Primary Sclerosing Cholangitis: An Atypical Presentation

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Abstract

Primary sclerosing cholangitis (PSC) is a chronic inflammatory disease of bile ducts. Patients with PSC usually present with fatigue, jaundice and pruritus. Ultimately it leads to cirrhosis of liver and portal hypertension. But it rarely presents with decompensated liver disease without any previous symptoms. Here we report a case of PSC which presented with features of decompensated liver disease with K-F rings in the eyes.

Introduction

Patients with primary sclerosing cholangitis usually present with pruritus, jaundice, upper abdominal discomfort. Raised serum alkaline phosphatase and beaded appearance of biliary systems on MRCP or Cholangiography are characteristics of PSC. Our case presented with features of decompensated liver disease with K-F ring. Serum ceruloplasmin and 24 hours urinary copper were within normal limits. Our case was likely to be a case of primary sclerosing cholangitis.

The Case

A 28 year old male patient presented with history of yellow discolouration of urine and conjunctiva for two months and swelling of abdomen and feet for 3 weeks. Jaundice was insidious in onset but progressive. Jaundice was not preceded by typical prodromal features but it was associated with anorexia, upper abdominal pain, high coloured urine and pruritus. Stool colour was normal and no steatorrhoea. There was no history of fever, weight loss, hematemesis, melena, any drug intake, blood transfusion, sexual exposure or any surgery. There was no history suggestive of any neuropsychiatric manifestations. There was no significant family history.

On general examination, there were mild pallor, jaundice, bipedal pitting edema, generalised hyperpigmentation of skin, engorged epigastric vein. Jugular venous pressure was not elevated. No spider, gynaecomastia or lymphadenopathy was detected. Blood pressure was 120/70mm of Hg. Pulse was 62/min. On G.I. System examination, dilated tortuous abdominal veins with flow away from umbilicus were observed. There was ascites. Hepatic span was 11cm with palpable left lobe of liver. Spleen was enlarged 4cm below the left costal margin along its axis. Other systems examination reveal no abnormality. But Slit Lamp examination of eyes revealed bilateral K-F rings (Figures 1 and 2).

Investigations showed Hb-8.6 gm/dl, total count-8400/cu.mm, N64, L36, E0, M0, ESR-66mm in 1st hour,platelets-1.1lacs/c.mm. Fasting Blood sugar-79mg/dl,urea-32mg/dl,creatinine-1.1mg/dl; Bilirubin-13.5mg/dl, direct bilirubin-10.9mg/dl, AST-112U/L, ALT-62U/L, ALP-880U/L, Serum protein-7.0gm/dl,Albumin-2.4 gm/dl and globulin 4.6gm/dl. Prothrombin time was 19.2 seconds with control 12.0 seconds and INR was 1.92. Ascitic fluid study showed SAAG-1.3gm/dl and cell count 80 cells/cmm with lymphocytes 70%.Ultrasonography of abdomen showed heterogenous echotextured liver, dilated IHBR and ascites with splenomegaly. Portal vein diameter was 11mm. Upper G.I. Endoscopy showed early esophageal varices. HbsAg, Anti HCV,ELISA for HIV were all non reactive. Serum ceruloplasmin was 42.7mg/dl (Normal-18-35mg/dl). 24 hour urinary copper was 17 µgm (normal<100 µgm). Magnetic resonance cholangiography (MRC) (Figure 3) revealed mildly dilated intrahepatic biliary channels with some segments showing focal dilatation and stenosis (beaded appearance) particularly at confluence of hepatic ducts. Calculi were seen at right hepatic duct. Proximal bile duct showed narrowing. No intraluminal mass was seen in common bile duct. MRC findings were suggestive of sclerosing cholangitis. Anti nuclear antibody, anti mitochondrial antibody,
Primary sclerosing cholangitis (PSC), a chronic inflammatory disease of the bile ducts, typically affects anywhere from the small interlobular to large intra- and extrahepatic bile ducts, and leads to progressive bile duct fibrosis and liver cirrhosis. No apparent etiology or disease association is found with PSC. But it can occur in association with inflammatory bowel disease, systemic fibrosing conditions or with other autoimmune disorders. Patients present with fatigue, pruritus, jaundice or abdominal discomfort in 60% of cases. Symptoms such as pruritus and right upper abdominal pain are the most common intermittent symptoms, occurring with considerable individual variation and resolving spontaneously, in most cases. Approximately 15% to 55% of PSC patients are asymptomatic at presentation. The natural history of the disease is currently being evaluated but is generally recognized to be slowly progressive, leading to complications of chronic cholestasis, portal hypertension and biliary cirrhosis. In unusual instances, patients with PSC present with complications of cirrhosis and portal hypertension. A cholestatic picture of liver function with an elevation in serum alkaline phosphatase level is the biochemical hallmark of PSC, although some patients may have normal alkaline phosphatase levels. Testing for specific autoimmune antibodies does not contribute to the diagnosis of PSC though multiple autoantibodies can be detected in PSC. Antinuclear antibodies and smooth muscle antibodies can be found in 20% to 60% of patients, usually in lower titres than those observed in autoimmune hepatitis. In contrast, antimitochondrial antibodies are seldom seen in patients with PSC. The prevalent autoantibody reactivity is a perinuclear antineutrophilic autoantibody (perinuclear antineutrophil cytoplasmic antibody), present in approximately 80% of patients, but lacking in diagnostic specificity. Cholangiography is considered to be the gold standard for the diagnosis of PSC. Cholangiography shows the multifocal strictures, irregularities (beading) of intra and/or extrahepatic bile ducts. Magnetic resonance cholangiography (MRC) for detecting PSC has emerged as an accurate, rapid, noninvasive alternative examination of the biliary tract, and is commonly used in multiple centres.

