



(CASE REPORT)



RAPIDLY PROGRESSIVE PRIMARY SCLEROSING CHOLANGITIS IN A YOUNG WOMAN WITHOUT IBD

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ABSTRACT

Background: Primary Sclerosing Cholangitis (PSC) is a progressive cholangiopathy characterized by multifocal stricturing and fibrosis of the intrahepatic and/or extrahepatic bile ducts. The disease generally follows an indolent course over several years; however, rapidly progressive variants leading to early cirrhosis and liver transplantation are rarely reported, particularly in young woman without associated inflammatory bowel disease (IBD).

Case Presentation: A 23-year-old woman presented with jaundice and pruritus in May 2025. Initial investigations demonstrated cholestatic liver dysfunction with elevated bilirubin and alkaline phosphatase levels. Viral, autoimmune, infectious, and metabolic etiologies were excluded. MRCP revealed multifocal biliary strictures with non-visualization of the common hepatic duct and bilateral hepatic ducts, associated with lobar atrophy and caudate lobe hypertrophy, highly suggestive of primary sclerosing cholangitis. Liver biopsy demonstrated biliary-pattern injury with periductal fibrosis consistent with PSC. Despite treatment with ursodeoxycholic acid and symptomatic therapy, she experienced rapid progression of jaundice and cholestasis within six months. Repeat evaluation revealed worsening of hyperbilirubinemia, coagulopathy, and established biliary cirrhosis. Following multidisciplinary assessment, the patient underwent deceased donor orthotopic liver transplantation with hepaticojejunostomy in December 2025. She remains clinically stable on follow-up. Incidentally, her father had ulcerative colitis for 25 years, developed sigmoid colon cancer in 2020 resulting in a total proctocolectomy with ileal pouch-anal anastomosis (IPAA).

Conclusion: This case highlights an aggressive phenotype of PSC with unusually rapid progression to end-stage liver disease in the absence of IBD. Early recognition of aggressive disease behavior and timely referral for transplantation is essential for favorable outcome.

Keywords: Primary Sclerosing Cholangitis; Cholestasis; Biliary Cirrhosis; Liver Transplantation; IBD, Ulcerative Colitis.

1 INTRODUCTION

Primary Sclerosing Cholangitis is a chronic cholestatic liver disorder characterized by progressive inflammatory and fibrotic destruction of intrahepatic and/or extrahepatic bile ducts. The disease predominantly affects young males and is strongly associated with inflammatory bowel disease, especially ulcerative colitis. The natural history is usually slowly progressive, with median transplant-free survival ranging between 10 and 20 years.

Rapid progression to biliary cirrhosis and liver failure within months is uncommon and sparsely reported in literature. Such cases pose diagnostic and therapeutic challenge, particularly when inflammatory bowel disease is absent. We report a rare case of rapidly progressive PSC in a young woman leading to end-stage liver disease requiring deceased donor orthotopic liver transplantation (LT) within months of diagnosis.

2 CASE PRESENTATION

A 23-year-old woman presented in May 2025 with complaints of jaundice and progressive pruritus of one week duration. There was no history of abdominal pain, fever, gastrointestinal bleeding, altered sensorium, weight loss, prior liver disease, alcohol intake, hepatotoxic drug exposure, blood transfusion, or high-risk behavior. Family history was significant for ulcerative colitis in her father, who had been under follow-up for 20 years and developed carcinoma colon for which proctocolectomy followed by IPAA was done in 2020.

On examination, the patient was icteric, hemodynamically stable, without ascites, pedal edema, hepatic encephalopathy, or peripheral stigmata of chronic liver disease.

Initial biochemical investigations revealed cholestatic liver dysfunction.

Viral serologies including hepatitis B surface antigen, anti-hepatitis C virus antibody, IgM HAV, IgM HEV and HIV 1 and 2 antibodies were non-reactive. Infectious workup including HSV-1 IgM, HSV-2 IgM, Epstein-Barr virus, and cytomegalovirus IgM was negative (Table 1).

