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Case Report ***Volume 1 | Issue 1***

KOS Journal of Clinical, Medical Case Reports and Case Studies
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A Rare Case of Primary Sclerosing Cholangitis in a 20-Year-Old Man

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Received: July 22, 2025; **Accepted:** July 24, 2025; **Published:** July 26, 2025

Citation: Guru YM, et al. (2025) A Rare Case of Primary Sclerosing Cholangitis in a 20-Year-Old Man. *KOS J Clin Med Case Rep Case Stud*. 1(1): 1-5.

1. Abstract

Primary Sclerosing Cholangitis is a rare, long-lasting cholestatic liver disease characterised by advanced inflammation and fibrosis of the intrahepatic and extrahepatic bile ducts. It is infrequent in young adults, making the primary diagnosis and management serious to prevent difficulties. A 20-year-old male presented with icterus, pruritus, fever, and yellowish discoloration of urine. Laboratory investigations exposed deranged liver function tests, including elevated bilirubin and cholestatic enzymes. The patient underwent Magnetic Resonance Cholangiopancreatography, which established characteristic features of PSC such as intrahepatic bile duct beading, multifocal strictures, periductal fibrosis, caudate lobe hypertrophy, and bile duct wall thickening. These results, along with the clinical presentation, established the diagnosis of Primary Sclerosing Cholangitis. The importance of early radiological evaluation, mainly MRCP, in diagnosing PSC, especially in atypical young adult presentations. Early detection facilitates timely involvement, helps monitor disease progression, and reduces the danger of serious difficulties such as cholangiocarcinoma, hepatocellular carcinoma, gallbladder carcinoma, and colorectal carcinoma. Radiologists and clinicians must remain cautious for PSC's imaging features to confirm rapid diagnosis and management.

2. Keywords

Primary Sclerosing Cholangitis, MRCP, Cholestatic Liver Disease, Biliary Strictures, Young Adult, Cholangiocarcinoma, Hepatobiliary Imaging, Caudate Lobe Hypertrophy, Periductal Fibrosis, Radiological Diagnosis

3. Introduction

Primary sclerosing cholangitis is a long-lasting, advanced cholestatic liver disease characterised by inflammation and fibrosis of the intrahepatic and extrahepatic bile ducts, in the end leading to multifocal bile duct strictures and liver cirrhosis. It is a relatively rare disorder with a prevalence of around 6 to 16 per 100,000 individuals in the Western population, though precise epidemiological data from Asia and India are lacking [1,2]. PSC has a strong predeliction for young to middle-aged men, most frequently presenting between the ages of 30 and 50 years [3]. However, occurrence in younger individuals, predominantly in the second decade of life, is unusual and contributes to diagnostic confusion due to the intersection of symptoms with other cholestatic conditions.

The aetiology of PSC remains poorly understood, but the disease is believed to result from a complex interaction of genetic predisposition, immune dysregulation, and environmental factors [4]. Around 70-80% of PSC patients have coexisting inflammatory bowel disease, predominantly ulcerative colitis [5]. This close association suggests a shared

pathogenic mechanism, possibly mediated through abnormal immune responses to gut microbiota or increased intestinal permeability, leading to hepatobiliary inflammation. However, PSC can also occur in the absence of IBD, mainly in paediatric or very young adult populations, as seen in rare case reports [6].

Clinically, PSC is insidious in start and frequently asymptomatic at early stages, with diagnosis frequently made incidentally on the basis of abnormal liver function tests, particularly cholestatic enzyme elevations such as alkaline phosphatase and gamma-glutamyl transferase [7].

Symptomatic patients may present with fatigue, pruritus, jaundice, right upper quadrant abdominal pain, and recurrent episodes of cholangitis. In advanced stages, complications include secondary biliary cirrhosis, portal hypertension, and hepatic failure. A unique feature of PSC is its association with increased danger for malignancies, most particularly cholangiocarcinoma, hepatocellular carcinoma and colorectal cancer in cases coexisting with IBD [8].

The diagnosis of PSC is primarily based on characteristic cholangiography results, typically obtained through magnetic resonance cholangiopancreatography, which establishes multifocal strictures and segmental dilations of the bile ducts, described as a "beaded appearance" [9]. Liver biopsy is generally not required unless small duct PSC is suspected or when overlap with autoimmune hepatitis is considered. Histopathology, when performed, reveals periductal concentric fibrosis, frequently referred to as "onion-skinning", which is pathognomonic but not always present [10].

