

Case Report

A case of cutaneous scleroderma with primary sclerosing cholangitis

Nandeesh H. P., Jeevan H. R., Deepak Suvarna., Chandra Babu D., Indrajit Suresh, Subashchandra B. J.¹, Savitha T. C.²

Departments of Gastroenterology, ¹Medicine, and ²Dermatology, J.S.S. Medical College & Hospital, Mysore, Karnataka, India

Abstract

Sclerosing cholangitis comprises of a spectrum of cholestatic conditions that are characterized by patchy fibrosis, inflammation and destruction of intra hepatic and extrahepatic ducts. We report a case of a 42 year old woman who presented with darkening of skin with yellowish discolouration of the eyes. Clinical examination revealed icterus, taut skin with hepatosplenomegaly. Liver function tests showed a cholestatic picture. Skin biopsy showed features of cutaneous scleroderma. MRCP and Liver biopsy was suggestive of sclerosing cholangitis.

Key words

Cholestatic liver disease, cutaneous scleroderma, primary sclerosing cholangitis

Introduction

Primary sclerosing cholangitis (PSC) is a chronic cholestatic liver disease with obliterative inflammatory fibrosis of the bile ducts. The most frequent is primary sclerosing cholangitis (PSC), an idiopathic disorder that usually occurs in association with inflammatory bowel disease but may develop independently. Secondary sclerosing cholangitis is similar to PSC but develops as a consequence to obstructive, toxic, ischaemic and neoplastic causes.^[1] The prevalence is estimated to be about 6 cases per 100,000 population.

PSC is strongly associated with inflammatory bowel disease, especially ulcerative colitis, and is often complicated by cholangiocarcinoma.^[2] It is also associated with other autoimmune disorders like Type 1 Diabetes mellitus, Thyroid disorders, Psoriasis, Rheumatoid arthritis, Celiac disease, SLE, Sarcoidosis.

The association between Scleroderma and PSC(<1%) is a rare occurrence. Systemic sclerosis (SS) is a multisystem

disorder characterized by fibrosis that involves the skin and a variety of internal organs, although hepatic involvement in scleroderma is rare.^[3] Therefore, hereby we report a case of primary sclerosing cholangitis with cutaneous scleroderma.

Case Report

A 42-yr old lady who had darkening of skin over face and extremities with Raynaud's phenomenon presented with jaundice, abdominal distension with upper GI bleed. There were no symptoms of sicca syndrome, mouth ulcers, photosensitivity, skin rashes, dysphagia, abdominal pain, diarrhoea, arthralgias, arthritis or eye problems. On examination she was icteric with presence of taut skin. Abdominal examination showed hepatosplenomegaly with mild ascites. All other systems were normal.

MRCP showed multi focal segments of narrowing and dilatation of intra hepatic and extra hepatic biliary system suggestive of sclerosing cholangitis [Figure 1].

Liver biopsy showed fragments of hepatic parenchyma displaying loss of architecture, hepatocytes with feathery degeneration and cholestasis in pseudo lobules. There was marked increase in fibrosis and chronic inflammation in portal tracts and focal areas showed bile duct proliferation with periductal fibrosis, suggestive of primary sclerosing cholangitis [Figure 2].

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Address for correspondence:

Dr. H. P. Nandeesh, Department of Gastroenterology, JSS Medical College Hospital, Mysore, Karnataka, India.

E-mail: nandeesh_gastro@yahoo.co.in

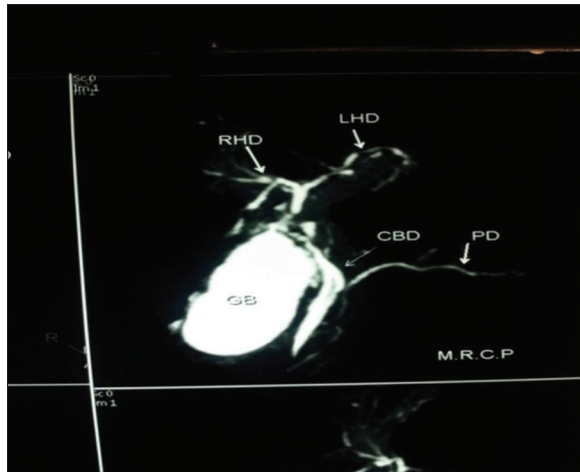


Figure 1: Magnetic resonance cholangiopancreatography

Skin biopsy was suggestive of cutaneous scleroderma. The patient was treated with Proton pump Inhibitor, Ursodeoxycholic acid, Beta blockers, Endoscopic Variceal Ligation, Fresh frozen plasma and Packed Red cell transfusion. Biliary stenting was deferred due to extensive biliary strictures at multiple levels and advanced liver disease (Child C). The option of Liver transplantation was considered [Table 1].

