ATYPICAL PRESENTATION OF AUTOIMMUNE PANCREATITIS

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ABSTRACT

Autoimmune pancreatitis (AIP) is an inflammation in the pancreas from an autoimmune etiology. It is often difficult to diagnosis with significant morbidity and mortality. It can present with two subtypes of AIP type 1 and type 2. There is limited data on the treatment of autoimmune pancreatitis. Most patients are respond to corticosteroid therapy.

KEYWORDS

Atypical, autoimmune, pancreatitis

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INTRODUCTION

Autoimmune pancreatitis (AIP) is characterized clinically by frequent presentation with obstructive jaundice with or without a pancreatic mass, histologically by a lymphoplasmacytic infiltrate and fibrosis and therapeutically by a dramatic response to steroids.[1] AIP is classified into Type 1 or Lymphoplasmacytic sclerosing pancreatitis without granulocytic epithelial lesions (GELs) and Type 2 or Idiopathic duct centric pancreatitis (IDCP) or AIP with GELs.[2] Type 1 seems to be a part of an IgG4-related systemic disease. Type 2 is usually limited to pancreatic involvement only inflammatory bowel disease (IBD) is associated more often with Type 2 (30%–48%).[3] Ulcerative colitis accounts for 73% of IBD in AIP-2.[4]

CASE REPORT

A 32-year-old young gentleman presented with episodic epigastric pain. radiating to the back for 5 months and jaundice with pruritus and high-colored urine for 4 days before the hospital visit. He also complained of the passage of mucoid stool occasionally mixed with blood, 2-3 episodes per day for 4 days.

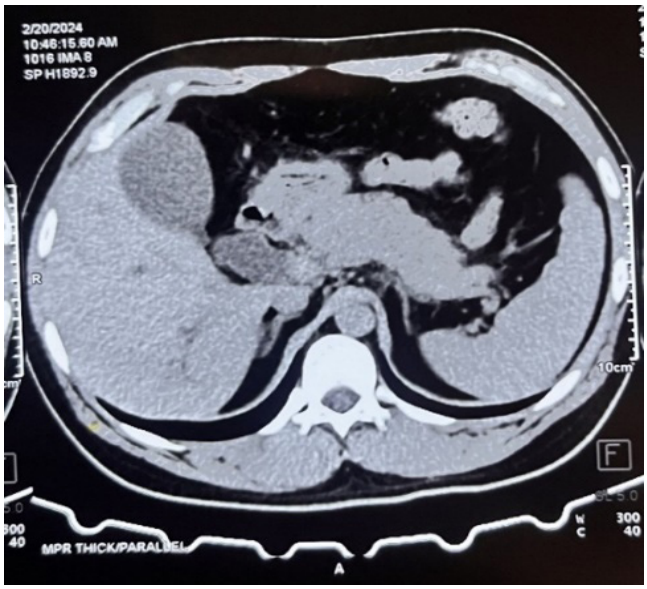

Fig. 1: CT scan showing enlarged pancreas

Blood investigations showed conjugated hyperbilirubinemia and raised liver enzymes and lipase. A computed tomography scan (Fig. 1) revealed a diffusely enlarged pancreas with effacement of the lobular contour, showing decreased enhancement during the early phase and delayed enhancement in the late phase. Diffuse narrowing of the main pancreatic duct and intrapancreatic portion of the common bile duct (CBD) with proximal CBD and IHBRs dilatation. EUS done showed bulky head, body, and tail of pancreas with normal Main pancreatic duct (MPD), dilated proximal CBD measuring 17.16 mm with distal CBD measuring 1.35 mm with compression noted in Mid CBD by the bulky pancreas. EUS-guided FNB from pancreatic tissue showed acini and ductal cells with fibrin and mixed inflammatory cells in the background. IgG4 level was significantly raised (2056.4 mg/dL).

