SAS Journal of Medicine
Abbreviated Key Title: SAS J Med
ISSN 2454-5112
Journal homepage: https://saspublishers.com

OPEN ACCESS

Gastroenterology

Small Duct Primary Sclerosing Cholangitis Ducts: Review and a Case Report

Asmaa N'khaili[1*], Riad Semlali[1], Fatima Ezzahra Chakor[1], Meryem Aouroud[1], Adil Ait Errami[1], Sofia Oubaha[2], Zouhour. Samlani[1], Khadija Krati[1]

[1]Department of Gastroenterology CHU Mohammed VI Marrakech, Morocco
[2]Laboratory of Physiology, Faculty of medicine and pharmacy of Marrakech, Morocco

DOI: 10.36347/sasjm.2021.v07i12.013 | **Received:** 19.11.2021 | **Accepted:** 21.12.2021 | **Published:** 30.12.2021

***Corresponding author:** Asmaa N'khaili

Abstract **Case Report**

Primary sclerosing cholangitis is a rare disease characterized by inflammation, fibrosis, and narrowing of the intrahepatic or extrahepatic bile ducts .Small bile duct PSC(SDPSC) is a subtype of PSC, the prevalence of which is estimated to be around 10% of all CSPs. We report the case of a patient 36 years old, admitted for generalized mucocutaneous jaundice and cholestasis, the etiology was not determined according to the assessments carried out, The MR cholangiography did not reveal any abnormalities explaining the cholestasis, the diagnosis of CSP was made according to the liver histology.

Keywords: Small bile ducts, inflammation, fibrosis, cholestasis.

1. INTRODUCTION

Primary sclerosing cholangitis is a rare disease, most commonly affecting males at a median age of 40 years, characterized by stenosing fibro-inflammatory lesions of the bile ducts. Liver biopsy is essential for the diagnosis of PSC when cholangio-MRI fails to diagnose, this is the case of the small duct primary sclerosing cholangitis, The diagnostic criteria are based on the biochemical characteristics of chronic cholestasis of unknown etiology, normal cholangiography, liver histology compatible with PSC [1].

We report the case of small duct primary sclerosing cholangitis diagnosed diagnosed through liver histology.

2. PATIENT AND OBSERVATION

A 36 year old male patient, reports a generalized cutaneous-mucous jaundice and cholestasis made up of dark urine, discolored stools, evolving for 4 months, associated with pruritus, a deterioration in general health . He had a normal physical examination, except the existence of frank mucocutaneous jaundice. Biologically, total bilirubin was 155.1iu / l predominantly direct at 95iu / l ALAT at 95uu / l (1.9N), ASAT at 69u / l (1.6N), PT at 65% PAL at 424 (3.2N) and GGT at 33ui / l (N) Laboratory results were negative for hepatitis A, B and C, Epstein-Barr virus (EBV), cytomegalovirus (CMV). The IgG 4 dosage was normal at 0.210 g / l. the immunological assessment carried out showed that the antinuclear antibodies, anti-mitochondrial antibodies, anti-sp100 AC; anti-gp210 AC, anti-smooth muscle, anti-hepatic-renal microsomal AC, AC anticytosol, AC anti SLA were negative. cholangiogram was without abnormalities apart from an angioma located at the level of segment II in subcapsular in clear T2 hypersignal, enhanced in the periphery and in lump in the arterial phase, homogenizing in the portal phase, measuring 10* 8mm, without dilation of the intrahepatic and extrahepatic bile ducts. The hepatic histology showed an obliterating cholangitis associated with a peribilar portal inflammation, a discreetly atrophic aspect of the bile ducts, an irregularity of the canalic caliber, with an important cholestasis associated with a ductopeniahe. The diagnosis of SDPSC was made. The screening colonoscopy performed was without abnormalities. Patient was put on ursodeoxicolic acid at a dose of 20 mg / kg / day.