Discussion

Systemic sclerosis is a widespread disorder of connective tissue that involves the skin (scleroderma) and various organs of the body, including the heart, lungs, kidneys, and gastrointestinal tract.^[4]

Sclerosing cholangitis comprises of a spectrum of cholestatic conditions that are characterized by patchy inflammation, fibrosis and destruction of the intrahepatic and extra hepatic bile ducts. These conditions are chronic, progressive disorders in which persistent biliary damage may lead to biliary obstruction, biliary cirrhosis and hepatic failure with associated complications.^[5] The first description of sclerosing cholangitis is credited to Delbet in 1924.^[6]

The diagnosis of PSC is based on cholangiographic findings in the setting of consistent clinical, biochemical, serologic and histologic findings as well as exclusion of secondary causes of sclerosing cholangitis. The characteristic cholangiographic findings are multi focal strictures and segmental dilatations in the absence of secondary causes of sclerosing cholangitis.^[7]

ERCP is considered the standard for establishing a diagnosis of PSC but carries a risk for complications of up to 10% in patients with PSC. Magnetic resonance cholangio- pancreatography has largely replaced ERCP as a diagnostic tool as a result of the improvements in image quality and its non invasive nature.^[8]

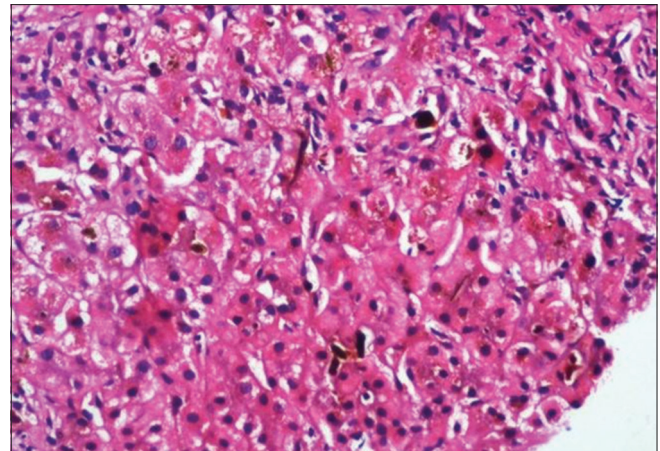


Figure 2: Liver biopsy –Low power field x 400

Table 1: Investigations

Haemoglobin: 8 gm/dl	USG abdomen: Hepatosplenomegaly, intrahepatic biliary radicles and common bile duct were normal, portal vein: 15 mm, splenic vein: 10 mm with mild ascites
TC : 6500 cells/mm ³	Upper GI endoscopy: Grade III to 1V esophageal varices with red colour signs, no gastric varices, endoscopic variceal ligation done
DC : N (70%) , L (30%)	Colonoscopy: Normal
Platelet count :1.5 lakhs/mm ³	ANA: Detected (anti centromere pattern suggestive of scleroderma)
PBS: Microcytic hypochromic anaemia	Anti ds DNA: Not detected
PT INR: 1.64, test (19), control (13)	ASMA: Negative
Total bilirubin: 7.10mg/dl	ANTI LKM1: Negative
Direct bilirubin: 4.10mg/dl	Anti Mitochondrial antibody: Negative
Total proteins: 6.0 gm/dl	pANCA: Negative.
Albumin: 3.1mg/dl	TSH: Normal
A/G ratio: 1.0	RA factor: Negative
AST: 122U/L	CK-NAC: Normal.
ALT: 63U/L	Iron indices: Normal
ALP: 378U/L	HRCT thorax: Normal
ECG, chest X-ray, ECHO: Normal	IgG4: 0.26g/dl (normal range: 0.03-2.0)
HIV, HBs AG , Anti HCV (ELISA): Non reactive	

USG=Ultrasonogram, TC=Total leukocyte count, DC=Differential count, GI=Gastrointestinal, ANA=Anti-nuclear antibody, ds DNA=Double stranded Deoxyribo nucleic acid, ASMA=Anti smooth muscle antibody, ANTI LKM1=Anti liver kidney microsome antibody, p ANCA= Peri nuclear anti-nuclear cytoplasmic antibody, TSH=Thyroid stimulating hormone, RA=Rheumatoid arthritis, AST=Aspartate amino transferase, CK-NAC=Creatinine kinase-N Acetyl Cysteine, ALT=Alanine amino transferase, ALP=alkaline phosphatase, HRCT=High resolution tomography, ECG=Electro cardio gram, ECHO=Echo cardio gram, HIV=Human immunodeficiency virus, HBs AG: Hepatitis B surface antigen, HCV=Hepatitis C virus, ELISA=Enzyme linked immuno sorbent assay

Although the cause of both SS and PSC is largely unknown, there is evidence of immune system abnormalities and of genetic influences in these disorders. The most commonly recorded association of the major histocompatibility complex (MHC) with scleroderma is with the HLA B8, DR3-containing haplotype and the high frequency of HLA DR3 encountered in PSC patients must be noted.^[9]

Primary sclerosing cholangitis in patients with systemic sclerosis might be the consequence of the widespread alteration of connective tissue presented in systemic sclerosis through the abnormal collagen deposition in the bile ducts epithelium.^[10]

To summarize, scleroderma is a multisystem disorder affecting skin, gastrointestinal system, lungs and hepatobiliary system. In a patient with cutaneous scleroderma with dearranged liver function test is worth evaluating for primary sclerosing cholangitis.

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