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Marginal Corneal Ulcer Revealing Sjögren's Disease

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I. Introduction

Corneal immunological disorders are characterized by chronic inflammation of immune origin. Although these conditions are not infectious in themselves, they can sometimes be triggered by infectious agents. The etiological diagnosis is often complex and prolonged. The presence of an ulcer at the limbus of the cornea is particularly suggestive of an inflammatory cause: indeed, peripheral inflammatory corneal ulcers are frequently autoimmune in origin. Their diagnosis must be established quickly due to the risk of perforation, and also because they may be the first manifestation of a serious systemic disease that has not yet been diagnosed. The most common pathologies in this context include rheumatoid arthritis and Wegener's disease.

Sjögren's disease is an autoimmune inflammatory condition that affects exocrine glands, such as the salivary and lacrimal glands. The infiltration of T lymphocytes into these glands leads to their destruction and a decrease in secretion, resulting in xerophthalmia (dry eyes) and xerostomia (dry mouth).

The aim of this article is to describe, through a clinical case, a rare ophthalmological manifestation that may reveal Sjögren's disease: the marginal corneal ulcer.

II. Case Description

We present the case of a 30-year-old female patient who came in with a red and painful eye for one month. Despite initial treatment with artificial tears and topical antibiotics, her condition did not improve. She has a history of iron deficiency anemia.

Upon admission, the patient complained of ocular pain, photophobia, and decreased visual acuity. Ophthalmological examination showed visual acuity of 3/10, conjunctival hyperemia in the right eye, and a peripheral ulcer in the temporal-inferior zone, measuring 3 mm in its longer axis and staining with fluorescein (Figure 1). Corneal sensitivity was preserved. The fluorescein test revealed superficial punctate keratitis at stage 4 according to the Oxford classification, with a reduced break-up time. The anterior chamber was calm, without signs of uveitis, the lens was clear, and the vitreous was filamentous. The fundus was normal.

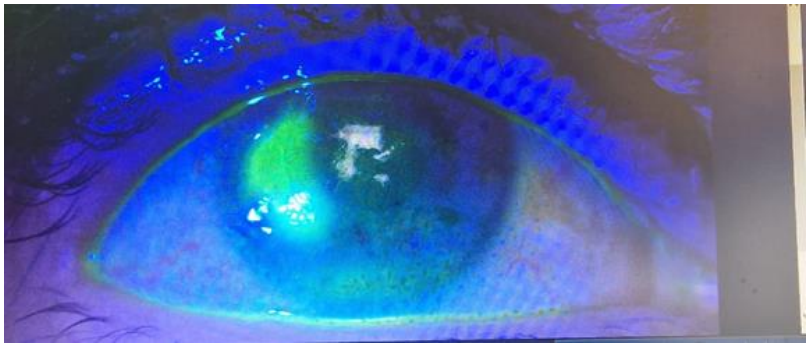

Figure 1: photo of the marginal ulcer staining with fluorescein

After a 48-hour therapeutic window with no treatment, a corneal sample was taken and returned sterile. Due to the ineffectiveness of antibiotic eye drops and the peripheral nature of the ulcer, we opted for local corticosteroid treatment (dexamethasone without preservatives) at a rate of 8 drops per day, combined with preservative-free artificial tears concentrated in sodium hyaluronate, and pain management with level 2 analgesics. The patient was referred to the internal medicine department for an etiological investigation, including infectious tests (Herpes, hepatitis B and C, HIV), inflammatory tests (ESR, CRP), and immunological tests (anti-SSA, anti-SSB antibodies, rheumatoid factor, anti-DNA antibodies).

Close follow-up showed a decrease in functional symptoms (photophobia, pain) without regression in the size of the ulcer, which remained fluorescein-positive (Figure 2). Treatment with autologous serum, diluted to 20% as eye drops, was initiated, with a dosage of six drops per day, along with an evening application of vitamin A ointment. The etiological investigation revealed positivity for anti-SSA antibodies, and a biopsy of the salivary glands showed lymphocytic sialadenitis with a focus score of ≥ 1. The diagnosis of Sjögren's disease was confirmed.

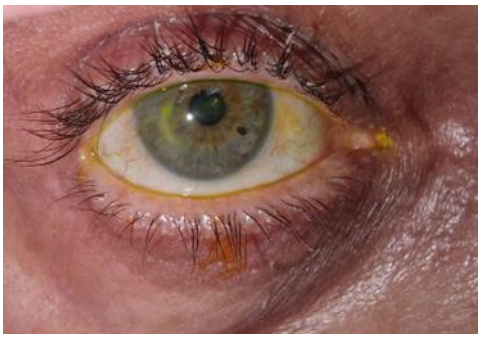

Figure 2: Photo showing the evolution of the ulcer after corticosteroid treatment

Clinical evolution was favorable with improvement in symptoms, a decrease in the size of the ulcer, and total healing, leaving a corneal opacity. The final visual acuity was 6/10, due to the development of a subcapsular cataract (Figure 3).