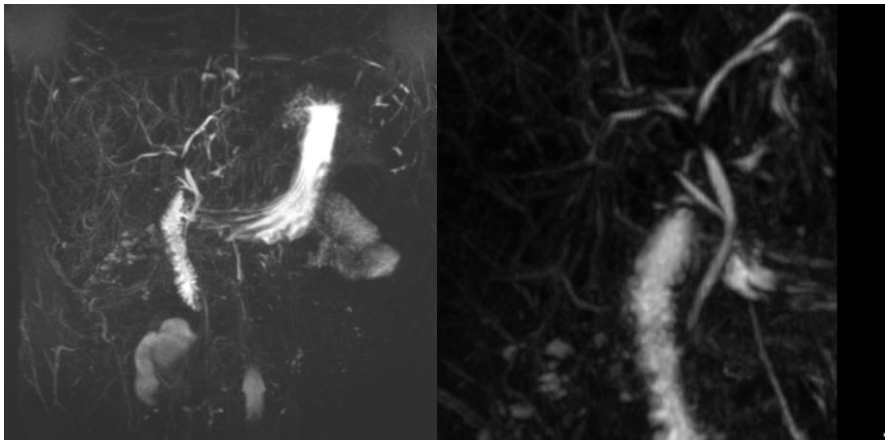

Figure 1: T2 sequence of the 3D MR cholangiography showing a normal and undilated aspect of the intra and extrahepatic bile ducts

3. DISCUSSION

Primary sclerosing cholangitis (PSC) is a chronic, progressive, cholestatic liver disease of unknown etiology. According to EASL recommendations, the diagnosis of PSC can be made in the presence of biological cholestasis associated with abnormalities typical of cholangiography and in the absence of a cause of secondary sclerosing cholangitis [2].

We can thus distinguish 3 subtypes of PSC: the classic form which affects the small and large bile ducts, the form which affects only the small bile ducts and the form associated with autoimmune hepatitis: affects small and large bile ducts [3].

MR Cholangiography is an essential step in terms of diagnosis of PSC, it makes it possible to visualize the radiological lesions typical of the bile ducts in terms of PSC, MR Cholangiographiy could replace the Endoscopic retrograde cholangiopancreatography ,due to its invasive nature [4], it makes it possible to show stenoses, most often multiple, alternating with usually moderate dilations of the intra and extrahepatic bile ducts. The aspects observed are in fact very variable, including diverticular or cystic aspects, best visualized using the 3D technique [5]. However, its normality does not exclude the diagnosis of sclerosing cholangitis. In 1985, the question of the existence of small bile duct involvement was first raised by Ludwig [6]. Studies carried out in 1991 led to the emergence of the term PSC of the small bile ducts in the presence of histology suggestive of PSC and in the absence of radiological anomalies of the bile ducts [7].

Liver biopsy is therefore essential for the diagnosis of PSC, when MR cholangiography does not reveal any abnormalities or for the diagnosis of overlap syndrome [8]. The most specific sign found on histology is "onion skin", which is described as fibrous obliteration and concentric replacement of connective tissue in the small bile ducts [9], the differential diagnosis can be made with other causes of macroscopically normal bile duct cholestasis and in particular PBC, sarcoidosis, drug-induced cholangitis and MDR3 deficits [10].

The prevalence of small canals PSC is estimated to be around 10% of all PSCs. Secondary involvement of the large bile ducts seems relatively rare since it is reported only in around 15% of cases after an average follow-up of around ten years [11]. The diagnosis of small duct PSC is still difficult, with regard to the presence of associated IBD, generally this is not a mandatory criterion for small duct PSC according to some studies [10].

The etiology of primary sclerosing cholangitis of small ducts is unknown. However, when considering small duct PSC as a subtype of PSC, dietary exposure, infection, toxins, and autoimmunity may be the primary contributing factors [12].

In case of no change to the form of large bile ducts, the risk of cholangiocarcinoma is low, no case has been described, which makes the prognosis better than the classic form of PSC [13].

With the intention of detecting cholangiocarcinoma earlier, the annual MR cholangigraphy is systematic in terms of classic form of PSC [5], The rate of monitoring by MR cholangiograohy may be reduced, in case of SDPSC while performing it at 1 year and then every 3 years because this form has a better prognosis with a minimal risk of cholangiocarcinoma and only rarely progresses to a classic form of CSP of the major ducts [13].

There is not enough data regarding the treatment of SDPSC, but according to the study by Charatcharoenwitthaya et al. concluded that treatment with AUDC might not delay disease progression but might improve biological cholestasis [14]. However, in patients with small duct PSC who present with terminal

liver disease, have a longer transplant-free survival than large duct PSC [15].