Table 1: Lab investigations

Parameter	05/05/2025	12/10/2025
Total bilirubin	4.80 mg/dL	13.58 mg/dL
Direct bilirubin	1.60 mg/dL	9.69 mg/dL
Indirect bilirubin	0.70 mg/dL	3.89 mg/dL
Alkaline phosphatase	348 U/L	292 U/L
SGOT (AST)	56 U/L	76.8 U/L
SGPT (ALT)	52 U/L	37 U/L
GGT	114 U/L	87.4 U/L
Total protein	3.80 g/dL	
PT	15.10 sec	
INR	1.08	2.18
Serum calcium	9.5 mg/dL	
Ceruloplasmin	43.80 mg/dL	
HBsAg	Non-reactive	
Anti- HCV	Non-reactive	
IgM HAV	Negative	
IgM HEV	Negative	
HIV 1/2	Non-reactive	
HSV-1 IgM	Negative	

HSV-2 IgM	Negative	
Epstein-Barr virus	Negative	
CMV IgM	Negative	

Autoimmune and metabolic evaluation revealed negative anti-mitochondrial antibody, ANA, LKM antibody, normal serum angiotensin-converting enzyme levels, normal serum calcium and ceruloplasmin level of 43.80 mg/dL.

Ultrasonography of the abdomen demonstrated mild focal intrahepatic biliary radicle dilatation (IHBRD), moderate hepatomegaly, enlarged caudate lobe, and a normal gallbladder.

MRCP demonstrated moderate atrophy of the right lobe of the liver with mild atrophy involving segments II and III of the left lobe. These regions showed T2 hyperintensity and T1 hypointensity. Reduced calibre of the right portal vein and its segmental radicles involving segments II and III was observed with associated minimal IHBRD. Moderate hypertrophy of segment IV and mild-to-moderate caudate lobe hypertrophy was noted. (Fig 4 and 5)

There was non-visualization of the common hepatic duct and bilateral hepatic ducts with multifocal strictures and narrowing associated with intervening and peripheral biliary dilatation involving both hepatic lobes. Tapering of the distal common bile duct was noted, while the remaining common bile duct measured 3.5 mm in diameter.

Triphasic CECT abdomen confirmed moderate right lobe atrophy, mild atrophy of segments II and III, perfusional changes, attenuated right and left hepatic ducts, and compensatory hypertrophy of segments IV, V and caudate lobe. Fibroscan demonstrated liver stiffness of 14.1 kPa with ultrasound attenuation parameter of 275 dB/m.

The patient was initiated on ursodeoxycholic acid and antihistamines for symptomatic management. Ultrasound-guided liver biopsy demonstrated maintained hepatic architecture with oedematous expanded portal tracts showing ductular proliferation and mild inflammatory infiltrate. Florid duct lesions were absent, but periductal fibrosis was identified. Interface hepatitis, emperipolesis, hepatocyte rosetting, steatosis, ballooning degeneration, Mallory–Denk bodies were not seen. Copper stain demonstrated positivity in periportal hepatocytes. Nodular regenerative hyperplasia suggestive of vascular flow abnormality was also noted. Histopathological impression favored biliary-pattern injury probably related to primary sclerosing cholangitis without definite overlap syndrome. Fibrosis stage was F2. (Fig 1)

