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Pancreatic Pseudotumors Associated with Multifocal Idiopathic Fibrosclerosis

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Abstract. Two patients with multifocal idiopathic fibrosclerosis and sclerosing cholangitis developed biliary obstruction due to a fibrotic pancreatic pseudotumor. The masslike fibrosis mimicked pancreatic carcinoma on sonography and cholangiopancreatography. In one patient sonography was successfully used to assess the response of the pseudotumor to corticosteroid therapy.

Key words: Pseudotumors, pancreatic – Multifocal idiopathic fibrosclerosis.

Multifocal idiopathic fibrosclerosis (MIF) is an uncommon fibroproliferative disorder with multiple manifestations including retroperitoneal fibrosis, sclerosing cholangitis, Riedel's thyroiditis, fibrotic pseudotumor of the orbit, Dupuytren's contractures, and fibrosis of the salivary glands (sicca syndrome) [1–3]. Sclerosing cholangitis is a well-known cause of obstructive jaundice in patients with MIF, but pancreatic fibrosis is also becoming increasingly recognized [2–5]. We present two patients with MIF, progressive jaundice, and masslike pancreatic fibrosis that mimicked pancreatic carcinoma.

Case Reports

Case 1

A 62-year-old man, was admitted to the hospital with increasing jaundice. His past medical history included a subtotal thyroidectomy 25 years earlier. The thyroid was found to be enlarged and infiltrated by benign fibrous tissue. The patient also had a long history of unilateral proptosis. A computed tomographic (CT) scan 2 years prior to admission revealed a retroorbital soft tissue mass, but a biopsy was never performed. Six months prior to admission the patient experienced submandibular pain and swelling. The right submandibular gland was excised. The specimen histologically showed only fibrosis and chronic sialadenitis.

During the present admission the serum bilirubin level was 27.3 mg/dl. An ultrasound scan (Fig. 1) showed a large sonolucent mass in the head of the pancreas. The extrahepatic and intrahepatic ductal systems were not grossly dilated. The liver demonstrated slightly increased echogenicity. Because of the possibility of pancreatic carcinoma, the patient underwent exploratory laparotomy. A 6-cm indurated mass encasing the common bile duct was found in the head of the pancreas. Multiple frozen and permanent biopsy specimens demonstrated extensive fibrosis and an occlusive plasma cell phlebitis. Elements of both acute and chronic inflammation were present with parenchymal fibrosis and atrophy. A liver biopsy specimen revealed cholestasis and changes suggestive of sclerosing cholangitis. Because of all the fibrosis it was impossible to perform an adequate operative cholangiogram. Following surgery a percutaneous transhepatic cholangiogram was performed (Fig. 1) to better assess the intrahepatic bile ducts. It showed an irregular stricture of the distal common bile duct and multiple stenoses of the intrahepatic ducts. The latter were suggestive of sclerosing cholangitis. The patient was placed on low-dose steroids, which resulted in return of the serum bilirubin level to normal. A repeat sonogram demonstrated complete resolution of the pancreatic mass, 12 weeks (Fig. 1), and 12 months after initiation of treatment.

Case 2

A 63-year-old man was admitted to the hospital with jaundice of 3 weeks' duration. His medical history was unremarkable except for mild ethanol abuse. There were no documented prior bouts of pancreatitis. Physical examination revealed Dupuytren's contractures and moderate hepatomegaly. Serum bilirubin (total) level was 24 mg/dl. Serum amylase level was normal. Sonography demonstrated gallstones, a small amount of ascites, and no biliary dilatation. The pancreas was poorly seen. Endoscopic retrograde cholangiopancreatography (ERCP) failed to opacify the biliary tree. The pancreas was enlarged and the pancreatic duct was diffusely encased (Fig. 2) with attenuation and splaying of secondary branches. There was no evidence to suggest changes of chronic pancreatitis. Exploratory laparot-