4. CONCLUSION

Primary small duct sclerosing cholangitis is a rare disease with uncertain prevalence due to limited studies. The scarcity of PSC of small bile ducts poses diagnostic problems. . Thus, it is necessary to eliminate the other etiologies which can generate chronic cholestasis by choosing the right laboratory tests and imagery. After ruling out known causes of chronic cholestasis, cholangiography and liver biopsy can be used to distinguish PSC from small and large bile ducts.

REFERENCES

1. Amini, A., & Nagalli, S. (2020). Pericholangitis. Stat Pearls Publishing, Treasure Island, Europe PMC 28 Jan 2020.
2. European Association For The Study Of The Liver. (2009). EASL Clinical Practice Guidelines: management of cholestatic liver diseases. *Journal of hepatology*, *51*(2), 237-267.
3. Prashanth, R., & Hrishikesh, S. (2021). Primary sclerosing cholangitis, Stat Pearls, Treasure Island, Stat Pearls Publishing.
4. European Society of Gastrointestinal E. (2017). European Association for the Study of the Liver. Electronic address eee, European Association for the Study of the L. Role of endoscopy in primary sclerosing cholangitis: European Society of Gastrointestinal Endoscopy (ESGE) and European Association for the Study of the Liver (EASL) Clinical Guideline. *J Hepatol*, 66, 1265-1281.
5. Schramm, C., Eaton, J., Ringe, K. I., Venkatesh, S., Yamamura, J., & MRI working group of the IPSCSG. (2017). Recommendations on the use of magnetic resonance imaging in PSC-A position statement from the International PSC Study Group. *Hepatology*, *66*(5), 1675-1688.
6. Ludwig, J. (1991, February). Small-duct primary sclerosing cholangitis. In *Seminars in liver disease* (Vol. 11, No. 01, pp. 11-17). © 1991 by Thieme Medical Publishers, Inc..
7. Angulo, P., & Lindor, K. D. (1999). Primary sclerosing cholangitis. *Hepatology*, *30*(1), 325-332.
8. Dyson, J. K., Beuers, U., Jones, D. E., Lohse, A. W., & Hudson, M. (2018). Primary sclerosing cholangitis. *The Lancet*, *391*(10139), 2547-2559.
9. Burak, K. W., Angulo, P., & Lindor, K. D. (2003). Is there a role for liver biopsy in primary sclerosing cholangitis?. *The American journal of gastroenterology*, *98*(5), 1155-1158.
10. European Association For The Study Of The Liver. (2009). EASL Clinical Practice Guidelines: management of cholestatic liver diseases. *Journal of hepatology*, *51*(2), 237-267.
11. Björnsson, E., Boberg, K. M., Cullen, S., Fleming, K., Clausen, O. P., Fausa, O., ... & Chapman, R. W. (2002). Patients with small duct primary sclerosing cholangitis have a favourable long term prognosis. *Gut*, *51*(5), 731-735.
12. LaRusso, N. F., Shneider, B. L., Black, D., Gores, G. J., James, S. P., Doo, E., & Hoofnagle, J. H. (2006). Primary sclerosing cholangitis: summary of a workshop. *Hepatology*, *44*(3), 746-764.
13. Weismüller, T. J., Trivedi, P. J., Bergquist, A., Imam, M., Lenzen, H., Ponsioen, C. Y., ... & International PSC Study Group. (2017). Patient age, sex, and inflammatory bowel disease phenotype associate with course of primary sclerosing cholangitis. *Gastroenterology*, *152*(8), 1975-1984.
14. Charatcharoenwitthaya, P., Angulo, P., Enders, F. B., & Lindor, K. D. (2008). Impact of inflammatory bowel disease and ursodeoxycholic acid therapy on small-duct primary sclerosing cholangitis. *Hepatology*, *47*(1), 133-142.
15. Björnsson, E., Olsson, R., Bergquist, A., Lindgren, S., Braden, B., Chapman, R. W., ... & Angulo, P. (2008). The natural history of small-duct primary sclerosing cholangitis. *Gastroenterology*, *134*(4), 975-980.