Short communication

Mooren’s ulcer: 30 years of follow-up


Centro de Oftalmología Barraquer, Institut Universitari Barraquer, Barcelona, Spain

Abstract

Case report: A 33-year-old Caucasian female presented with epiphora, ocular pain, and foreign body sensation in both eyes for one month. Examination revealed bilateral peripheral corneal ulcers. The patient had been treated with immunomodulators, and she was treated in the left eye with peripheral semi-circular keratoplasty, penetrating keratoplasty, conjunctival–corneal–scleroplasty, buccal mucosal graft, tibial osteo-keratoprosthesis and finally, retinal detachment.

Discussion: Mooren’s ulcer is an immunological corneal disease. This lesion must be treated initially with immunomodulators. Surgical treatment should be considered when a risk of corneal perforation is present, when the perforation appears, or under acute necrosis.

® 2016 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.

Úlceras de Mooren: 30 años de seguimiento

Resumen

Caso clínico: Mujer de 33 años de raza blanca que consultó por sensación de cuerpo extraño, epífora y dolor intenso, de un mes de evolución, en ambos ojos (AO). El examen biomicroscópico objetivó una úlcera corneal periférica bilateral. Precisó tratamiento con inmunomoduladores y fue intervenida en el ojo izquierdo de queratoplastia en corona semi-circular, queratoplastia penetrante, conjuntivo-córneo-escleroplastia, recubrimiento de mucosa bucal, osteo-queratoprótesis tibial y, finalmente, de desprendimiento de retina.

Discusión: Las úlceras de Mooren son una afección corneal inmunológica que requiere tratamiento inmunomodulador, reservándose el quirúrgico ante el riesgo inminente de perforación, cuando esto ha ocurrido, o en los casos de necrosis aguda.

® 2016 Sociedad Española de Oftalmología. Publicado por Elsevier España, S.L.U. Todos los derechos reservados.


* Corresponding author.

E-mail address: ferranvilaplanamira@gmail.com (F. Vilaplana).

2173-5794/© 2016 Sociedad Española de Oftalmología. Published by Elsevier España, S.L.U. All rights reserved.
Introduction

Mooren’s ulcer is a chronic inflammatory process of the marginal cornea. It can be either unilateral or bilateral (50%). It affects the conjunctive-scleral–corneal limbo, and symptoms include pain, photophobia and epiphora. It grows circumferentially, and can affect the entire cornea and lead to perforation. It is largely found in developing countries. Its etiology is usually multifactorial; physical, chemical or surgical trauma and infectious or inflammatory process can be triggering factors, but the causal mechanism is immunological. There are multiple therapies, but current treatments are based on immunomodulation, with surgery reserved for cases with imminent risk of perforation, perforation or acute corneal necrosis.

Case report

33-year old woman who came to emergency room (December 1984) with sensation of foreign object, epiphora and intense pain for the last month in both eyes (BE). Visual acuity was 0.85 with correction of +1; –0.25 × 0° in the right eye (RE) and 0.8 with correction of +1; –2.5 × 0° in the left eye (LE). The biomicroscopic examination revealed peripheral corneal ulcers of 6–7 h in RE and 6–9 h in LE (Fig. 1A and B).

The cultures made in blood, chocolate and Sabouraud agar were negative. Internal medicine and laboratory studies showed no concomitant systemic disease.

In the RE the exacerbations were controlled with topical corticosteroids (disodium dexamethasone phosphate 1 mg/1 ml). The conjunctival biopsy of the LE showed an immunological process with collaborating T-cells and macrophages.

The patient received treatment, according to the phase, based on topical and systemic corticosteroids (120 mg methyl prednisolone with decreasing doses), topical 2.5% cyclosporine A 3 times a day in BE, oral azathioprine (75 mg/day) and intravenous cyclophosphamide (1.2 g in 250 cc of saline).

Due to poor anatomical evolution and risk of perforation, we decided to perform surgery on the LE with peripheral semi-circular keratoplasty (June 1985) (Fig. 2A and B). After 4 months (October 1985) and after acute donor tissue necrosis, the patient presented corneal perforation (Fig. 3A and B) for which she underwent emergency penetrating keratoplasty (12 mm). We subsequently observed necrosis of the graft and relapse of the process, indicating conjunctival–corneal–scleroplastic (16 mm) (June 1986) (Fig. 4A and B), with an unsatisfactory immediate outcome, evolving to corneal edema. As a result of the new relapse of the process, and in view of imminent risk of perforation, we performed penetrating keratoplasty (10 mm) with extracapsular cataract removal (September 1987) (Fig. 5A and B). Due to the new surgical failure, it was decided to perform a buccal mucosal graft (October 1987) (Fig. 6A). After 22 years of observation with no signs of reactivation, she underwent tibial osteo-keratoprosthesis (September 2009) (Fig. 6B) and was finally operated for retinal detachment (September 2009) by silicone band buckle, air exchange vitrectomy, using a contact lens for vitreal–retinal surgery in patients who have undergone osteo-odonto-keratoprosthesis (Nadal and Barraquer). Her visual acuity at the last control (May 2015) was 0.9 with correction of +2.00; –2.00 × 60° in RE and 0.04 with no improvement with correction in LE.

Fig. 1 – (A) 6–7 h peripheral corneal ulcer in RE. (B) 6–9 h peripheral corneal ulcer in LE.

Fig. 2 – (A) Peripheral corneal thinning. (B) Peripheral semi-circular keratoplasty.
Discussion

Mooren’s ulcer is an immune condition with largely T-cells and macrophages, which can be determined with a simple conjunctival biopsy. Treatment must be primarily with immune modulators, despite which it can evolve to perforation. The utility of topical 2% cyclosporine as adjuvant treatment, which we used 30 years ago, has recently been shown.

Lee et al. tested the use of topical 0.02% tacrolimus, due to its immune suppression effect, for chronic inflammatory processes of the ocular surface, with good results, although only one of the 12 patients was diagnosed with Mooren’s ulcer. Xie et al. used topical treatment in the form of 0.1% FK506.
eye drops in 15 eyes of 9 patients, with good results; however, follow-up was only 22 months, with no recurrences in that period.

The risk of perforation is highly variable, ranging from 10% to 65% depending on the author; youth is a risk factor in this case.

Surgery is indicated in cases of risk or perforation and in acute necrosis. Lamellar keratoplasty can be useful, but it only worked for a short period of time in our patient. Amniotic membrane transplantation is ineffective, and should only be used in acute situations with extreme corneal thinning, and as a coadjuvant with immune-modulating therapies. The Cochrane studies found no scientific evidence for recommending any specific type of treatment for this condition.

In sum, Mooren’s ulcer is a corneal condition with an immune component that requires an immune-modulating treatment; topical therapies with tacrolimus and FK506 can be useful, and surgery should be reserved for imminent risk of perforation, perforation or cases of acute necrosis.

**Conflicts of interest**

None of the authors has any relationship, economic or other, with pharmaceutical companies that could be involved in the surgical materials used.

**Acknowledgments**

Thanks to Andrés Maeso and the Photography Department of Centro de Oftalmología Barraquer for their help in the photographic archive.

**References**