In spite of advances in diagnostic imaging and supportive care, there is presently no curative medical therapy for PSC. Ursodeoxycholic acid, once considered a mainstay of treatment, has shown limited efficacy in altering disease progression or preventing complications when used at high doses [11]. Liver transplantation remains the definitive treatment for patients who develop end-stage liver disease or cholangiocarcinoma, though PSC is one of the leading signs for liver transplantation in young adults in many centres worldwide [12].

The occurrence of PSC in a 20-year-old male, as in the present case, is clinically significant due to its rarity in this age group and the implications it carries for long-term management. Early-onset PSC may represent a distinct clinical subset, with some studies suggesting more aggressive disease progression and increased danger of problems compared to adult-onset PSC [4]. Moreover, the psychosocial and quality-of-life impact in young patients is considerable, given the chronicity of the disease, the need for lasting surveillance, and the possibility of liver transplantation at an early age.

The recognition of PSC in young adults requires a high index of suspicion, especially when confronted with unexplained cholestasis or recurrent episodes of ascending cholangitis. It also demands a multidisciplinary method involving hepatologists, gastroenterologists, radiologists, and transplant surgeons for optimal management. Continued investigation into the genetic and immunological mechanisms fundamental to PSC is essential, particularly in younger populations, to develop targeted therapies and improve patient consequences.

This case report presents the clinical presentation, diagnostic contests, and management deliberations of a rare instance of PSC in a 20-year-old male, emphasising the importance of initial recognition and complete care.

4. Case presentation

A 20-year-old male presented to the Department of Medicine at Sri Devaraj Urs Medical College, Kolar, with complaints of icterus, pruritus, fever, and yellowish discoloration of urine. Clinical examination exposed signs suggestive of hepatobiliary pathology, prompting additional laboratory investigations. His liver function tests were altered, showing raised bilirubin levels and raised cholestatic enzymes, including alkaline phosphatase and gamma-glutamyl transferase. Considering the clinical and biochemical results, the patient was referred to the Department of Radiodiagnosis for unconventional imaging to assess the underlying cause of cholestasis. An MRCP was performed using a 1.5 Tesla Siemens® Magnetom Avanto MRI scanner equipped with an 18-channel coil system. The imaging protocol included T2-weighted sequences to assess the liver parenchyma and bile ducts, MRCP sequences for detailed assessment of intrahepatic and extrahepatic biliary tree, and contrast-enhanced sequences when required to assess ductal wall improvement and parenchymal differences.

Radiological interpretation was absorbed on identifying characteristic features of Primary Sclerosing Cholangitis, such as biliary duct irregularities, multifocal strictures, irregular ductal dilatation, and liver parenchymal fibrosis.

The MRCP and MRI results in this patient exposed classic imaging features of Primary Sclerosing Cholangitis. There was irregular beading of the intrahepatic bile ducts, caused by irregular strictures and dilatations, and periductal fibrosis, which appeared hyperintense on T2-weighted images, especially in the peripheral regions of the liver. Confluent fibrosis was renowned in the central parts of the liver, which appeared hypodense on CT and hyperintense on T2-weighted MRI sequences.

Figure 1 demonstarting an MRI Upper Abdomen T1-weighted image showing dilated intrahepatic biliary radicals. The black arrow indicates dilated central intrahepatic bile ducts, while the white arrow points to dilated peripheral intrahepatic bile ducts.

Figure 2 shows MRCP images of the intrahepatic biliary radicals and both the right and left hepatic ducts. The bile ducts appear mildly dilated and irregular, with changes more pronounced in the left lobe of the liver. The left hepatic duct measures approximately 3.5 mm, and the right hepatic duct measures around 2.4 mm, which indicates biliary ductal dilatation and irregularity consistent with primary sclerosing cholangitis.

Figure 1: MRI Upper abdomen TIW image shows dilated central (black arrow) and peripheral (white arrow) intrahepatic biliary radicals.