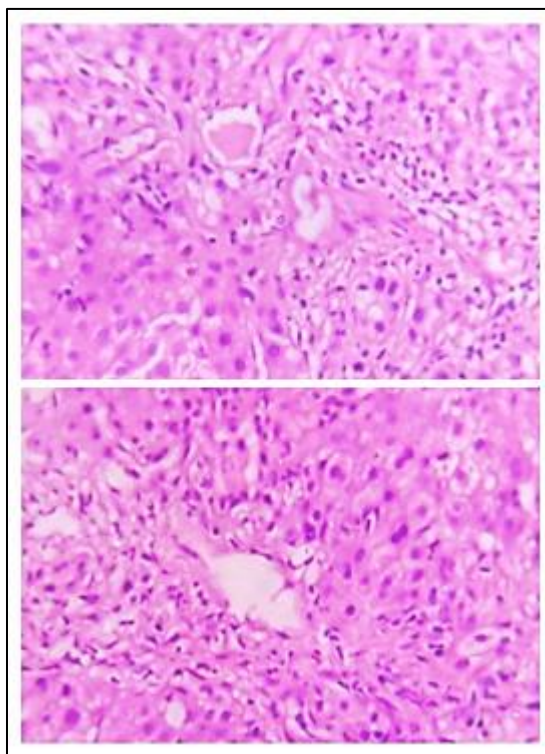


Figure 1: Liver biopsy before liver transplant

Colonoscopy and colonic biopsies performed to evaluate associated inflammatory bowel disease, was negative for features of inflammatory bowel disease. (Fig 2)

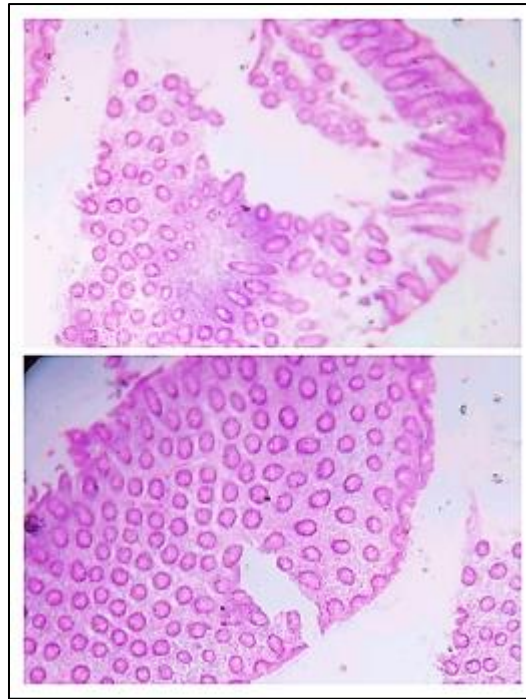


Figure 2: Colon biopsy of patient before liver transplant

Biopsy from the colonic mucosa shows mild inflammation- no features of inflammatory bowel disease – Ulcerative Colitis noted.

