A RARE CASE REPORT ON INFLAMMATORY BOWEL DISEASE AND PRIMARY SCLEROSING CHOLANGITIS

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ABSTRACT

Inflammatory bowel (IBD), including Crohn's disease and Ulcerative colitis, is generally associated with hepatobiliary disorders. Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease characterised by inflammation and fibrosis in the bile ducts. This overlap presents different diagnostics and therapeutic challenges. A 41-year-old female patient presented with alterations in bowel habits, dyspeptic symptoms, weight loss, intermittent abdominal pain and mild swelling. The MR-cholangiopancreatogram (MRCP) showed Bile duct strictures suggestive of early PSC also having a positive ANA. Endoscopy reports revealed infective colitis and colonoscopy as Grade III pancolitis, ultimately leading to the confirmed diagnosis of an IBD-PSC overlap syndrome. Initially this condition is treated with Mesalamine (oral and suppository), folic acid supplements and supportive treatments. This report highlights the diagnostic challenges, monitoring strategies, and the importance of a multidisciplinary approach in managing this challenging patient population. It also provides a brief overview of the prevalence rates and clinical characteristics of IBD-PSC and PSC-IBD presentations.

Keywords: Primary sclerosing cholangitis (PSC), inflammatory bowel disease (IBD), MR-cholangiopancreatogram, ulcerative colitis.

INTRODUCTION

Inflammatory Bowel Disease (IBD), primarily Ulcerative Colitis (UC) and Crohn's disease (CD), are chronic, relapsing inflammatory disorders of the gastrointestinal tract [1]. Primary Sclerosing Cholangitis (PSC) is a progressive cholestatic liver disease characterized by inflammation and fibrosis of the bile ducts, leading to strictures and ultimately biliary cirrhosis and liver failure [2]. The coexistence of IBD and PSC, represents a unique clinical and pathological entity with distinct features, complications and surveillance needs [3].

The relationship between IBD and PSC is bidirectional, but the prevalence rates differ depending on which condition is diagnosed first. IBD + PSC; Among patients with IBD, the prevalence of PSC is estimated to be 2-7.5% [4]. The presence of PSC in IBD patients is often associated with a more extensive colitis, a milder colonic inflammatory phenotype, and a higher risk of colorectal cancer [5]. PSC + IBD; Conversely, a substantial majority of patients with PSC (approximately 70-80%) have co-existing IBD, with UC being the predominant type (over 80% of IBD in PSC patients are UC) [2]. In these cases, the IBD is often mild or even asymptomatic, and may affect a greater extent of the colon. The liver disease typically dictates the prognosis in PSC-IBD patients [1].

The exact pathogenesis of this association remains unclear, genetic predispositions, immune dysregulation, and altered gut microbiome are hypothesized to play a role. Diagnosis involves liver function tests (typically elevated ALP), MRCP imaging for biliary strictures and colonoscopy for colitis assessment and cancer surveillance. Differential diagnoses including autoimmune hepatitis, IgG4-related sclerosing cholangitis, secondary sclerosing cholangitis, drug induced liver injury, and cholangiocarcinoma were considered and excluded based on clinical, serological, and radiological findings [6].

There is no curative treatment for PSC aside from liver transplantation. IBD is managed with standard medications, while PSC requires close monitoring, supportive care and endoscopic intervention when needed [5].

Management of IBD includes 5-Aminosalicylic acid, corticosteroids, immunomodulators, Biologic Agents. Currently, no approved medical therapy halts PSC progression, but management focuses on monitoring, symptoms control and complication prevention.

CASE REPORT

A 43-year-old female patient presented with complaint of 3 months of alteration in bowel habits, dyspeptic symptoms and weight loss (15 kg over 3 months). Initial laboratory findings indicate systemic inflammation with an elevated ESR (80 mm/hr), total count 11440 and Hb
10.2 gm%. Upper endoscopy revealed Reflux Esophagitis, Hiatus Hernia and Antral Gastritis. Colonoscopy revealed Grade I Pan Colitis, consistent with Ulcerative Colitis. Biopsy from the colon demonstrated dense inflammatory cell infiltration, cryptitis and crypt abscesses. The High-Power review (HPR) further revealed focal active colitis, occasional cells exhibiting crypt architectural distortion, mucin depletion, and basal plasmacytosis, findings that confirmed chronic active Ulcerative Colitis.

Initial treatment included Mesalamine (oral and suppository forms), Folic acid supplementation, a supportive care. Symptoms improved partially, with reduced stool frequency and improved haemoglobin, but systemic inflammation persisted.

Routine liver function tests indicated a progressive rise in liver enzymes, AST 243 U/L, ALT 108 U/L and GGT 163. This biochemical pattern raised suspicion for Primary Sclerosing Cholangitis (PSC). These findings prompted further evaluation for hepatobiliary involvement, and MRI with MRCP was performed. The imaging study showed a tortuous appearance of the cystic duct and common bile duct with multiple short-segment stenosis and dilations, without evidence of stones or wall thickening. Periportal lymphadenopathy, mild hepatomegaly, and a focal hyperintense lesion measuring 8.6 mm in segment VIII were noted, along with a bulky spleen. There were no features of parenchymal fibrosis or cirrhosis. These findings were suggestive of early Primary Sclerosing Cholangitis.

