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SJOGREN'S SYNDROME AND PRIMARY BILIARY CHOLANGITIS OVERLAP: GOOD RESPONSE TO HYDROXYCHLOROQUINE TREATMENT

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BACKGROUND

Sjögren's syndrome (SS) can overlap with autoimmune diseases. The prevalence of SS in primary biliary cholangitis (PBC) is reported in 3.5 – 100% of cases. Both diseases present similar clinical manifestations and are characterized by immune-mediated destruction of exocrine glands with participation of Th17 response. Ursodeoxycholic acid (UDCA) is the first-line drug for PBC and significantly reduces the likelihood of liver transplantation and death. Hydroxychloroquine (HCQ) is used for some SS manifestations, but no previous studies reported its impact on hepatic evolution in SS-PBC overlap patients.

CASE REPORT

A 60-year-old woman presented on the rheumatology service with three previous episodes of encephalitis of unknown etiology with exclusion of infectious diseases and reported inflammatory arthralgia of wrist, metacarpophalangeal and proximal interphalangeal joints associated with xerostomia and alopecia. Exams to investigate SS were requested and showed: ANA cytoplasmic reticular 1/160, polyclonal hypergammaglobulinemia: 1.75 (VR: 0;55–1.46 g/dL), anticardiolipin-IgM: 56 (VR: < 40 MPL-U/mL); minor salivary gland biopsy with area of 6 mm², 2 inflammatory foci-score and degree of inflammation of 3; Schirmer test: right eye: 3 mm, left eye: 0 mm. She had negative tests for lissamine green, anti-Ro, anti-La, anti-Sm and anti-dsDNA antibodies. Complementary exams revealed elevated transaminases and cholestasis enzymes: aspartate aminotransferase (AST): 60 (VR < 35 U/L), alanine aminotransferase (ALT): 65 (VR < 35 U/L), alkaline phosphatase (FA): 198 (VR: 30-120 U/L), gamma-glutamyltransferase (GGT): 261 (VR < 38 U/L) and she was referred to a hepatologist, whose diagnosis was primary biliary cholangitis with positive antimitochondrial antibody: 1/160 (VR: nonreactive) and liver biopsy consistent with PBC. The results of other antibodies and viral hepatitis serologies were all negative. It was prescribed HCQ (5 mg/kg/day), omega-3 fatty acid (2 pills/day), AAS 100 mg/day, cholecalciferol 7000 UI/week and UDCA (15 mg/kg/day); however, the patient did not initiate UDCA because it has not yet been released via the Unified Health System (SUS). Despite this, after hydroxychloroquine treatment it was observed reduction in all transaminases and cholestasis enzymes: AST: 35 (VR < 35 U/L), ALT 35 U/L (VR < 35 U/L), GGT: 156 (VR < 38 U/L) and FA: 107 (VR: 30-120 U/L).

CONCLUSION

Ursodeoxycholic acid is the first-line drug to treat PBC and it has also been used to SS-PBC overlap. Biochemical response to UDCA is considered a strong predictor of long-term clinical outcomes. Hydroxychloroquine is used for some SS manifestations, but there are no studies evaluating its impact on the levels of liver enzymes in patients with SS-PBC overlap. We presented a case whose decreased transaminases and cholestatic enzymes after HCQ treatment without concomitant UDCA; being possible to raise the hypothesis that hydroxychloroquine may be another therapeutic option that could improve liver prognosis in overlap SS-PBC patients.