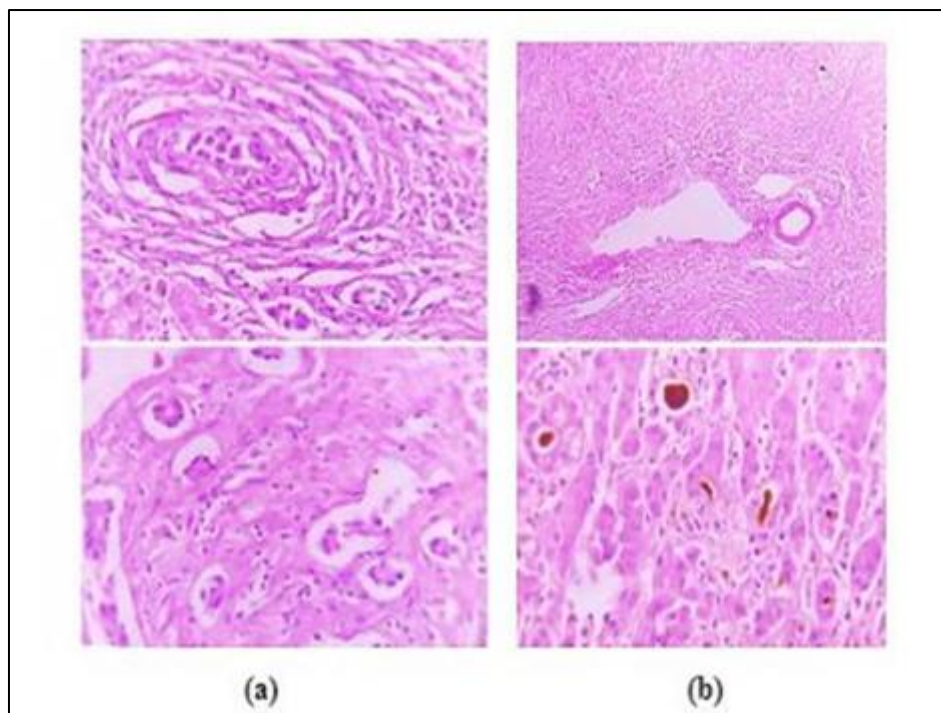


Figure 3a & 3b: Liver biopsy of Explant

Biopsy from liver explant shows periductal fibrosis, concentric minor with mild inflammation in portal, hepatocyte shows bile stasis or cholestasis

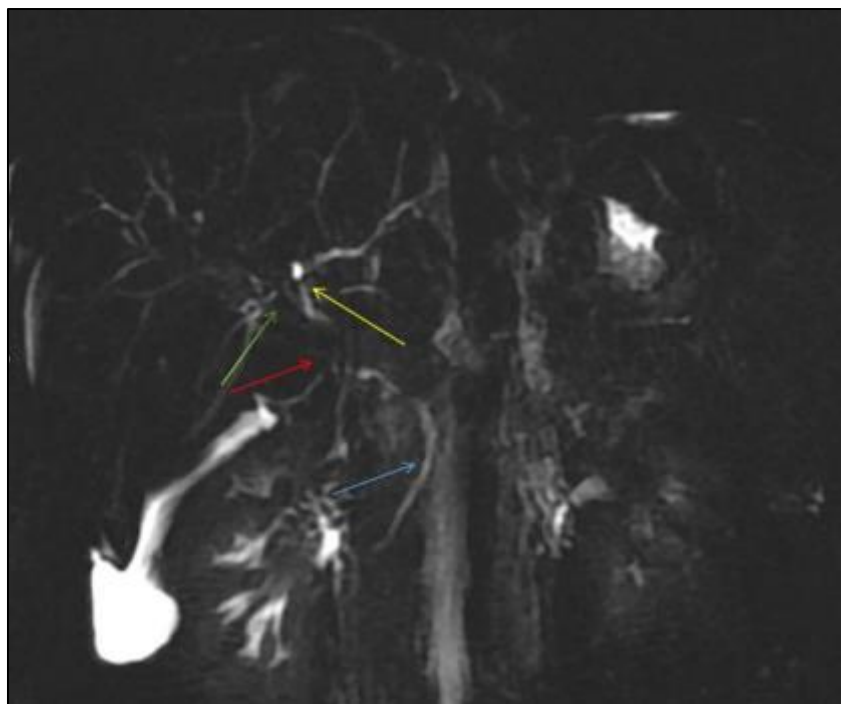


Figure 4: MRCP image

- Red arrow- Non visualized proximal CBD and part of hepatic duct.
- Green arrow - Non visualized RHD.
- Yellow arrow - Stricture of LHD
- Blue arrow- Mid -distal CBD is normal

