

## Case Report on Sclerosing Cholangitis

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**Abstract:** Sclerosing cholangitis is a cholestatic liver disease of chronic duration. The main feature of the disease is inflammation and fibrosis of bile duct leading to cirrhosis and final treatment is liver transplantation with less life expectancy. It is most commonly seen in patients with inflammatory bowel disease (IBD). Primary sclerosing cholangitis is of unknown etiology and of immune mediated disease. There are newer techniques like ERCP (Endoscopic Retrograde Cholangiopancreatography) and MRC (Magnetic Resonance Cholangiography) for diagnosing sclerosing cholangitis. There is no proven medical treatment for sclerosing cholangitis. The disease can lead to complications such as dominant strictures, secondary bacterial infection and malignancy mainly cholangiocarcinoma. Liver transplantation is the only therapy recommended for sclerosing cholangitis.

**Keywords:** Sclerosing Cholangitis, Diagnosis, Therapy, Cholangiocarcinoma, Liver Transplantation

### INTRODUCTION

Sclerosing cholangitis is a disease causing inflammation and fibrosis of bile ducts. This leads to obstruction in the flow of bile to intestine and also can lead to cirrhosis of liver [1]. Sclerosing cholangitis is idiopathic and mostly assumed to be auto immune origin, though doesnot respond to immuno suppressants [2]. The sclerosing cholangitis is multifactorial. There are associations between sclerosing cholangitis and human leukocyte

antigen (HLA). The levels of serum alkaline phosphatase are increased markedly. Sclerosing cholangitis have auto antibodies and abnormal immunoglobulins. Approximately 80% of people with sclerosing cholangitis have perinuclear Anti-Neutrophil Cytoplasmic Antibodies (ANCA). Antinuclear antibody and anti-smooth muscle antibody are found in 20-50% of patients [4-7]. Sclerosing cholangitis have high associations with inflammatory bowel diseases including ulcerative colitis and Crohn's disease [3].

### CASE REPORT

A 46 years old female came to emergency department with complaints of fever on and off for past one month, pain abdomen for 15 days and vomiting and decreased appetite for past 3 days. Patient also had H/O easily fatigability and generalized itching increased more in past few days. On General examination she was conscious, oriented and co-operative, afebrile. Patient had pallor and icterus and also B/L pedal edema was present. Vitals are normal. On local examination patient had mildly distended abdomen with right hypochondriac tenderness and hepatomegaly. Other systemic examinations were found to be normal. His blood sample was taken and sent for biochemical analysis.

### LAB INVESTIGATIONS

#### CBC

Haemoglobin- 8.9gm% , TLC-26330, N-91% L-3.9% E-0.4%, RBC -3.20 million, MCV- 84.9 fl, MCH-27.28pg, MCHC- 32.8, Platelet – 2.44 lakhs/cu.mm

#### LFT

Bilirubin (T) and (D): 12.5mg/dl and 10mg/dl, bilirubin indirect: 2.3mg/dl, SGOT: 66IU/L, SGPT: 22IU/L, ALP: 775 IU/L, total protein: 7.3g/dl, albumin: 2.2 g/dl, globulin: 5.1g/dl, A:G ratio: 0.4, Gamma GT(GGT): 192 IU/L

#### RFT

Urea – 50mg%, Creatinine – 1 mg%  
Blood sugar – 110mg%

#### Serology

ANCA- positive [6].

#### Urine routine

Colour: yellow, appearance: clear, pH:8.

Proteins, sugar, bile salts and bile pigments: nil

Ascitic fluid tapping:

Total count: 502cells/cu.mm

Differential count: neutrophils: 20% and lymphocytes: 80%

### Ultrasonography Abdomen

- Chronic liver parenchymal diseases
- GB sludge
- Common bile duct appears dilated with sludge
- Hepatosplenomegaly with ascites
- Umbilical hernia

### TREATMENT

Medical therapies have no benefits over sclerosing cholangitis

- Tacrolimus have shown subsequent improvement in therapy in biochemical values of ALP(8)
- Ursodeoxycholic acid has also shown changes in histological appearance. But not yet proven to have survival outcome(9,10,11)

### Surgical therapies

- Biliary surgery: strictures can be removed surgically by dilatation or by choledochojejunostomy. But this therapy has become out-fashioned after recent advances in endoscopic techniques.
- Liver transplantation: has shown improved survival rates and also liver transplantation must be considered before the advance stages of disease [21].

### DISCUSSION

Sclerosing cholangitis is rare in India. It can be seen in only 4% of patients with IBD. Males are usually more prone for the disease and commonly associated with IBD. Sclerosing cholangitis shows a marked increase in serum autoantibodies levels. ANCA – Anti Neutrophil Cytoplasmic Antibodies are present in 87% of patients with sclerosing cholangitis and anticardiolipin (aCL) in 66% and antinuclear antibodies (ANA) in 53%. It has increased risk of hepatobiliary malignancy mainly cholangiocarcinoma [13] and life incidence of CCA is 6-23%. ERCP- endoscopic retrograde cholangiopancreatography is successful demonstrating the biliary tree in 95% of cases. Procedure related complications for ERCP occur in 3-8% including abdominal pain, pancreatitis, bleeding, common bile duct perforation, biliary sepsis and death [14,15]. MRC- Magnetic resonance cholangiography has been found more accurate comparable to ERCP [16-20].

### CONCLUSION

Sclerosing cholangitis is an immune mediated liver disease associated with morbidity and mortality. There are no proper medical treatments available for sclerosing cholangitis, yet many therapies are yet to be

proven. Studies are done for development of therapies and to improve health and halt progress of the disease, thereby decreasing incidence of complications and advanced liver diseases and need for transplantation.

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