Autoimmune pancreatitis is a recently recognized clinical entity characterized by narrow strictured main pancreatic duct on ERCP, diffusely enlarged sausage-shaped pancreas on CT scan and MRI, seropositivity for antinuclear antibodies, hypergammaglobulinemia, and excellent response to steroids. We report a 25-year-old man and a 53-year-old man with this condition. [Indian J Gastroenterol 2004; 23: 181-183]

Key words: Autoimmune disorders

Since Sarles et al1 reported a particular type of pancreatitis with hypergammaglobulinemia, many investigators have suggested that an autoimmune mechanism may be involved in some patients with chronic pancreatitis. An association of pancreatitis with other autoimmune diseases like diabetes mellitus, sicca syndrome, primary sclerosing cholangitis, primary biliary cirrhosis, and systemic lupus erythematosus has been described.2 Recently, cases of autoimmune pancreatitis without systemic autoimmune syndrome have been reported, which has led to the concept of autoimmune-related pancreatitis or autoimmune pancreatitis (AIP).3

Case Reports

Case 1: A 25-year-old teetotaller presented with 4 months of recurrent abdominal pain suggestive of pancreatitis, and cholestatic symptoms for a week prior to presentation. He had no past or family history of pancreatitis. On examination he had icterus. Laboratory tests (Table) confirmed the presence of cholestasis. Tests for antinuclear antibodies (ANA) and double-stranded DNA (ds DNA) were positive. Ultrasonography showed diffusely enlarged pancreas. CT scan (Fig 1) showed an enlarged, hypodense, sausage-shaped pancreas with hypodense rim. ERCP showed a smooth stricture in the distal bile duct and a narrow, irregular main pancreatic duct with multiple strictures in the head region.

The patient was started on prednisolone 40 mg daily. After 6 weeks, when steroids were tapered to 20 mg daily, he was totally asymptomatic, his liver chemistry had normalized and his serum tested negative for ANA. Repeat CT scan showed significant reduction in size and well-enhancing pancreas. Repeat ERCP showed a normal pancreatic duct and an incomplete stricture in the distal common bile duct. He was advised to taper and stop steroids and is well on follow up one year later.

Case 2: A 53-year-old teetotaller presented with two episodes of pain suggestive of pancreatitis over 5 months and jaundice with cholestatic symptoms for 6 weeks. He had no past or family history of pancreatitis. On examination he was icteric. Laboratory data (Table) showed high ESR, hypergammaglobulinemia, normal serum amylase, high blood sugar and cholestatic picture on liver biochemistry. ANA was negative.

Ultrasonography revealed bulky hypoechoic pancreas with a hypoechoic lesion in the head of the pancreas. T2-weighted axial MRI showed a hypointense thin capsule surrounding the

| Table: Laboratory data before and after steroid therapy |
|-----------------|-----------------|-----------------|-----------------|-----------------|
|                 | Pre steroid     | Post steroid    | Pre steroid     | Post steroid    |
| Bilirubin (0.5-1 mg/dL) | 11.2            | 0.7             | 4.2             | 0.5             |
| Total protein (5-7 g/dL) | 7.4             | 6.8             | 7.4             | 7.3             |
| Globulin (2.5-3.5 g/dL)  | 3.0             | 2.6             | 3.9             | 3.0             |
| AST (5-40 IU/L)         | 176             | 21              | 150             | 32              |
| ALT (5-35 IU/L)         | 379             | 25              | 116             | 20              |
| SAP (40-125 U/L)        | 376             | 104             | 370             | 114             |
| Amylase (<200 U/L)      | 142             | 122             | 114             | 412             |
| Post prandial glucose (≤200 mg/dL) | 125          | 194             | 296             | 142             |
| ESR (mm/1st h)          | 22              | 18              | 115             | 82              |

ANA - Positive Negative Negative Negative
SAP - serum alkaline phosphatase. ANA - anti nuclear antibodies. Values in parentheses are normal range.