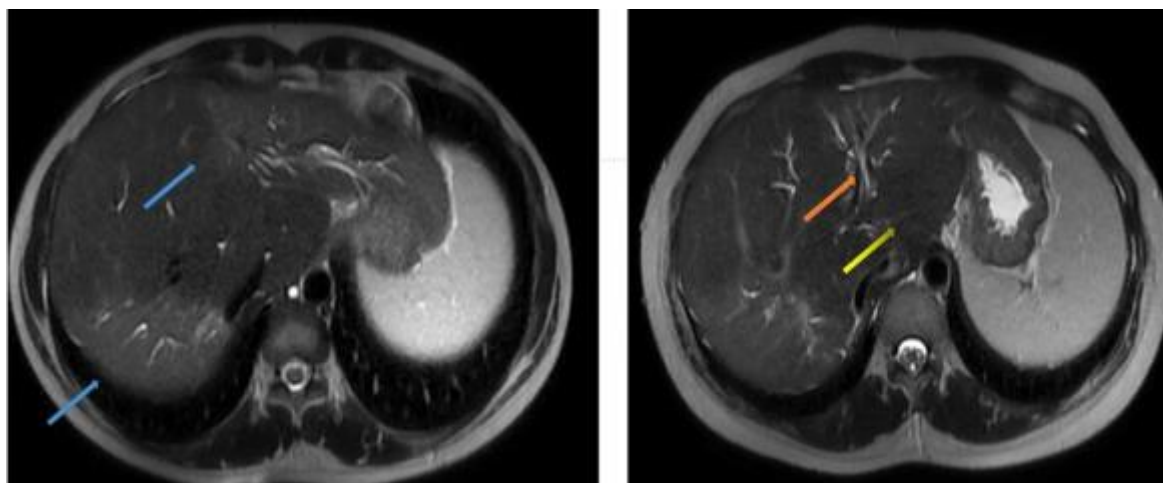


Figure 5: MRI Liver- Axial T2-weighted images

- Blue arrows - T2 hyperintensity with mild atrophy
- Yellow arrow - Caudate hypertrophy with normal signal
- Orange arrow - Minimal IHBRD

Despite medical management, there was progressive worsening of jaundice and pruritus over the subsequent six months. Owing to rapid disease progression, she was referred for liver transplantation evaluation in August 2025.

Pre-transplant evaluation performed on 12/10/2025 demonstrated significant clinical deterioration. Laboratory investigations revealed, serum bilirubin of 13.5 mg/dL

HBsAg, anti-HCV, HIV I and II antibodies, anti-HBc total antibody, IgM anti-HAV, HBV DNA, and HCV RNA were negative or undetectable. ANA profile was negative. Alpha-fetoprotein and carcinoembryonic antigen levels were normal, while

CA 19-9 was mildly elevated at 46.70 U/mL. Repeat ultrasonography revealed volume redistribution of the liver with right lobe atrophy, hypertrophy of caudate lobe and segment IV, heterogeneous echotexture with undulated margins, multifocal peripheral IHBRD measuring 2–4 mm, mild splenomegaly, and hypertrophied hepatic artery. Portal vein flow remained hepatopetal.

Chest radiography, computed tomography thorax, pulmonary function tests, and cardiac evaluation including electrocardiography were normal. Bubble contrast echocardiography demonstrated 3–5 bubbles appearing in the left atrium at the sixth cardiac cycle. Pre-transplant liver biopsy revealed established biliary cirrhosis secondary to primary sclerosing cholangitis.

Following multidisciplinary assessment involving hepatology, transplant surgery, anesthesiology, cardiology, nephrology, pulmonology, psychiatry, gynecology, ophthalmology, and otorhinolaryngology teams, the patient was deemed suitable for liver transplantation.

She underwent deceased donor orthotopic whole liver transplantation with hepaticojejunostomy on 02/12/2025. Postoperative recovery was uneventful, and the patient continues to do well on follow-up with stable graft function. (Fig 3a,3b)

Given the family history of ulcerative colitis, patient's father underwent evaluation for PSC, which was negative. Her brother presented with diarrhea and weight loss; colonoscopy demonstrated edematous colonic mucosa with terminal ileal inflammation. Histopathological examination suggested nonspecific findings. Targeted gene sequencing performed to evaluate pathogenic genetic variants of IBD did not reveal any significant abnormalities. Fecal microbiota analysis of patient's brother revealed: Normal Gut Health Index (GHI) Score (0.73), Microbiome Diversity Score (3.76), Butyrate Producing Capacity Score (6.98), Inflammatory Index (1.28) and absent Dysbiosis.

