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PERIPHERAL ULCERATIVE KERATITIS REFRACTORY IN AN OVERLAP OF SJÖGREN'S SYNDROME AND SARCOIDOSIS: A CASE REPORT

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BACKGROUND

Sjögren's syndrome and sarcoidosis are diseases not usually overlap. The peripheral ulcerative keratitis (PUK) is the most common form of a corneal melt in rheumatoid arthritis, but is also described in vasculitis, e.g., granulomatosis with polyangiitis. There are few cases in association with sarcoidosis and Sjögren's, and sarcoidosis is a exclude criteria for Sjögren's.

CASE REPORT

A 44-year-old female patient went to a medical evaluation with complaints of ocular and oral dryness. She had positive anti-La and anti-Ro antibodies and a labial biopsy with epithelial dysphasia and chronic sialadenitis – compatible with Sjögren's syndrome. She presented negative tests for hepatitis B and C, HIV and HTLV 1 and 2. The patient had a score of 7 points in the 2016 ACR-EULAR criteria for Sjögren's syndrome with 3 points for a positive biopsy, 3 points for anti-SSA positivity and 1 point for positive Schirmer test. In 2012 and later again in 2014, she developed refractories erythematous maculopapular skin lesions in the palpebral area, knees and left forearm. She was submitted to a skin biopsy that was of granulomatous dermatitis with sarcoid pattern – highly suggestive of sarcoidosis. In that point, she was diagnosis with overlap Sjögren's syndrome and sarcoidosis. In 2015, the patient was submitted to a renal biopsy to investigate the cause of her decreasing renal function, that shown renal tubular acidosis type 1, and had a good response to the treatment with azathioprine and bicarbonate reposition. In 2017, the patient developed a severe case of ceratitids in the right cornea. Infectious causes, such as bacterial infection, syphilis and toxoplasmosis were discarded. She had a negative antineutrophil cytoplasmic antibodies (ANCA) serology and the rheumatoid factor was negative. There were no findings of erosive lesions in the joint X-ray of hands, feet and knees. While in ocular activity of PUK, the patient received treatment with topical and oral corticoids, azathioprine and cyclosporine without improvement of ocular symptoms. She had a good response to mycophenolate – treatment with anti-TNF and rituximab was unnecessary. Nowadays the patient presents a mild bulbar conjunctival hyperemia and diffuse corneal opacity without signs of disease activity.

CONCLUSION

In our case, PUK is associated with an overlap of Sjögren's syndrome and sarcoidosis. The ocular manifestation did not respond to the initial treatment with azathioprine and methotrexate and responsive to mycophenolate. The authors declare that the patient was oriented and signed the written informed consent form.