Fig 1: Case 1: Spiral CT of pancreas post steroid therapy, arterial phase, showing diffusely enlarged, sausage-shaped, poorly enhancing gland surrounded by thin capsule; main pancreatic duct is not dilated.
pancreatic body. CT scan revealed a bulky and sausage-shaped pancreas with decrease in contrast enhancement. In addition, there was mild intrahepatic biliary dilatation with thickened wall of the common bile duct. ERCP (Fig 2) showed main pancreatic duct attenuation and irregularity, distal bile duct stricture, multiple areas of intrahepatic biliary dilatation, finely serrated wall of bile duct, and pruning of peripheral bile ducts suggestive of sclerosing cholangitis and narrow-duct pancreatitis. Fine-needle aspiration of head of pancreas revealed only lymphoid cell aggregates. Gallium scan revealed increased accumulation of the isotope in the pancreas.

He was started on 40 mg prednisolone daily. Two months later he was totally asymptomatic, liver biochemical tests had normalized, and sugar level was better controlled. Repeat MR scan showed disappearance of the hypointense rim suggesting that the inflammatory capsule had resolved. Repeat CT scan showed significant reduction in the size of the pancreas with better enhancement. ERCP also showed smooth wall of the common bile duct, and resolution of the peripheral changes of sclerosing cholangitis noted in the previous ERCP. The pancreas duct had also increased in caliber.

Discussion

More than 150 cases have been reported as AIP or pancreatitis with narrow pancreatic duct in the Japanese literature. There have been no reports from India so far. Whether this reflects the rarity of the disease or a failure to recognize it as a clinical entity is unknown.

AIP has been reported in association with other autoimmune-related diseases such as Sjögren’s syndrome, sclerosing cholangitis, inflammatory bowel disease, systemic lupus erythematosus, Hashimoto’s thyroiditis, and retroperitoneal fibrosis. There is a male predominance, with most patients being older than 50 years. Patients present with mild abdominal pain, weight loss, and obstructive jaundice. Laboratory data usually show increased levels of serum pancreatic enzymes, hypergammaglobulinemia, and presence of autoantibodies such as antinuclear antibodies, and antibodies against lactoferrin, carbonic anhydrase type II and rheumatoid factor. Rise in IgG levels and elevation of IgG-containing immune complexes have been observed in a majority of patients. Diabetes mellitus Type 2 is often (43%-68%) observed in AIP.

Radiological findings are pathognomonic and different from classical chronic pancreatitis. The pancreas is diffusely enlarged, and appears “sausage-like”, with a capsule-like rim that appears as low density on CT, hypointense on T2-weighted MR images, and shows delayed enhancement on dynamic MRI. Pancreatic calcification or pseudocyst is seldom seen. ERCP shows segmental or diffuse irregular narrowing of the main pancreatic duct and distal bile duct strictures. Histology shows fibrotic changes with infiltration of lymphocytes and plasmocytes mainly around the pancreatic duct.

Clinical, biochemical and radiological response to steroids in AIP occurs in 2-3 weeks, with normalization of serum bilirubin within one month. A trial of steroid therapy has been suggested in patients where clinical suspicion of AIP is high and fine-needle aspiration biopsy is not diagnostic of malignancy.

Both our patients presented with pain suggestive of pancreatitis and obstructive jaundice. Our first case had an intrapancreatic biliary stricture, which has been described in the majority of patients with AIP. Cholangiogram in the second patient showed strictures of both intra- and extrahepatic bile ducts similar to primary sclerosing cholangitis; this has been described in association with AIP. In contrast to primary PSC, features that suggest AIP with sclerosing cholangitis are occurrence in older age group, high prevalence of autoantibodies, characteristic imaging of the pancreas, and good response to steroids. Steroid therapy is usually effective for stricture of CBD as well as the pancreatic duct. Normalization of pancreatic histology and improvement of type 2 diabetes mellitus on steroid therapy has also been shown.

Awareness of existence of this rare variant of chronic pancreatitis and the ability to make a diagnosis based on clinical, radiological and biochemical findings, avoids unnecessary resective procedures for suspected pancreatic adenocarcinoma in addition to providing complete recovery with steroids.

References


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