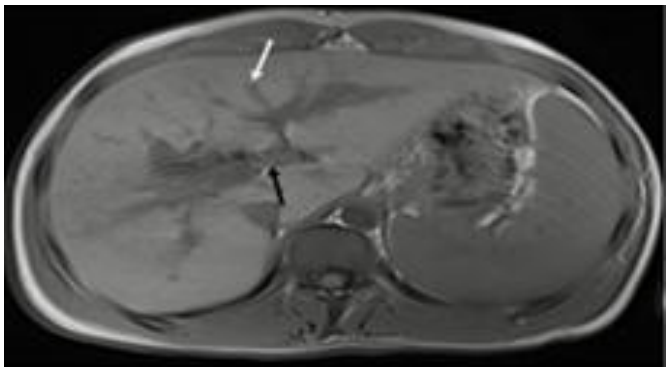
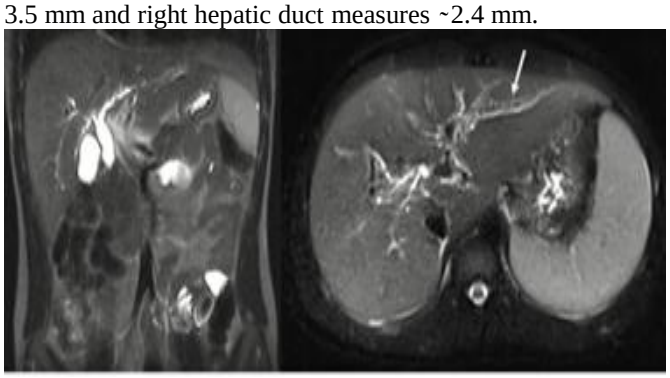


Figure 2: Intrahepatic biliary radicals, right & left hepatic ducts: Appears mildly dilated and irregular more prominent in left lobe of liver (white arrow). Left hepatic duct measures ~ 3.5 mm and right hepatic duct measures ~2.4 mm.



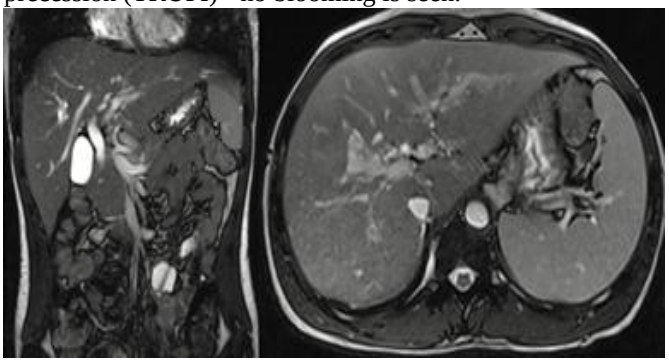
Figures 1 and **Figure 2**: MRI Upper abdomen T1W image shows dilated central and Intrahepatic biliary radicals, right and left hepatic ducts

There was thickening and hyperenhancement of the bile duct walls, signifying active inflammation. In addition, hypertrophy of the caudated lobe was observed, a result frequently seen in long-lasting stages of PSC. Another important observation was the visualisation of a greater than expected number of peripheral intrahepatic ducts, representing peripheral ductal strictures. These imaging results correlated well with the clinical presentation and biochemical abnormalities, confirming the diagnosis of Primary Sclerosing Cholangitis.

Figure 3 shows True Fast Imaging with Steady-State Free Precession sequences. These images demonstrate clear visualization of the biliary tree without any signal loss or blooming artifacts. The ducts are well delineated, allowing for accurate assessment of biliary structures. No extrinsic compression or obstructing lesion is seen, supporting a primary ductal abnormality rather than secondary causes.

Figure 4 shows a Maximum Intensity Projection MRCP image, highlighting the mildly dilated and irregular central and peripheral intrahepatic biliary radicals. Multiple short segment strictures are alternating with ductal dilatations, resulting in the characteristic “beaded appearance”. This imaging feature is a classic mark of Primary Sclerosing Cholangitis and reflects chronic inflammatory changes with progressive fibrosis and segmental narrowing of the bile ducts.

Figures 3: True Fast Imaging with steady-state-free precession (TRUFI) - no blooming is seen.



Figures 4: MIP Image – Mildly dilated and irregular central and peripheral intrahepatic biliary radicles with multiple alternating short segment strictures, giving beaded appearance (white arrow).

Figures 3 and **Figure 4**: True fast imaging with steady-state-free precession and MIP image.