Repeat colonoscopy continued to show diffuse pan colitis, and liver enzymes remained elevated (ALP 203 U/L, GGT 116 U/L), Antinuclear Antibody revealed positive at 3+ intensity, confirmed the presence of IBD-PSC overlap. The patient continues on Mesalaminemaintenance therapy, along with Ursodeoxycholic acid 300 mg and vitamin supplements. UC was stable. PSC is in early stage, requiring gastroenterology monitoring and multidisciplinary follow up.

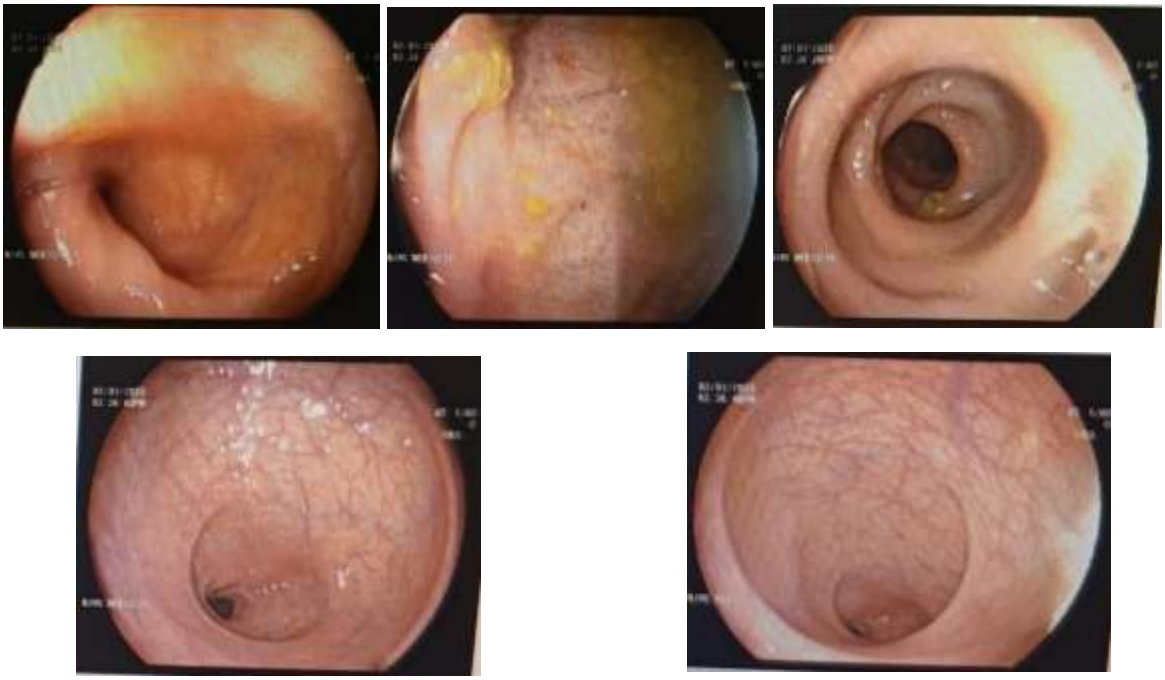

Figure 1: Oesophago-Gastro-Duodenoscopy

DISCUSSION

The coexistence of IBD and PSC represents a rare but clinically significant overall syndrome. Characteristically, intestinal inflammation may remain mild or well-controlled, whereas hepatobiliary disease progresses, independent of intestinal activity. Pathogenesis is believed to involve genetic predisposition, immune dysregulation, and gut-liver axis dysfunction, as discussed in key reviews and hypotheses [7,8]. Persistent elevation of cholestatic liver enzymes in IBD patients should raise suspicion for PSC. Conversely, PSC patients require regular colonoscopic screening for subclinical IBD [2].

Autoimmune markers, such as ANA further highlight the autoimmune contribution to hepatobiliary injury and may influence therapeutic response [9,10]. In overlap syndromes, long term risks include cirrhosis, portal hypertension, cholangiocarcinoma, and colorectal carcinoma, emphasizing the need for comprehensive biochemical, imaging, and endoscopic surveillance. Multidisciplinary management, involving gastroenterologists is fundamental for timely diagnosis and optimized patient outcomes.

CONCLUSION

This case report highlights the diagnostic complexity in patients with inflammatory bowel disease who develop unexplained liver enzyme abnormalities suggestive of primary sclerosing cholangitis. The demonstration histopathology report combined with positive ANA and characteristic MRCP findings, supports the diagnosis of an IBD-PSC overlap syndrome. Early recognition, supported by vigilant biochemical, imaging, and endoscopic surveillance, is crucial due to the potential progression to cirrhosis, portal hypertension, cholangiocarcinoma and colorectal cancer.

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