitis, is often seen at CT (27,33) (Figs 3, 7). A capsule-like low-attenuation rim has also been described in 12%–80% of reported cases of lymphoplasmacytic sclerosing pancreatitis (Fig 10) (19,27,28,30–32). On delayed phase images, a capsule-like low-attenuation rim may show subtle delayed enhancement (31,33).
In contrast to other forms of chronic pancreatitis, lymphoplasmacytic sclerosing pancreatitis does not commonly manifest with parenchymal calcifications and pseudocysts (3,28,36,37). However, intraductal calcifications may occur late in the course of the disease (9,31,33,38), and formation of pseudocysts associated with lymphoplasmacytic sclerosing pancreatitis has also been reported (39).

Major pancreatic vascular involvement is uncommon in lymphoplasmacytic sclerosing pancreatitis compared with pancreatic adenocarcinoma (27), although cases with venous occlusion or narrowing, particularly the splenic vein, have been reported (9,30,32) (Fig 10). Major arterial involvement—such as narrowing of the celiac artery and the superior mesenteric artery—has not been reported. At conventional angiography, however, irregular narrowing and encasement of the small peripancreatic arteries were reported in up to 57% of cases (28,35). Poor opacification of portal or splenic veins due to stenosis or obstruction with collateral veins was also reported in 23% of cases at conventional angiography (28).

Enlarged peripancreatic lymph nodes may also be observed on CT images. Sahani et al (27) reported that nine of 25 patients with lymphoplasmacytic sclerosing pancreatitis had enlarged peripancreatic nodes that measured more than 1 cm in diameter on the short axis on CT scans.

**Biliary Tract Findings**

Stricture of the common bile duct is often seen in patients with lymphoplasmacytic sclerosing pancreatitis, particularly in patients whose pancreatic head is affected, with a reported frequency of 33%–90% (5,27,28,30,31,40). At endoscopic...
retrograde cholangiopancreatography (ERCP), smooth stenosis of the distal common bile duct localized in the pancreatic head, with dilatation of the more proximal common bile duct, is the most common finding in patients with lymphoplasmocytic sclerosing pancreatitis (27,41) (Figs. 3, 11). Narrowing of the intrapancreatic common bile duct is thought to be induced mainly by compression of the swollen pancreas (7). Coronal three-dimensional or reformatted CT images help to delineate smooth, beaklike stenosis of the common bile duct in the intrapancreatic portion and dilatation of the proximal common bile duct (Figs. 3, 11). Stenosis or strictures of the proximal and middle extrahepatic bile duct and the intrahepatic bile duct—findings that resemble those seen in primary sclerosing cholangitis—have also been observed at ERCP and magnetic resonance (MR) cholangiopancreatography (7,27,28).

At CT, thickening and enhancement of the gallbladder wall and/or the common bile duct wall have been described, and these findings correspond to the inflammatory infiltrate and fibrosis observed microscopically (Figs. 3, 11) (30,32,42). However, when a biliary stent is present at the time of CT scanning, these biliary tract findings may be obscured due to pneumobilia or changes related to stent placement.

Extrapancreatic Findings
Involvement with multiple organ systems has been reported, and these findings may be seen on CT images. Such changes include, but are not limited to, retroperitoneal fibrosis, renal involvement, lung disease, and mediastinal adenopathy (27,43).

Findings at Other Imaging Modalities
Patients with lymphoplasmocytic sclerosing pancreatitis typically show diffuse or segmental narrowing of the main pancreatic duct at ERCP. The secondary branches are usually not visualized (7–9).

MR imaging may reveal diffuse pancreatic enlargement with hypointensity on T1-weighted images. The low-attenuation capsuleslike rim described at CT is hypointense on T2-weighted images and shows delayed contrast enhancement, which suggests fibrous tissue rather than a fluid
collection or phlegmon (31). MR cholangiopancreatography may show stenosis of the bile ducts mainly in the intrapancreatic area, which results in dilatation of the proximal biliary tract (7). Sclerosing changes of the intrahepatic bile ducts or common bile duct that are similar to primary sclerosing cholangitis are sometimes observed (7,44). Although stenosis of the main pancreatic duct may be observed, determining whether it is due to lymphoplasmacytic sclerosing pancreatitis or to pancreatic carcinoma may be difficult with MR cholangiopancreatography (40).

At ultrasonographic (US) examination, the pancreas appears hypoechoic and diffusely enlarged with a so-called sausagelike appearance (8,9,28). It may also appear as a hypoechoic mass in the affected site (5,28). Endoscopic US may show diffuse hypoechoic pancreatic enlargement or a focal, irregular hypoechoic mass (20). Structure of the common bile duct in the pancreatic head, widespread bile duct wall thickening, and diffuse strong enhancement of the bile duct system, including the gallbladder, proximal bile duct, and distal bile duct, have also been seen with endoscopic US when contrast material is used (41). Farrell et al (20) reported that findings from endoscopic US-guided fine-needle aspiration may support the diagnosis of lymphoplasmacytic sclerosing pancreatitis in combination with endoscopic US findings and clinical data.

Gallium scintigraphy (45,46) and fluorine 18 fluorodeoxyglucose positron emission tomography (47) have been reported to show increased uptake in the affected site of the pancreas during the active stage of the disease, and such findings may be confused with the increased uptake seen in pancreatic cancer or lymphoma.