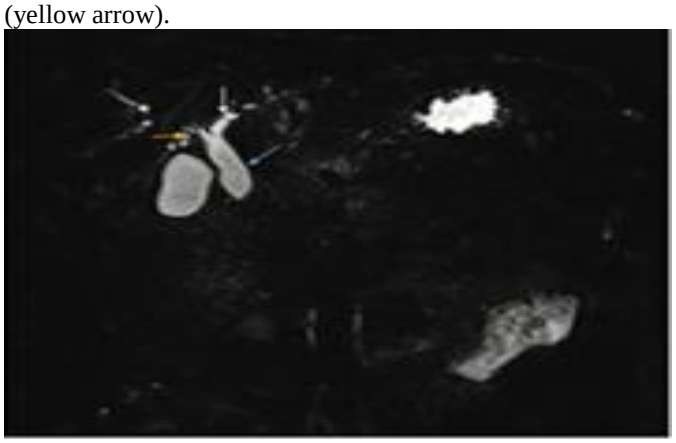
Primary Sclerosing Cholangitis is a rare but important cause of continuing cholestatic liver disease, particularly unusual in young adults. This case underscores the importance of primary imaging with MRCP for prompt diagnosis and staging of PSC. Early results are important to initiate appropriate management, monitor disease development, and prevent complications such as cholangiocarcinoma, hepatocellular carcinoma, gallbladder carcinoma, and colorectal carcinoma. Radiological assessment plays a serious role in the management of PSC, and awareness of its imaging features can suggestively impact patient results by facilitating early therapeutic involvements and improving quality of life.

Figure 5 demonstrates a T2-weighted MRCP image showing dilated common hepatic duct and common bile duct. The white arrow points to the CHD, which measures approximately 11 mm, and the blue arrow indicates the CBD, measuring about 12.5 mm, both of which are dilated beyond normal reference ranges. The cystic duct appears normal, as indicated by the yellow arrow. Prominently, the ductal walls are smooth and regular, and no intraductal mass is seen, ruling out obstructive lesions such as cholangiocarcinoma or choledocholithiasis.

Figure 6 presents True Fast Imaging with Steady-State Free Precession sequences, which provide high-resolution images of the biliary system without blooming artifacts. The bile ducts appear clearly outlined, further confirming the presence of dilated intrahepatic and extrahepatic ducts without any evidence of obstructive masses.

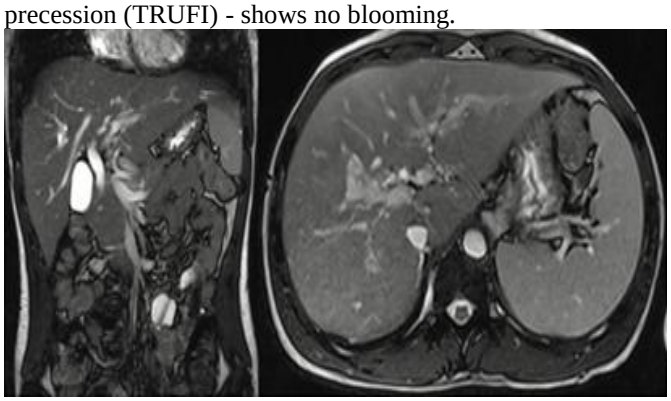
Figures 5 and **Figure 6**: T2W and TRUFI.

Figures 5: T2 W - Common hepatic duct (CHD) (white arrow) & common bile duct (CBD) (blue arrow) appears dilated CHD measures~ 12.5 mm. Cystic duct: Normal (yellow arrow).



Duct wall appears smooth and regular.
No intraductal mass noted.

Figures 6: True Fast Imaging with steady-state-free precession (TRUFI) - shows no blooming.



5. Discussion

Primary sclerosing cholangitis is an exclusive cholestatic liver disease that presents diagnostic and management competitions, particularly when it occurs in young adults. Its clinical, biochemical, and radiological features can overlap with other hepatobiliary conditions, including primary biliary cholangitis, autoimmune hepatitis, IgG4-related sclerosing cholangitis, secondary sclerosing cholangitis, and Caroli's disease. Differentiating PSC from these conditions is serious as the prognosis, problems, and therapeutic methods differ suggestively.

6. Primary Sclerosing Cholangitis vs. Primary Biliary Cholangitis

Even though PSC and PBC are both chronic cholestatic liver diseases, they differ in pathogenesis, demographics, and clinical course. PBC predominantly affects middle-aged women, whereas PSC is more communal in men, mainly in the third to fifth decades of life [13]. However, PSC presenting in a 20-year-old male, as in the present case, is relatively rare. PBC is characterised by autoimmune-mediated destruction of small intrahepatic bile ducts, whereas PSC involves both intrahepatic and extrahepatic bile ducts, leading to multifocal strictures [14].

