

Autoimmune Diseases Associated with Primary Biliary Cirrhosis: A Retrospective Study of 51 Patients

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Abstract : Introduction: Primary biliary cirrhosis (PBC) is a cholestatic cholangitis of unknown etiology. It is frequently associated with autoimmune diseases, which explains their systematic screening. The aim of our study was to determine the prevalence and the type of autoimmune disorders associated with PBC and to assess their impact on the prognosis of the disease. Material and methods: It is a retrospective study over a period of 16 years (2000-2015) including all patients followed for PBC. In all these patients we have systematically researched: dysthyroidism (thyroid balance, antithyroid autoantibodies), type 1 diabetes, dry syndrome (ophthalmologic examination, Schirmer test and lip biopsy in case of Presence of suggestive clinical signs), celiac disease (celiac disease serology and duodenal biopsies) and dermatological involvement (clinical examination). Results: Fifty-one patients (50 women and one men) followed for PBC were collected. The Mean age was 54 years (37-77 years). Among these patients, 30 patients (58.8%) had at least one autoimmune disease associated with PBC. The discovery of these autoimmune diseases preceded the diagnosis of PBC in 8 cases (26.6%) and was concomitant, through systematic screening, in the remaining cases. Autoimmune hepatitis was found in 12 patients (40%), defining thus an overlap syndrome. Other diseases were Hashimoto's thyroiditis (n = 10), dry syndrome (n = 7), Gougerot Sjogren syndrome (n=6), celiac disease (n = 3), insulin-dependent diabetes (n = 1), scleroderma (n = 1), rheumatoid arthritis (n = 1), Biermer Anemia (n=1) and Systemic erythematosus lupus (n=1). The two groups of patients with PBC with or without associated autoimmune disorders were comparable for bilirubin levels, Child-Pugh score, and response to treatment. Conclusion: In our series, the prevalence of autoimmune diseases in PBC was 58.8%. These diseases were dominated by autoimmune hepatitis and Hashimoto's thyroiditis. Even if their association does not seem to alter the prognosis, screening should be systematic in order to institute an early and adequate management.

Keywords : autoimmune diseases, autoimmune hepatitis, primary biliary cirrhosis, prognosis

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