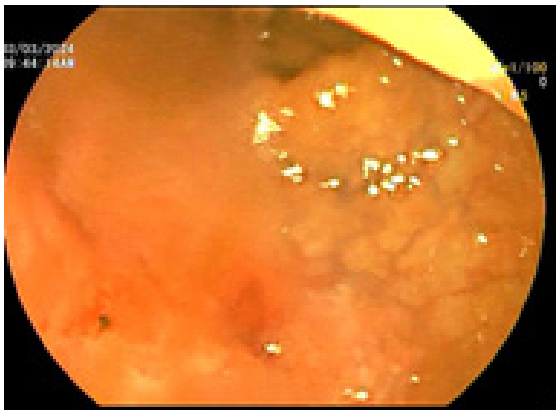

Fig. 2: Colonoscopy examination revealing rectal erythema and ulcer

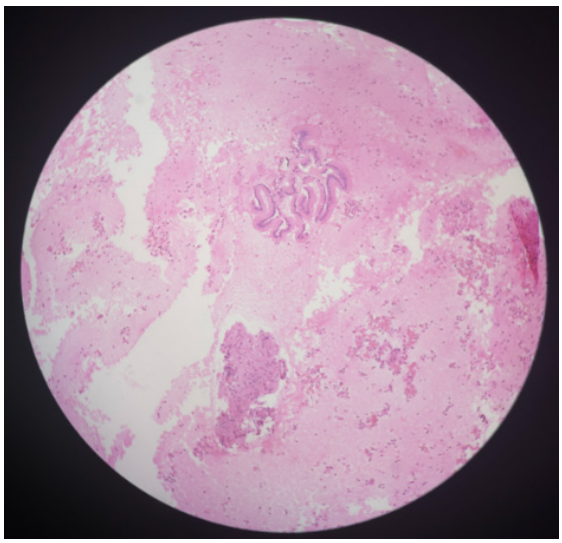

Fig. 3: Biopsy showing cryptitis, polymorphs, lymphoplasmacytic in lamina propria with mild crypt distortion

Magnetic resonance cholangiopancreatography was done due to the persistence of clinical presentation, which also revealed similar findings. A colonoscopy (Fig. 2) revealed rectal erythema with superficial ulcers. Biopsies showed cryptitis, polymorphs, and lymphoplasmacytosis in the lamina propria with mild crypt distortion suggestive of early Ulcerative colitis. Oral prednisolone was started at a dose of 40 mg daily. Rectal bleeding stopped, and bilirubin began to fall within a week and normalized after 8 weeks.

Ultrasonography showed a normal pancreas. IgG4 levels normalized. Steroid dose was reduced (tapered) down by 5 mg weekly.

DISCUSSION

Autoimmune pancreatitis is a distinct form of pancreatitis characterized clinically by frequent presentation with obstructive jaundice with or without a pancreatic mass, histologically by a lymphoplasmacytic infiltrate and fibrosis, and therapeutically by a dramatic response to steroids. Patients with obstructive jaundice with a diffusely enlarged pancreas (especially with a capsule-like rim) without pancreatic ductal dilatation/cutoff or pancreatic low-density mass on CT/MRI are highly likely to have AIP. In such patients, the presence of less collateral evidence is required to make the diagnosis of AIP. Marked elevation of serum IgG4 (>2 times the upper limit of normal) is strongly suggestive of AIP in the setting of obstructive jaundice/ pancreatic mass. Elevation in serological markers is not sufficient to make a diagnosis of AIP unless seen in the setting of typical imaging findings. As mentioned earlier, Type 1 often can be diagnosed without histology, but Type 2 requires an adequate histological specimen to make a definitive diagnosis. In patients with appropriate collateral evidence of AIP, response to steroids can confirm a strong suspicion of AIP. However, steroid trials as a means to diagnose AIP are to be used sparingly and should not be used as a substitute for a thorough search for an etiology. Steroid therapy leads to a reduction in IgG4 levels in AIP. However, falsely elevated IgG4 in pancreatic cancer and other non-AIP states also can decrease with steroid therapy. Therefore, a "response" of IgG4 to steroid treatment cannot be used to diagnose AIP. The distinction between the two types of AIP is often not straightforward. Clinical presentation may vary, and EUS-FNB is only approximately 83.3% sensitive.[5] Our patient, South Asian man with high levels of IgG4, suggested Type 1. On the contrary, young age with concurrent IBD-like presentation favoured Type 2 AIP. Type 1 AIP is more common in East Asia. However, Type 2 AIP is being increasingly diagnosed in Japan and Korea. This possibly could represent the underdiagnosis of Type 2 AIP.[6]

Despite intensive workup, the diagnosis of autoimmune pancreatitis can still be challenging. Some cases may present atypically with mixed clinical presentations, and the findings may not correlate adequately to classify it as Type 1 or Type 2 autoimmune pancreatitis despite serology, biopsy and correlation with extra-organ involvement and may need a prolonged follow-up despite good response with the steroids.

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