Serologically, PBC is connected with anti-mitochondrial antibodies in over 90% of cases, while PSC lacks disease-specific autoantibodies, although perinuclear anti-neutrophil cytoplasmic antibodies may be present in up to 80% of PSC patients [15]. MRCP is the preferred imaging modality for PSC, showing the classical "beaded" bile ducts. In difference, imaging in PBC is often normal or non-specific, as the disease primarily affects small ducts not visualised on cholangiography [16]. In addition, PBC has approved medical therapy with ursodeoxycholic acid, which can slow disease development, whereas no disease-modifying treatment is available for PSC [17].

7. PSC vs. Autoimmune Hepatitis

Autoimmune hepatitis is another differential diagnosis due to underlying clinical structures, such as elevated liver enzymes and possible autoimmune markers. AIH typically presents with a hepatocellular design of liver enzyme elevation, whereas PSC shows a cholestatic pattern with predominant alkaline phosphatase elevation [6]. Liver biopsy in AIH exposes interface hepatitis with plasma cell infiltration, in difference to the periductal fibrosis seen in PSC [18].

A subset of patients may develop an overlap syndrome between PSC and AIH, mainly in paediatric and young adult populations. This intersection can confuse diagnosis and management, often requiring immunosuppressive therapy together with surveillance for PSC-related problems [19]. Recognising this overlap is essential in young patients presenting with features of both conditions.

8. PSC vs. IgG4-Related Sclerosing Cholangitis

IgG4-related sclerosing cholangitis must also be considered, especially when bile duct strictures are noted on imaging. IgG4-SC typically affects older males and is often associated with autoimmune pancreatitis and elevated serum IgG4 levels [20]. Different PSC, IgG4-SC responds dramatically to corticosteroid therapy, making initial differentiation dangerous. Biopsy results in IgG4-SC reveal dense lymphoplasmacytic infiltrates rich in IgG4-positive plasma cells, obliterative phlebitis, and storiform fibrosis, features not seen in PSC [21].

9. PSC vs. Secondary Sclerosing Cholangitis

Secondary sclerosing cholangitis refers to bile duct injury caused by identifiable aetiologies such as biliary tract surgery, ischemic injury, recurrent pancreatitis, or infections like AIDS-related cholangitis. SSC can mimic PSC radiologically, but is usually connected with a clear precipitating factor [22]. In difference, PSC is idiopathic and advanced in spite of no obvious cause. SSC management involves treating the underlying cause, whereas PSC has no specific curative treatment other than liver transplantation.

10. PSC vs. Caroli's Disease

Caroli's disease is a congenital disorder characterised by segmental dilation of large intrahepatic bile ducts. Similar to PSC, it can present with recurrent cholangitis and jaundice in young individuals. However, imaging in Caroli's disease reveals saccular or cystic dilation rather than the diffuse multifocal strictures and "beading" typical of PSC [23]. Caroli's disease may also be associated with congenital hepatic fibrosis and renal cystic diseases, additional assisting in differentiation.

11. Clinical Implications

In the present case, the diagnosis of PSC in a 20-year-old

male emphasises the importance of distinguishing PSC from these mimicking conditions. While PBC and AIH are more common in females, IgG4-SC and SSC typically have secondary causes or distinct serological profiles. The potential for PSC to progress to cirrhosis, portal hypertension, and cholangiocarcinoma mandates vigilant lasting follow-up.

Assuming the lack of disease-modifying therapies, initial diagnosis focuses on symptom management, surveillance for malignancy, and preparation for liver transplantation when needed. The young age at presentation may imply a longer disease development with cumulative problems, and the need for more investigation into disease-modifying treatments for PSC, especially in early-onset cases.

12. Conclusion

Primary Sclerosing Cholangitis is a continuing, advanced cholestatic liver disease noticeable by inflammation and fibrosis of the bile ducts. Its occurrence in young adults is rare, making this case important. Early diagnosis through MRCP is vital for accurate staging, monitoring disease development, and detecting problems such as cholangiocarcinoma, hepatocellular carcinoma, gallbladder carcinoma, and colorectal carcinoma. Appropriate imaging facilitates the opening of supportive treatment to improve quality of life and delay difficulties. In this case, the important role of radiological evaluation in PSC emphasises the need for increased clinical and radiological awareness for initial detection and management.

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