Our case presented with features of decompensated liver disease. On examination there was features K-F ring with chronic liver disease. K-F ring may be found in Wilson’s disease as well as other conditions like primary biliary cirrhosis, chronic aggressive hepatitis. But serum ceruloplasmin and 24 hour urinary copper estimation all were within normal limits. Features of chronic liver disease with raised ALP then lead us to do a MRCP which revealed the diagnosis of Primary Sclerosing Cholangitis. So patient of PSC presenting with features of decompensated liver disease without any previous symptoms is unusual.

Discussion

Primary sclerosing cholangitis (PSC), a chronic inflammatory disease of the bile ducts, typically affects anywhere from the small interlobular to large intra- and extrahepatic bile ducts, and leads to progressive bile duct fibrosis and liver cirrhosis. No apparent etiology or disease association is found with PSC. But it can occur in association with inflammatory bowel disease, with systemic fibrosing conditions or with other autoimmune disorders. Patients present with fatigue, pruritus, jaundice or abdominal discomfort in 60% of cases. Symptoms such as pruritus and right upper abdominal pain are the most common intermittent symptoms, occurring with considerable individual variation and resolving spontaneously, in most cases. Approximately 15% to 55% of PSC patients are asymptomatic at presentation. The natural history of the disease is currently being evaluated but is generally recognized to be slowly progressive, leading to complications of chronic cholestasis, portal hypertension and biliary cirrhosis. In unusual instances, patients with PSC present with complications of cirrhosis and portal hypertension. A cholestatic picture of liver function with an elevation in serum alkaline phosphatase level is the biochemical hallmark of PSC, although some patients may have normal alkaline phosphatase levels. Testing for specific autoimmune antibodies does not contribute to the diagnosis of PSC though multiple autoantibodies can be detected in PSC. Antinuclear antibodies and smooth muscle antibodies can be found in 20% to 60% of patients, usually in lower titres than those observed in autoimmune hepatitis. In contrast, antimitochondrial antibodies are seldom seen in patients with PSC. The prevalent autoantibody reactivity is a perinuclear antineutrophilic autoantibody (perinuclear antineutrophil cytoplasmic antibody), present in approximately 80% of patients, but lacking in diagnostic specificity. Cholangiography is considered to be the gold standard for the diagnosis of PSC. Cholangiography shows the multifocal strictures, irregularities (beading) of intra and/or extrahepatic bile ducts. Magnetic resonance cholangiography (MRC) for detecting PSC has emerged as an accurate, rapid, noninvasive alternative examination of the biliary tract, and is commonly used in multiple centres.

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