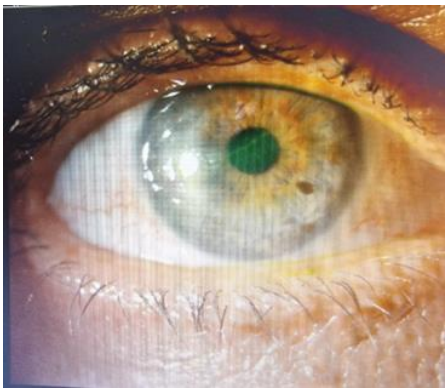

Figure 3: Photo at the end of treatment showing a corneal opacity

III. Discussion

Marginal corneal ulcer is a serious ocular complication, often associated with various autoimmune diseases. In some cases, corneal involvement may occur several years after the onset of systemic disease, while in others, it may manifest first. The presence of a corneal ulcer in patients with systemic autoimmune disorders may indicate potential progression of a life-threatening disease.

Several autoimmune diseases are linked to marginal ulcers, with rheumatoid arthritis being the most common. Cases of Wegener's granulomatosis, relapsing chondritis, systemic lupus erythematosus, and nodular polyarteritis and its variants have also been observed. Marginal ulcers can develop at any stage of these conditions, usually appearing after other manifestations, although they can sometimes be the revealing sign of an autoimmune disease.

In our case, the marginal ulcer was a revealing symptom of Sjögren's disease. This chronic autoimmune condition is characterized by inflammation and destruction of the exocrine glands, particularly the lacrimal and salivary glands. It presents with various ocular symptoms, such as foreign body sensations, variations in tear production, itching, and blurred vision. Clinical examination often reveals conjunctivitis, keratinization, and punctate or filamentary keratitis. Mooren's ulcer, although rare, represents an atypical manifestation of Sjögren's disease, often associated with a dry syndrome and decreased tear secretion, leading to a sandy sensation in the eyes.

Diagnosis relies on specific criteria, including the positivity of anti-SSA/Ro antibodies, focal lymphocytic sialadenitis, ocular staining scores, results from the Schirmer test, and unstimulated salivary flow rates. These criteria are essential for diagnosing and managing Sjögren's syndrome, ensuring prompt intervention for ocular complications such as ulcers.

Therapeutic management represents a considerable challenge, with limited success for both medical and surgical treatments, which can lead to blindness in severe cases. General treatment is managed by the internist, based on the severity of the condition and the presence of other autoimmune diseases, including corticosteroids or, in severe forms, immunosuppressants like hydroxychloroquine and pilocarpine, which promote salivary secretion. Close collaboration is crucial to manage complex cases and optimize immunosuppression.

Locally, preservative-free tear substitutes rich in hyaluronic acid are recommended. The nocturnal application of vitamin A ointment contributes to patient comfort. Rapid occlusion of all four lacrimal puncta is advised to retain secreted tears and maintain substitutes in place. Options include silicone plugs or absorbable internal prostheses for the lacrimal ducts, to be renewed every six months. For managing the ulcer, autologous serum can also be used. This involves using the patient's autologous serum, diluted to 20%, as eye drops. This serum is rich in growth factors (EGF, NGF, TGF-β) that promote epithelial healing, as well as molecules such as vitamin A, fibronectin, neuropeptide P, and lysozyme. The usual dosage is one drop four times a day over the long term. Its tolerance is excellent, due to a pH and osmolarity similar to those of tears. Being non-preserved, it is crucial to respect the cold chain and ensure good hygiene during its use. Its clinical effectiveness is significant, leading to improvement in symptoms and ocular surface, helping patients accept the constraints of its application. Autologous serum does not have a specific regulatory status and is considered either a magistral preparation or a biological drug. Its preparation requires cooperation between the hospital service that draws the blood and the hospital pharmacy that prepares the eye drops.

IV. Conclusion

Primary Sjögren's syndrome is an autoimmune disease that primarily affects exocrine glands, such as the salivary and lacrimal glands. Additionally, systemic involvement is common. Primary Sjögren's syndrome is of particular interest to ophthalmologists, as it constitutes an important differential diagnosis in conditions associated with dry eye disease. Furthermore, ocular tests for more precise diagnosis and monitoring of primary Sjögren's syndrome have become increasingly important, and new therapies for local and systemic treatment are evolving due to a better understanding of the immunological mechanisms and molecular pathways involved in the pathogenesis of primary Sjögren's syndrome.

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