Corneal Melting in Undiagnosed Goujerot Sjogren’s Syndrome: A Case Report

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ABSTRACT

Keratolysis or corneal melting is a very rare situation that might occur in different inflammatory systemic diseases such as Sjogren syndrome. In the case of sterile corneal ulcers, characterized by the absence of ocular inflammation, the optimal treatment has not yet been established. We report the case of a unilateral aseptic keratolysis revealing primitive Sjogren guzzler syndrome and having responded well to the treatment. Clinical case: a 56-year-old woman who presented with a notion of inflammatory polyarthralgia for 4 years with self-medication by oral non-steroidal anti-inflammatory drugs NSAID presented a month before her first examination, a reduced visual acuity with foreign body sensation in both eyes. After a month of ophthalmic examination for a corneal ulcer, the visual acuity dropped to HM, and the examination found a corneal perforation measuring less than 1 mm with a positive spontaneous seidel and an athalamia. A treatment with cyanoacrylate glue was performed along with medical treatment. A labial biopsy performed found a lymphoplasmacytic infiltration of the exocrine gland in favor of a primitive Gougerot Sjogren syndrome. Gougerot-Sjögren syndrome is a systemic autoimmune disease characterized by damage to the exocrine glands, in particular the lacrimal and salivary glands. The treatment of corneal perforations is based on different choices such as cyanoacrylate glue, amniotic graft, conjunctival recovery, and keratoplasty.

Keywords: Corneal perforation, inflammatory diseases, Sjögren's syndrome.

1. INTRODUCTION

The ocular manifestations of autoimmune diseases are mainly dry syndrome, scleritis and keratitis. Keratolysis in the absence of an infective cause may be an indication of systemic autoimmune disease [1]. Aseptic keratolysis, which is much rarer, is characterized by its central or paracentral location and is a real therapeutic emergency [2]. It is associated with significant visual morbidity [3].

Aseptic keratolysis can manifest in patients with preexisting tear-film instability resulting from keratoconjunctivitis sicca (KCS) as part of primary or secondary Sjögren’s syndrome (SS) [4]. The purpose of our work is to recall the clinical picture and the diagnosis procedure, to insist on the gravity of this event, as well as to update the therapeutic component, through a clinical case.

2. CASE REPORT

We report the case of a 56-year-old woman who presented with a notion of inflammatory polyarthralgia for 4 years with self-medication by oral non-steroidal anti-inflammatory drugs NSAID, who presented a reduced visual acuity with foreign body sensation in both eyes for a month.

The ophthalmological examination found a visual acuity of 0.10 with a slight conjunctival hyperemia in both eyes.

The right eye presented para-central corneal ulcer of 1 × 1 mm with diffuse KPS (Fig. 1a). The left eye had a para central corneal ulcer of 1.5 mm/1 mm with diffuse KPS. (Fig. 1b).

Corneal sampling with the mycological, parasitological and bacteriological study were carried out and were negative. Schirmer’s test was at 2 mm in both eyes.
The patient was put on lubricating agents, and after an internist consultation, a high dose of corticosteroid therapy was prescribed.

The evolution was marked by the healing of the ulcer on the right eye. The left eye was complicated with a corneal melting after 20 days of treatment, the clinical examination of this eye finds: a VA=HM, a corneal perforation <1 mm, a positive spontaneous seidel and an athalamie (Fig. 2).

A treatment with cyanoacrylate glue was performed along with medical treatment. The biological tests found: rheumatoid factor, antinuclear antibodies and native anti-DNA antibodies, all negative.

A labial biopsy performed found a lymphoplasmacytic infiltration of the exocrine gland in favor of a primitive Gougerot Sjogren syndrome.

The clinical course was marked by epithelial healing of the ulcer in both eyes, leaving corneal opacities with a neovascular appeal that regressed under local corticosteroid therapy (Figs. 3a and 3b). Visual acuity is at 0.7 in the right eye and 0.5 in the left eye.

3. Discussion

Non-infectious corneal melt is a rare yet potentially blinding entity as it may lead to perforation with devastating consequences. Several systemic disorders, including SS and other connective tissue diseases, have been identified as possible predisposing factors [1]. SS is a chronic inflammatory disorder characterized by exocrine gland dysfunction and a variable systemic course. Lymphocytic infiltration of the lacrimal and salivary glands results in the classic sicca complex characterized by dry eyes KCS and dry mouth (xerostomia). SS may occur alone (primary) or in association with other autoimmune diseases [5].

The presence of an aseptic central corneal ulcer without an infiltrate should suggest an inflammatory origin in relation to a systemic disease [6]. Corneal ulcers in the context of autoimmune or inflammatory pathologies are mainly peripheral. The diagnosis should be made in the presence of any appearance of die-casting, central or paracentral site and should be investigated first for rheumatoid arthritis (RA) or Sjogren’s syndrome (GS). Aseptic keratolysis is a real evolutionary turning point in the disease.
Local corticosteroid therapy is controversial and systemic treatment is most often necessary to achieve ulcer healing. It will be necessary to combine an immunosuppressant as well as local lubricating treatments, sometimes with the use of a therapeutic lens or an amniotic membrane graft [6].

Tzamalis et al. reported in their case report; a case of sterile corneal ulceration leading to perforation as the initial presentation of primary SS, which was treated effectively with a bandage contact lens, autologous serum eye drops, topical RGTA (poly-carboxy methylglucose sulfate), steroids, and systemic immunosuppression, avoiding surgical intervention [1].

4. Conclusion

Corneal ulcers in the context of autoimmune or inflammatory pathologies are mainly peripheral. The diagnosis should be made in the presence of any appearance of die-casting, central or paracentral site and should be investigated first for rheumatoid arthritis (RA) or Sjögren’s syndrome (GS). Aseptic keratolysis is a rare and serious ocular complication and a real evolutionary turning point in the disease [2] with significant performative potential. The treatment mainly uses corticosteroids and requires close collaboration with the internists.

Conflict of Interest

Authors declare that they do not have any conflict of interest.

References