Case Report
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Behcet's syndrome associated with Sjogren's syndrome, an infrequent combination: Case report and literature review

Abstract

58-year-old woman with a history of xerophthalmia and xerostomia, asthenia and alopecia beginning at the age of 27, oral ulcers and recurrent episodes of bilateral parotitis. She attends rheumatology at the age of 52 (year 2016), without treatment. Laboratorial, ophthalmological and oral cavity examinations were performed, along with the biopsy of 2 minor salivary glands. Sjogren's syndrome (SS) was concluded, excluding other differential diagnosis. Hydroxychloroquine 400 mg/day was indicated, improving symptoms. After three years (year 2019), she added polyarthritis, large and recurrent oral ulcers and the appearance of genital ulcers. Behcet's syndrome (BS) was suspected and a genetic study was performed: HLA-B27 histocompatibility antigen negative, HLA Class I, locus B molecular typing: HLA B* 35 and HLA B*51 positive (nomenclature B35; B51 positive). This case is reported due to the infrequent association of SS and BS, both fulfilling the diagnostic criteria. A literature review was performed. Both pathologies were scarcelyn reported together in the literature. A common pathophysiological mechanism cannot be defined at the moment. It is intended to highlight the broad thinking of differential diagnoses in everyday situations in rheumatology. A high index of suspicion, a profuse history and correct physical examination are essential to arrive at the diagnosis.

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Introduction

58-year-old woman with a history of xerophthalmia and xerostomia, asthenia and alopecia beginning at the age of 27, persistent and daily, with a sensation of grittiness in the eyes, she also presented oral ulcers and recurrent episodes of bilateral parotitis. Requires use of tear substitutes several times a day. Requires fluid intake to swallow dry food. She attends rheumatology at the age of 52 (year 2016), without treatment. Laboratory is performed observing erythrocyte sedimentation rate of 59, C3 fraction of complement 117, C4 fraction of complement 16, Ac. Antinuclear (ANA) 1/320 by Hep-2 technique, with positive anti-Ro >200 and positive anti-La, negative anti-SM and anti-RNP. Ophthalmological examination: Tindal negative. Shimmer test positive: < 5 mm/5 min in both eyes. Lissamine green staining test > 5 in both eyes, evaluated by ophthalmology. Salivary flow without stimulation <0.1 ml/min and evaluated by stomatology.

A biopsy of 2 minor salivary glands of the lower hemi-lip was performed (2002). Macroscopic description: four slightly nodular pieces of soft tissue measuring up to 0.4 cm in diameter with a yellowish-pink hue. Pathological description: fragment of fibrous tissue and four units of minor salivary glands with focal and diffuse lymphocytic infiltrates involving the stroma and not compromising the epithelial component. Quantitative representation of the lymphocyte component with a score greater than 1 foci/4 mm2 was performed and compatibility with Sjogren's syndrome was concluded.

Serologies: HIV, HCV, VHBAgS, VHBanticore, VDRL negative.

No history of head and neck irradiation, no evidence of amyloidosis, no transplant history, no evidence of other clinical signs compatible with IgG4 disease. No history of taking other drugs or substances. Sjogren's syndrome was diagnosed and Hydroxychloroquine 400 mg/day was indicated, improving symptoms.

After three years (year 2019), she added polyarthritis of small metacarpophalangeal, proximal interphalangeal and bilateral carpal joints, for which Methotrexate 15 mg/week was added. Later (2020), she presented a new episode of bilateral parotitis that resolved with the administration of corticosteroids. Due to persistent, large and recurrent oral ulcers and the appearance of genital ulcers, Behcet's syndrome (BS) was suspected and a genetic study is performed: HLA-B27 histocompatibility antigen negative, HLA Class I, locus B molecular typing: HLA B* 35 and HLA B*51 positive (nomenclature B35; B51 positive).

The ophthalmological examination revealed: visual acuity without correction in the right eye: 6/10 and in the left eye 7/10, which improved with a pinhole. On external examination, eyebrows, eyelashes and eyelid skin preserved, without lesions. Ocular motility and photomotor and consensual reflex were preserved. Negative relative afferent pupillary defect, pupillary reflexes focused on primary gaze position. Biomicroscopy in both eyes: decreased tear meniscus, superficial keratitis. Patent lacrimal points, white sclera, pink conjunctiva without adherence, transparent cornea, no keratic precipitates observed. No cells in anterior chamber. Negative flare. Trophic iris, without nodules. Isochoric, reactive centered pupils without anterior or posterior synechiae, transparent lens. Intraocular pressure right eye 12 mmHg and left eye 11 mmHg. indirect binocular ophthalmoscopy in both eyes: pink papilla, with clear edges, cup 2/6. Brightness and macular architecture preserved. No alterations are observed in the posterior pole or in the peripheral retina. A-V crossings preserved, caliber and trajectory vessels preserved. optical coherence tomography: preserved macular architecture in both eyes. Central thickness right eye 282 um and left eye 284 um.

Discussion

This case is reported due to the infrequent association of Sjogren's syndrome and Behcet's syndrome, both fulfilling the diagnostic criteria.[1–3]

The literature in Pubmed was reviewed using controlled terms from the Virtual Health Library and Decs/Mesh and with the use of Boolean operators: Sjögren's Syndrome / Sjogren's Syndrome / Sjogren's Syndrome AND Behçet's Syndrome / Behcet Syndrome / Behçet's Syndrome, obtaining few reports.[4–7]

In the reported case, the first diagnosis, maintained for years, was Sjogren's syndrome (SS), with periodic rheumatological and ophthalmological controls. In SS, the appearance of oral ulcers is possible given the dryness of the mucous membranes, friction with the teeth and the alteration of the usual oral flora, which predisposes to thrush and superinfections, situations that overlapped the diagnosis of BS for many years, added to the fact that no ocular lesions suggestive of BS appeared.

In this particular case, probably the embarrassment of referring genital ulcers in the different consultations, did not alert the diagnosis to a greater extent. Years later, after not finding a gynecological diagnosis for the genital ulcers and a new anamnesis, it was possible to detect them, suspect BS and proceed to the diagnosis. At the ocular level, she did not present uveitis or other related alterations.[8]

Conclusion

Both pathologies were scarcely reported together in the literature. A common pathophysiological mechanism cannot be defined at the moment. The fact of diagnosing an autoimmune disease such as Sjogren's syndrome does not rule out the appearance and overlap of another rheumatological disease, such as Behçet's Syndrome, whose frequency is less common.[9–14] It is intended to highlight the broad thinking of differential diagnoses in everyday situations in rheumatology such as SS, with infrequent manifestations over the years that do not improve. A high index of suspicion, a profuse history and correct physical examination are essential to arrive at the diagnosis.

Acknowledgments

None.

Conflicts of interest

The authors declare no conflicts of interest.

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