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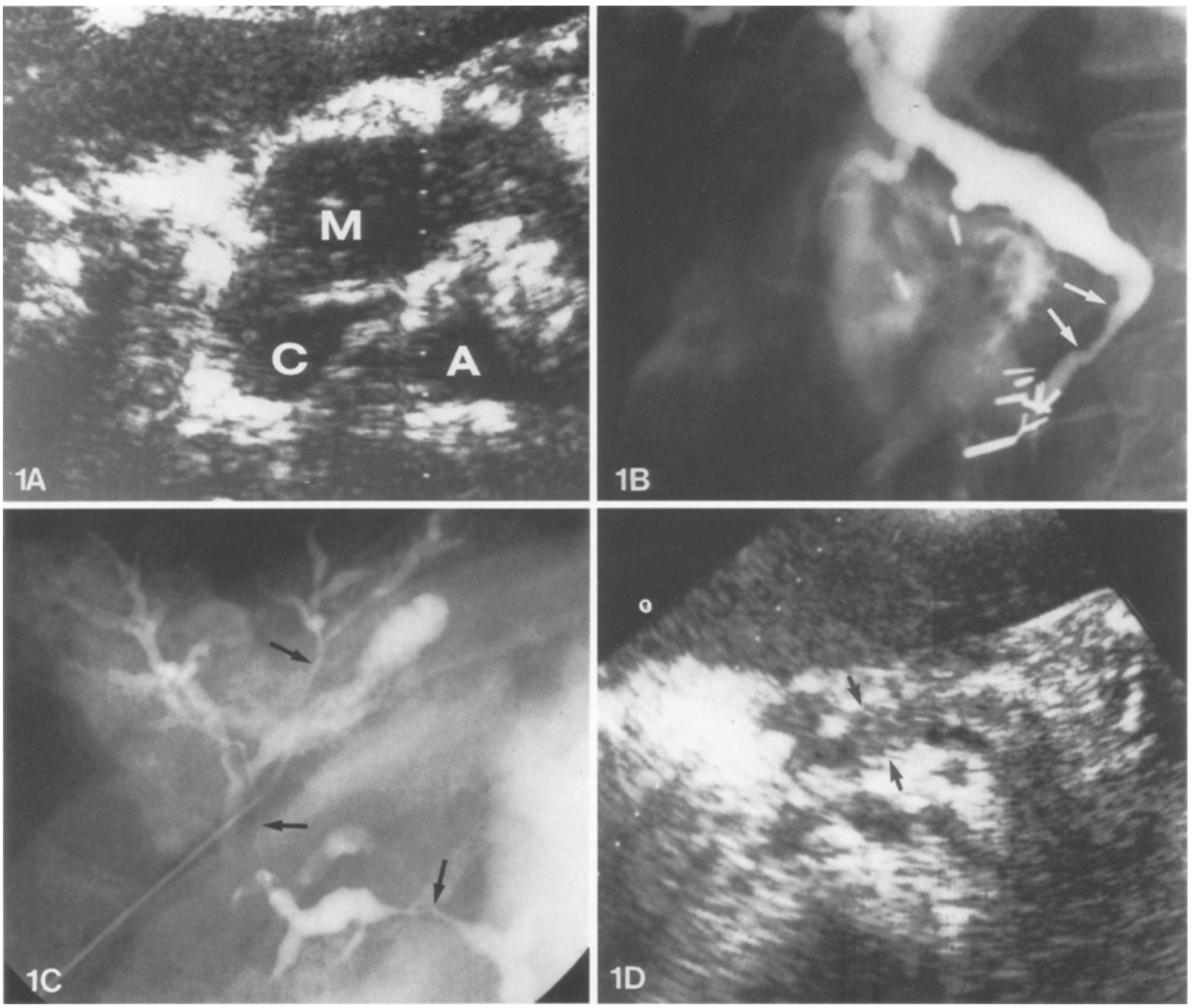

Fig. 1. *Case 1.* Sonographic demonstration of masslike pancreatic fibrosis. **A** Transverse sonogram illustrates a large sonolucent pancreatic mass (*M*) anterior to the inferior vena cava (*C*). *A*, aorta. **B** Percutaneous transhepatic cholangiogram shows a long irregular stricture of the distal common bile duct (*arrows*). There is also fine irregularity of the extrahepatic duct contour suggestive of sclerosing cholangitis. **C** Percutaneous transhepatic cholangiogram (oblique view) shows multiple encased intrahepatic ducts (*arrows*). The differential diagnosis includes sclerosing cholangitis, multicentric cholangiocarcinoma, and metastatic tumor. The intrahepatic ductal changes proved to be due to sclerosing cholangitis. The distal common bile duct abnormality was due to biopsy-proven pancreatic fibrosis. **D** Transverse sonogram following 12 weeks of corticosteroid therapy reveals complete resolution of the pancreatic mass. Note the normal pancreatic contours (*arrows*).

omy and open pancreatic biopsy revealed dense fibrosis diffusely replacing the pancreas and extending proximally along the common bile duct. A liver biopsy specimen showed cirrhosis, ductal proliferation, and periportal fibrosis associated with sclerosing cholangitis.

