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Research Article

Seronegative Primary Biliary Cholangitis: Epidemiological, Clinical, Therapeutic and Progressive Features

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Abstracts: ***Introduction:*** Seronegative PBC (specific AMA antibodies are negative) is a particular clinical form of PBC.***The aim*** of this study was to determine the clinical-biological and evolutionary features of seronegative PBCs. ***Methods:*** The medical data of 42 PBC patients were evaluated and the AMA positive and negative groups were compared. ***Results:*** 42 cases of PBC were collected. These were 41 women and 1 man. The mean age at diagnosis was 46 years (25-70 years). AMA Abs were most often positive (n = 34). Other antibodies were found in 22 patients. In our series, 9.75% of cases had seronegative PBC. By comparing the 2 groups of PBC with or without AMA Abs, depending on age, sex, degree of fibrosis and response to AUDC, no statistically significant difference was noted. ***Conclusion:*** In our series, the prevalence of seronegative PBC was 9.75%. Its clinical-biological and evolutionary profile under treatment does not seem to differ from that of seropositive PBC.

Keywords: primary biliary cholangitis, AMA seronegative, AMA seropositive.

INTRODUCTION

Primary biliary cholangitis (PBC) is a chronic cholestatic disease of autoimmune origin whose diagnosis is based on the combination of biochemical signs of cholestasis, immunological signs (anti-mitochondria antibodies (AMA) or anti gp-210 and anti sp-100 if the AMA Abs are negative) and / or characteristic histological signs of PBC. Seronegative PBC (specific antibodies are negative) is a particular clinical form.

The aim of this study was to determine the clinical-biological and evolutionary features of seronegative PBC.

PATIENTS AND METHODS

This is a retrospective study of 42 patients with primary biliary cholangitis in the hepato-gastroenterology department over a period of 8 years, extending between January 2012 and December 2019.

RESULTS

42 cases of PBC were collected. These were 41 women and 1 man. The mean age at diagnosis was 46 years (25-70 years). The main clinical signs were: asthenia (n = 32), jaundice (n = 28), pruritus (n = 26) and hepatomegaly (n = 18). 6 patients were asymptomatic at the time of diagnosis. Biology found cholestasis syndrome (n = 41) and cytolysis (n = 24). AMA Abs were most often positive (n = 34). Other antibodies were found in 22 patients. In our series, 9.75% of cases had seronegative PBC.

Liver biopsy (n = 20) found advanced fibrosis in 26% of cases, destructive lymphocytic cholangitis in 90% of biopsies and interface hepatitis in 35% of biopsies performed.

All patients were put on ursodeoxycholic acid. The course was marked by a complete (n = 22) or incomplete (n = 17) response.

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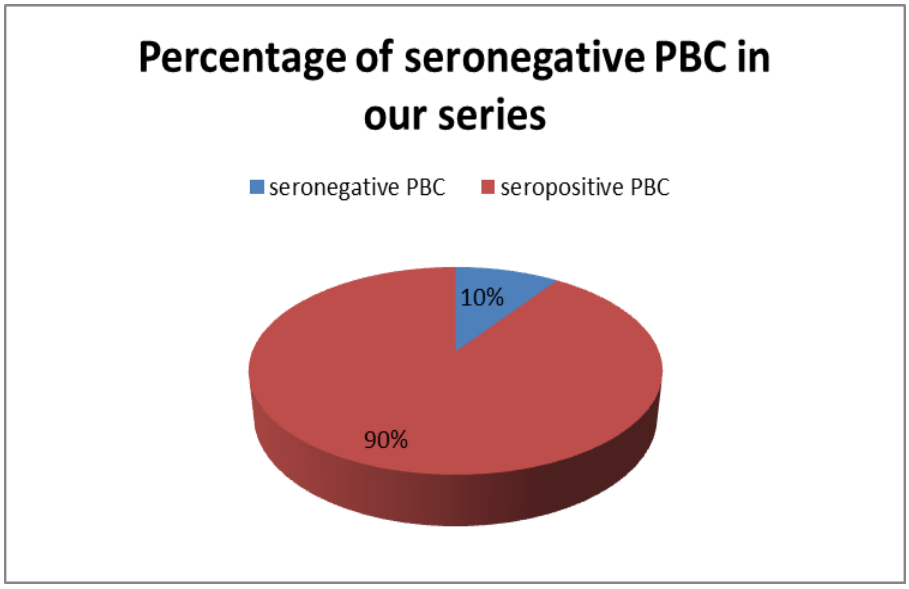

Figure 1: percentage of seronegative PBC in our series

By comparing the 2 groups of PBC with or without AMA Abs, depending on age, sexe, degree of fibrosis and response to AUDC, no statistically significant difference was noted.

DISCUSSION

Most patients with PBC test positive for AMA, but in 10% to 20% of cases it can be absent. AMA-seronegative patients are often positive for antinuclear antibodies (ANAs), including the PBC-specific ANAs sp100 and gp210, as well as recently described anti-kelch-like 12 and anti-hexokinase 1 antibodies (Norman, G. L. *et al.*, 2015).

Liver biopsy is necessary in case of negative anti-mitochondrial antibodies, in the absence of other etiology of cholestasis. It is useful to specify the activity and stage of the disease and to eliminate differential diagnosis such as sarcoidosis, primary sclerosing cholangitis, autoimmune hepatitis, lymphomas, drugs ... (Louie, J. S. *et al.*, 2020).

Histology in AMA-seronegative should demonstrate identical histological findings to AMA-positive disease (Louie, J. S. *et al.*, 2020).

Epidemiological, clinical and evolutionary profile of AMA-seronegative patients is similar to AMA-seropositive PBC as reported by J.H. Ben *et al.*, who compared 2 groups of PBC with or without AMA, depending on age, gender, degree of fibrosis and response to AUDC and noted no statistically significant difference (Ben, J. H. *et al.*, 2016).

M. Ayari *et al.*, reported the same results; No significant difference was found between the groups with or without AMA for demographic data, response to treatment or globe score (Ayari, M. *et al.*, 2019).

CONCLUSION

In our series, the prevalence of seronegative PBC was 9.75%. Its clinical-biological and evolutionary profile under treatment does not seem to differ from that of seropositive PBC.

REFERENCES

1. Ayari, M., Ayadi, S., Mabrouk, E. B. H., Zaaimi, Y., Yosra, S., Mouelhi, L., & Dabbeche, R. (2019). La cholangite biliaire primitive séronégative: prévalence, particularités cliniques, thérapeutiques et pronostiques. *La Revue de Médecine Interne*, *40*, A170.
2. Ben, J. H., Gharbi, O., Kchir, H., Maamouri, N., Chaabouni, H., & Ben, M. N. (2016). La cirrhose biliaire primitive séronégative: particularités épidémiologiques, cliniques, thérapeutiques et évolutives. *La Revue de Médecine Interne*, *37*, A145.
3. Louie, J. S., Grandhe, S., Matsukuma, K., & Bowlus, C. L. (2020). Primary Biliary Cholangitis: A Brief Overview. *Clinical Liver Disease*, *15*(3), 100-104. Published 2020 Apr 4. doi:10.1002/cld.867
4. Norman, G. L., Yang, C. Y., Ostendorff, H. P., Shums, Z., Lim, M. J., Wang, J., ... & Rothschild, K. J. (2015). Anti-kelch-like 12 and anti-hexokinase 1: novel autoantibodies in primary biliary cirrhosis. *Liver International*, *35*(2), 642-651.