3 DISCUSSION

PSC is a chronic cholangiopathy characterized by multifocal biliary strictures resulting in progressive cholestasis, fibrosis, and cirrhosis. The disease demonstrates strong association with inflammatory bowel disease in nearly 70% of patients. The present case is unusual due to multiple reasons: female gender, absence of inflammatory bowel disease, aggressive progression within months, requirement of liver transplantation shortly after diagnosis and presence of family history of UC.

Table 2: Causes of Secondary Sclerosing Cholangitis

Benign	Malignant
Eosinophilic cholangitis	Ampullary adenocarcinoma
Mast cell cholangitis	Cholangiocarcinoma
Sarcoidosis	Gallbladder cancer
Bile duct injury	Hepatocellular cancer
Mirizzi syndrome	Lymphoma
AIDS cholangiopathy	Metastatic cancer
Recurrent pyogenic cholangitis	Pancreatic adenocarcinoma
Intra-arterial chemotherapy	
Ischemic cholangiopathy	
Portal hypertensive cholangiopathy	
Vasculitis	
Chronic pancreatitis	
IgG4 cholangitis	
ICI toxicity	

The diagnosis of PSC was established based on characteristic cholangiographic findings and compatible histopathology after exclusion of secondary causes of sclerosing cholangitis. (Table 2)

MRCP findings including multifocal strictures, peripheral biliary dilatation, non-visualization of major ducts, lobar atrophy, and compensatory caudate hypertrophy were highly suggestive of advanced PSC.

Liver biopsy demonstrated biliary-pattern injury with periductal fibrosis and absence of overlap features. Nodular regenerative hyperplasia identified on histopathology may reflect associated vascular flow abnormalities secondary to chronic cholangiopathy and portal vascular remodeling.

Rapid progression in PSC has been associated with severe cholestasis, dominant strictures, extensive intrahepatic involvement, elevated bilirubin, and advanced fibrosis at presentation. In the present case, bilirubin increased from 4.8 mg/dL to 13.58 mg/dL within months despite therapy, accompanied by worsening coagulopathy and progression to established biliary cirrhosis.

Although elevated serum IgG was noted during pre-transplant evaluation, autoimmune overlap syndrome was not supported by histopathology or serological profile. IgG4-related sclerosing cholangitis was considered unlikely due to the characteristic cholangiographic pattern, absence of pancreatic involvement, and lack of supportive serology.

Currently, liver transplantation remains the only definitive therapy for advanced PSC. Indications include decompensated cirrhosis, intractable pruritus, recurrent cholangitis, and progressive hepatic dysfunction. Outcomes following transplantation are generally favorable, with reported five-year survival rates exceeding 80%.

The present case emphasizes the importance of early transplant referral in aggressive PSC phenotypes, even in patients without classical risk factors or inflammatory bowel disease.

4 CONCLUSION

Rapidly progressive PSC is an uncommon but clinically important variant of primary sclerosing cholangitis that may progress to end-stage liver disease within months. Early diagnosis through characteristic cholangiographic and histopathological findings, exclusion of secondary etiologies, close monitoring, and timely transplantation are essential to optimize outcomes. This case additionally highlights the occurrence of aggressive PSC in a young female without associated inflammatory bowel disease.

Learning Points

- PSC can demonstrate aggressive progress leading to LT within months.
- In the absence of IBD, PSC may progress rapidly.
- MRCP plays a crucial role in diagnosing and assessing the extent of disease.
- Histopathology is used in excluding overlap syndromes and secondary causes.
- Early referral for LT should be considered in rapidly progressive disease.

STATEMENTS ON COMPLIANCE WITH ETHICAL STANDARDS

Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of informed consent

Written informed consent was obtained from the patient for publication of this case report and associated clinical details.

Author Contributions

All authors contributed to clinical management, data acquisition, manuscript preparation, and final approval of the manuscript.

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