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RAPID COMMUNICATION

Involvement of pancreatic and bile ducts in autoimmune pancreatitis

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Abstract

AIM: To examine the involvement of the pancreatic and bile ducts in patients with autoimmune pancreatitis.

METHODS: Clinical and cholangiopancreatographic findings of 28 patients with autoimmune pancreatitis were evaluated. For the purposes of this study, the pancreatic duct system was divided into three portions: the ventral pancreatic duct; the head portion of the dorsal pancreatic duct; and the body and tail of the dorsal pancreatic duct.

RESULTS: Both the ventral and dorsal pancreatic ducts were involved in 24 patients, while in 4 patients only the dorsal pancreatic duct was involved. Marked stricture of the bile duct was detected in 20 patients and their initial symptom was obstructive jaundice. Six patients showed moderate stenosis to 30%-40% of the normal diameter, and the other two patients showed no stenosis of the bile duct. Although marked stricture of the bile duct was detected in 83% (20/24) of patients who showed narrowing of both the ventral and dorsal pancreatic ducts, it was not observed in the 4 patients who showed involvement of the dorsal pancreatic duct alone ($P = 0.0034$).

CONCLUSION: Both the ventral and dorsal pancreatic and bile ducts are involved in patients with autoimmune pancreatitis.

Key words: Autoimmune pancreatitis; Dorsal pancreatic duct; Ventral pancreatic duct

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INTRODUCTION

Autoimmune pancreatitis is a recently described clinical entity in which autoimmune mechanisms are involved in the pathogenesis[1-3]. It is characterized by the following clinical features: elderly male preponderance, the frequent initial symptom of jaundice, laboratory findings of serum elevation of gammaglobulin or IgG, especially IgG4, presence of autoantibodies, and a favorable response to steroid therapy. Histological findings are dense lymphoplasmacytic infiltration and fibrosis of the pancreas.

Frequently observed radiological imaging features are the enlargement of the pancreas and irregular narrowing of the main pancreatic duct. Most of the cases with autoimmune pancreatitis reveal a diffusely enlarged pancreas with a sausage-like appearance. Typical cases also show diffuse irregular narrowing of the entire main pancreatic duct. A focal type of autoimmune pancreatitis, showing focal swelling with a localized narrowing of the main pancreatic duct and upstream dilatation, is regarded as a variant of autoimmune pancreatitis[4].

Stenosis of the bile duct frequently occurs with autoimmune pancreatitis. The stenotic portion of the bile duct is usually the lower bile duct; however, the middle and upper extrahepatic bile duct and intrahepatic bile duct are sometimes involved. There are few precise reports of cholangiopancreatographic findings in autoimmune pancreatitis[5-7]. In the present study, we have aimed to examine the involvement of the pancreatic and bile ducts in patients with autoimmune pancreatitis.

MATERIALS AND METHODS

Patients

Between 1975 and 2003, a total of 28 patients (22 men and 6 women, average age 68.4 years) were diagnosed with autoimmune pancreatitis by the following

clinicopathological criteria: irregular narrowing of the main pancreatic duct on endoscopic retrograde pancreatography ($n = 28$); pancreatic enlargement on ultrasound or computed tomography ($n = 27$); hypergammaglobulinemia in excess of 2.0 g/dL ($n = 15$); elevated serum IgG4 in excess of 135 mg/dL ($n = 15$); presence of autoantibodies ($n = 15$); characteristic histological findings in the pancreas ($n = 11$); and responsiveness to steroid therapy ($n = 15$).

Methods

All patients underwent endoscopic retrograde cholangiopancreatography, and 10 patients also underwent magnetic resonance cholangiopancreatography. In all the patients treated with steroids, cholangiopancreatography was performed before and after the treatment. For the purposes of this study, we divided the pancreatic duct system into three portions: the ventral pancreatic duct; the head portion of the dorsal pancreatic duct including the accessory pancreatic duct; and the body and tail of the dorsal pancreatic duct. Clinical and cholangiopancreatographic findings were evaluated.

RESULTS

Both the ventral and dorsal pancreatic ducts were involved in 24 patients, 6 of whom showed segmental narrowing of the head portion of the dorsal pancreatic duct. In four patients, only the dorsal pancreatic duct was involved, two of whom showed segmental narrowing of the body and tail of the dorsal pancreatic duct.

Stenosis of the bile duct was detected in 26 patients, including 23 patients in the lower portion of the duct, 1 patient in the middle portion, 1 patient in the lower and intrahepatic portions, and 1 patient in the middle and intrahepatic portions. A marked stricture of the bile duct was detected in 20 patients and their initial symptom was obstructive jaundice. The other 6 patients showed moderate stenosis to 30%-40% of the normal diameter.

Although marked stricture of the bile duct was detected in 83% (20/24) patients who showed narrowing of both the ventral and dorsal pancreatic ducts, it was not observed in the four patients who showed only the involvement of the dorsal pancreatic duct ($P = 0.0034$).

DISCUSSION

The typical pancreatographic appearance of autoimmune pancreatitis is diffuse irregular narrowing of the main pancreatic duct involving both the ventral and dorsal pancreatic ducts[1-3]. Some cases of autoimmune pancreatitis show segmental irregular narrowing of the main pancreatic duct[4-7]. The segmental and diffuse types of autoimmune pancreatitis appear to represent a spectrum of the same disease rather than separate entities, as some studies[5,8] have reported the progression of segmental narrowing to diffuse narrowing on serial endoscopic retrograde pancreatography without steroid treatment. The pancreas develops by the fusion of the dorsal and ventral pancreatic primordial buds. The main pancreatic duct is formed by the fusion of the ventral and dorsal pancreatic ducts, while the accessory pancreatic duct is formed from the proximal portion of the dorsal pancreatic duct[9,10]. In the present study, we have divided the pancreatic duct system embryologically into three parts: the ventral pancreatic duct, the head portion of the dorsal pancreatic duct including the accessory pancreatic duct, and the body and tail of the dorsal pancreatic duct. Both the ventral and dorsal pancreatic ducts were involved in 24 patients, while 4 patients exhibited pathologic changes only in the dorsal pancreatic duct. Although the pathogenesis of autoimmune pancreatitis is unclear, autoimmune mechanisms might have occurred only in the dorsal pancreas in these 4 patients.

Stenosis of the bile duct frequently occurs with autoimmune pancreatitis. While the stenotic portion of the bile duct is usually the lower bile duct, the upper extrahepatic and intrahepatic bile ducts are sometimes involved. In the present study, a marked stricture of the bile duct was observed in 83% (20/24) patients with narrowing of both the ventral and dorsal pancreatic ducts, while 4 patients without the involvement of the ventral pancreatic duct showed no stenosis or moderate stenosis of the bile duct and no clinical manifestation of obstructive jaundice. Diffuse infiltrations by numerous lymphocytes and plasma cells with marked fibrosis in the pancreas and bile duct characterize the histological findings of autoimmune pancreatitis[7,11]. Periductal inflammation and marked fibrosis are the main causes of the bile duct stricture in autoimmune pancreatitis; however, fibrosis of the ventral pancreas around the lower bile duct may also be a contributing factor. In terms of the anatomical relationship between the dorsal pancreas and the common bile duct, obstructive jaundice rarely occurs without the involvement of the ventral pancreas.

In conclusion, both the ventral and dorsal pancreatic ducts are involved in a majority of the patients with autoimmune pancreatitis, while some patients show the involvement of only the dorsal pancreatic duct. Although obstructive jaundice due to stenosis of the bile duct commonly occurs with autoimmune pancreatitis, it rarely occurs in patients without the involvement of the ventral pancreas.

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