Discussion

Multifocal idiopathic fibrosclerosis is a fibroproliferative disorder known to involve multiple organ systems. When this disorder is associated with sclerosing cholangitis it is a frequent cause of jaundice and abnormal liver function tests. The two cases presented here illustrate that the extensive fibrosis produced in patients with this entity may not only result in sclerosing cholangitis but also may infiltrate the pancreas and extrinsically encase the bile duct as it courses to the duodenum. Sclerosing cholangitis may display thickening of the duct wall up to several millimeters [4] but in the absence of pancreatic involvement by fibrosclerosis is not usually associated with bulky, masslike fibrosis.

Pancreatic fibrosis due to MIF has been previously reported in 5 patients [2–5]. All 5 patients

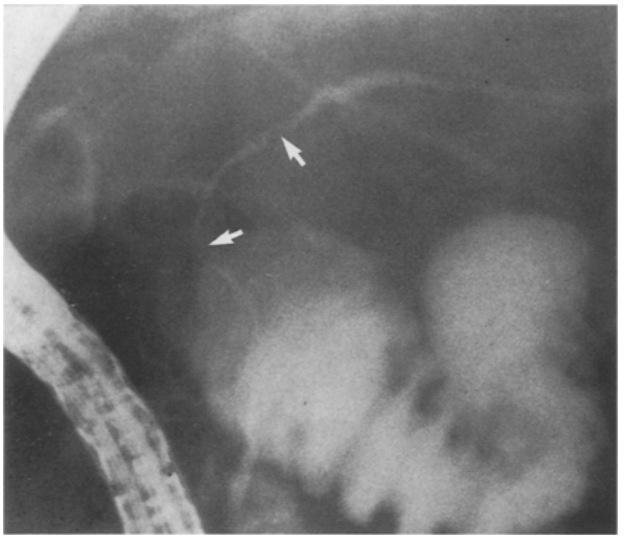

Fig. 2. *Case 2.* Abnormal pancreatogram in pancreatic fibrosis. ERCP (pancreatic duct injection) shows attenuation of the main pancreatic duct (*arrows*). Displacement and pruning of secondary branches were also identified. The pancreas was shown at open biopsy to be extensively infiltrated by fibrosis.

presented with hyperbilirubinemia and had operative or histologic evidence of associated sclerosing cholangitis. In 2 of these patients the cholangiographic picture failed to suggest sclerosing cholangitis and an atypical focal stricture that mimicked tumor encasement was present. In case 1 the intrahepatic cholangiographic findings were suggestive of sclerosing cholangitis, but the extrahepatic ductal changes were nonspecific and could have been due to a benign or malignant process. If the cholangiogram reveals morphologic changes suggestive of sclerosing cholangitis in the presence of a pancreatic mass, the radiologist may suggest the diagnosis of a fibrotic pancreatic pseudotumor, but biopsy remains mandatory to exclude carcinoma.

Sonography was performed in 2 of the 5 previously reported patients with MIF and pancreatic fibrosis. Unfortunately the appearance of the pseudotumor was not described in either patient. In our case 1, the mass appeared sonolucent and was indistinguishable from pancreatic carcinoma or focal pancreatitis. Following treatment with steroids, the pancreatic fibrosis resolved leaving the pancreas of normal echo texture. To the best of our knowledge this has not been previously reported in the sonographic literature. It is well known that steroids may be of value in the treatment of some patients with retroperitoneal fibrosis [7] and it is therefore not surprising that pancreatic fibrosis may respond similarly.

Only one of the previously reported patients with pancreatic fibrosis underwent pancreatography. In that patient amputation of the pancreatic duct was present mimicking carcinoma [3]. The patient in case 2 has splaying and attenuation of the pancreatic primary and secondary ducts rather than amputation of the main duct. This appearance, although atypical for carcinoma is nonspecific. The pancreatogram, therefore, may not be helpful in differentiating carcinoma from pancreatic fibrosis. The ability in this setting to opacify the pancreatic duct should not be used by nonradiologists as justification for ERCP over antegrade percutaneous cholangiography.

Neither noninvasive nor invasive imaging has proven very useful in providing a specific diagnosis in patients with MIF and pancreatic fibrosis. Cholangiographic changes characteristic of sclerosing cholangitis in the presence of a pancreatic mass allow the radiologist to suggest the diagnosis of a fibrotic pancreatic pseudotumor. At the present time, however, biopsy remains essential for establishing the diagnosis. Although not performed in our patients, percutaneous biopsy and close noninvasive imaging follow-up during steroid therapy might provide an alternative to open biopsy.

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