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Case Report

Case Report: Reynolds Syndrome

Introduction

Reynolds et al. described in 1971, 6 patients with classical primary biliary cholangitis, who presented concomitantly with varying degrees of scleroderma. Since then this association has been called Reynolds Syndrome.

This disease is more common in women and it presents as a systemic scleroderma localized type CREST (calcinosis, Raynaud's phenomenon, alterations in esophageal motility, sclerodactyly and telangiectasia). To establish this diagnosis is not necessary to gather all the elements, only two can diagnose an incomplete CREST [1].

The liver symptoms are almost unnoticed by patients and, this is the reason why the medical consultation is delayed [2]

Due to the fact that it is a rare and underdiagnosed disease, we present the case of a female patient who consulted for itch and lesions in the skin compatible with scleroderma, and subsequently, during the study, hepatic alterations were found.

Case Report

We present a 45-year-old woman patient, born and residing in Quito, housewife, with no relevant personal or family history, who came to our clinic due to severe pruritus episodes of several months of evolution. She was treated by several dermatologists with antihistamines without improvement. Physical examination revealed hyperpigmentation predominant in face (Figure 1), sclerosis predominantly on the back of both hands (Figure 2) and hypo and hyperpigmented lesions with the appearance of salt and pepper. In the rest of the skin there is generalized xerosis. In addition, slightly icteric scleras and ragades are evident in the perioral region (Figure 1).

The laboratory exams showed the following results: transaminases TGO: 54.5 U/L, TGP: 103.6 U/L, GGT: 502U/L, Direct Bilirrubin 0.73 mg/dl, Indirect Bilirrubin: 0.10 mg/dl, Total Bilirrubin: 0.83 mg/dl.

Due to the alteration of the liver function, we request tests hepatitis A, B and C serological tests, which were negative, we also requested a hepatic ultrasound reports: Liver of normal size and shape, increased echogenicity due to increased fat deposit without focal lesions (Figure 2).

Facing a patient with cholangitis without a history of chronic ingestion of alcohol or medication and without evidence of viral hepatitis, we suspected a picture of immune origin, so we requested anti-antimitochondrial bodies that were positive. Immunological analysis also found ANA and anti-centromere antibodies Ro positive.

Discussion

Primary biliary cholangitis (PBC) is a chronic inflammatory autoimmune cholestatic liver disease, which when untreated will culminate in end-stage biliary cirrhosis [3].

Figure 1:

Figure 2:

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The frequency of association between systemic sclerosis and primary biliary cholangitis is 5-10% and usually the first manifests as a CREST syndrome [4, 5]. In limited systemic sclerosis, 8% of antimitochondrial antibodies and 9-29% anti-Ro antibodies have been reported [6].

Primary biliary cholangitis begins insidiously, frequent pruritus accompanied by fatigue, or that is accidentally discovered when performing a liver profile that evidences high concentrations of alkaline phosphatase plus the presence of antimitochondrial antibodies [7]. In the case of our patient she only showed incoercible pruritus. It is also important to emphasize that our case is compatible with CREST syndrome, which was considered incomplete because it didn't present calcinosis. The respective analysis found ANA and anticentrome antibodies positive.

Survival of patients with primary biliary cholangitis without treatment is about 10 years. It seems that he would have a more benign course when he is associated with scleroderma [8].

The treatment of choice is ursodeoxycholic acid. Other drugs used are immunosuppressants and antifibrotics [9].

The prognosis of the disease depends on the evolution of primary biliary cholangitis. Pharmacologic approaches in practice, to reduce the impact of the progressive nature of disease, currently include licensed therapies (ursodeoxycholic acid and obeticholic acid) and off-label therapies such asfibric acid derivatives and budesonide [5]. In the case of our patient she clinically improved her pruritus, the hepatic panel and icteric scleras.

Conclusions

Patients with systemic autoimmune diseases may present evidence of concomitant liver disease [7]

Hepatic impairment in these diseases has been well documented, but is generally rare. The severity is variable and may be manifested only with a mild asymptomatic alteration of the transaminases or it might debut with cirrhosis with severe hepatic insufficiency [9].

It should be noted that patients with systemic autoimmune diseases have a high relative risk for developing liver disorders such as autoimmune hepatitis, primary biliary cirrhosis, nodular regenerative hyperplasia, even vascular syndromes such as Budd-Chiari syndrome [4].

This is why we must study hepatic function tests in these patients to detect alterations that can often coexist silently. Similarly, having a patient with alterations in the liver profile performed a thorough skin examination to determine the presence or absence of scleroderma, which would guide us to a Reynolds syndrome [8].

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