CT Features to Help Differentiate Pancreatic Cancer

Lymphoplasmacytic sclerosing pancreatitis has several typical characteristics on CT scans that may be useful in differentiating the condition from pancreatic cancer. When diffuse enlargement of the pancreas with mild peripancreatic stranding is seen on CT images of patients with obstructive jaundice without clinical features of acute pancreatitis, lymphoplasmacytic sclerosing pancreatitis should be considered as a potential diagnosis. Clinical, serologic, and other imaging studies should be carefully evaluated.

In patients with focal enlargement of the pancreas, diagnosing lymphoplasmacytic sclerosing pancreatitis is more challenging. When a patient with obstructive jaundice presents with a “mass” in the pancreatic head, differentiating between lymphoplasmacytic sclerosing pancreatitis and pancreatic cancer can be extremely difficult (48). The mass may also be seen in the body or tail of the pancreas, and it may simulate pancreatic cancer. CT findings more typically seen in pancreatic cancer than in lymphoplasmacytic sclerosing pancreatitis include (a) significant dilatation of the main pancreatic duct proximal to the narrowed segment, (b) atrophy of the pancreatic parenchyma proximal to the mass or focal enlargement, and (c) involvement of the major peripancreatic vessels. In patients with segmental narrowing of the main pancreatic duct due to lymphoplasmacytic sclerosing pancreatitis, the main pancreatic duct proximal to the segmental narrowing typically shows minimal or no dilatation (19,28). Previous studies that compared ERCP images of focal lymphoplasmacytic sclerosing pancreatitis and those of pancreatic cancer showed that the caliber of the main pancreatic duct proximal to the stricture is smaller in patients with lymphoplasmacytic sclerosing pancreatitis (<4 mm in 67% of patients and 4–6 mm in 33%) than in those with pancreatic ductal adenocarcinoma (<4 mm in 4%, 4–6 mm in 22%, and >6 mm in 74%) (30,35,49). Major peripancreatic vascular involvement, particularly arterial involvement, is an uncommon CT finding in lymphoplasmacytic sclerosing pancreatitis. Atrophy of the pancreas proximal to the mass is often seen in patients with pancreatic cancer, but it is usually not found in lymphoplasmacytic sclerosing pancreatitis (28). Although the acinar parenchyma becomes atrophic in lymphoplasmacytic sclerosing pancreatitis, it is replaced with fibrous tissue (36), which probably explains why the overall size of the pancreas does not usually change. Fine-needle aspiration biopsy may remain a necessary step to confirm the diagnosis (7).

Smooth narrowing of the distal common bile duct can be seen in patients with lymphoplasmacytic sclerosing pancreatitis; however, pancreatic cancer may display similar findings. Wakabayashi et al (35) reported that there were no significant differences among patients with focal lymphoplasmacytic sclerosing pancreatitis and those with pancreatic cancer in terms of the frequency of common bile duct stenosis and its character and length as seen with ERCP. Procacci et al (33) reported that CT findings can be used to correctly diagnose lymphoplasmacytic sclerosing pancreatitis with an accuracy of 92.5%. They evaluated seven patients with lymphoplasmacytic sclerosing pancreatitis and 20 patients with other pancreatic diseases (acute or chronic pancreatitis and adenocarcinoma of the pancreas). The criteria for a diagnosis of lymphoplasmacytic sclerosing pancreatitis included
focal or diffuse enlargement of the pancreas, possible capsule-like rim, possible stenosis or complete obstruction of the biliary duct, and possible stenosis (either focal or diffuse) of the main pancreatic duct. The affected pancreatic parenchyma was isodense to relative to the spleen and adjacent unaffected parenchyma on unenhanced CT images and hypoattenuating on arterial phase, venous phase, and delayed phase images (33). Procacci and colleagues reported one false-negative diagnosis of lymphoplasmacytic sclerosing pancreatitis in a case of chronic pancreatitis superimposed with lymphoplasmacytic sclerosing pancreatitis that showed diffuse calcifications in the pancreas. They also recorded one false-positive diagnosis for lymphoplasmacytic sclerosing pancreatitis in a case of mild edematous acute pancreatitis with imaging findings similar to those of lymphoplasmacytic sclerosing pancreatitis.

**Conclusions**

In conclusion, lymphoplasmacytic sclerosing pancreatitis is a unique clinical entity that is becoming more frequently recognized in clinical practice. Because patients with lymphoplasmacytic sclerosing pancreatitis often present with obstructive jaundice, their disease has been frequently misdiagnosed as pancreatic cancer. However, because lymphoplasmacytic sclerosing pancreatitis responds to steroid therapy, it is important to recognize this entity in order to avoid surgery. Characteristics seen on CT images that suggest lymphoplasmacytic sclerosing pancreatitis include diffuse or focal enlargement of the pancreas, absence of significant pancreatic atrophy, absence of substantial main pancreatic duct dilatation, and in some cases a rim of low attenuation surrounding the pancreas. However, diagnosis of this disease can be difficult, especially with patients who present with a focal mass or a masslike enlargement of the pancreas. When these CT findings are encountered, lymphoplasmacytic sclerosing pancreatitis should be considered as a potential diagnosis, and clinical, other imaging, and serologic data should be carefully evaluated. Fine-needle aspiration biopsy may remain a necessary step for excluding malignancy and for final definitive diagnosis.

**References**


Lymphoplasmacytic Sclerosing Pancreatitis (Autoimmune Pancreatitis): Evaluation with Multidetector CT

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