Case report

CORNEAL MELTING

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Summary

Corneal melting is a form of corneal inflammation that involves the outer part of the cornea. It may be associated with a variety of autoimmune diseases. It is often difficult to make the right diagnosis without detailed tests. We present three cases of peripheral ulcerative keratitis (PUK) with different systemic disease and discuss conservative and surgical methods of treatment.

Key words: corneal melting, peripheral ulcerative keratitis

Introduction

Corneal melting, also known as peripheral ulcerative keratitis (PUK) belongs to the group of peripheral keratopathies. PUK is a form of ocular inflammation that involves the outer part of the cornea [1]. The most severe complication is the progress of marginal corneal thinning leading to perforation.

The pathogenesis remains unclear in most of these disorders but autoimmune mechanisms are most likely involved. The peripheral cornea has distinct morphologic and immunologic characteristic that predispose it to inflammatory reactions. Unlike the central cornea, the peripheral cornea is closer to the limbal conjunctiva where capillary beds provide both nutrients, and immunocompetent cells such as macrophages, Langerhans cells, lymphocytes, plasma cells. As a result, any inflammation of the peripheral cornea may be caused by the invasion of a microorganism, or the deposition of immune complex, produced by local or systemic immune diseases. The activated complement pathway increases the vascular permeability and recruits the inflammatory cells. The neutrophils and macrophages infiltrate the cornea and secrete collagenases and proteases, thus leading to destruction of peripheral corneal stroma.

Approximately 50% of all noninfectious PUK are associated with collagen vascular diseases.

The history includes foreign body sensation with or without pain, tearing, photophobia and reduced visual acuity.

Physical examination reveals crescent-shaped

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corneal ulcer in a zone of 2 mm of the limbus, the overlying epithelium is absent and in the affected area the underlying stroma is thinned, the edge is sharp and not infiltrated. The ulceration may or may not be associated with cellular infiltrates in the stroma, and adjacent conjunctiva can be minimally or severely inflamed. In severe cases, the destruction of the cornea progresses centrally and circumferentially. The process is usually unilateral and limited to one sector but may also be bilateral and extensive.

The treatment of PUK is determined by the severity of findings within the cornea and the extra-ocular disease. The goal of treatment is to decrease the melting of the stroma. Systemic corticosteroids are the cornerstone of therapy. When applied topically, they have a variable effect. In more severe cases immunosuppressive medications have been used, alone or in combination. In cases of impending perforation surgery is the choice of treatment [2, 3].

The aim of this report is to assess the difficulties in the diagnostic process and methods of treatment of PUK.

Clinical cases presentation

Case 1

A 39-year-old female presented with a history of irritation for 7 years and gradually decreased visual acuity of the right eye. She had had a systemic disease – Acne rosacea, for 4 years. Slit lamp examination revealed peripheral corneal ulcer with a thinned stroma, engaging the whole circumference of the cornea, with superficial new blood vessels and clear inner edge, mild blepharitis and injection of the conjunctiva (Fig.1). Left eye was normal.

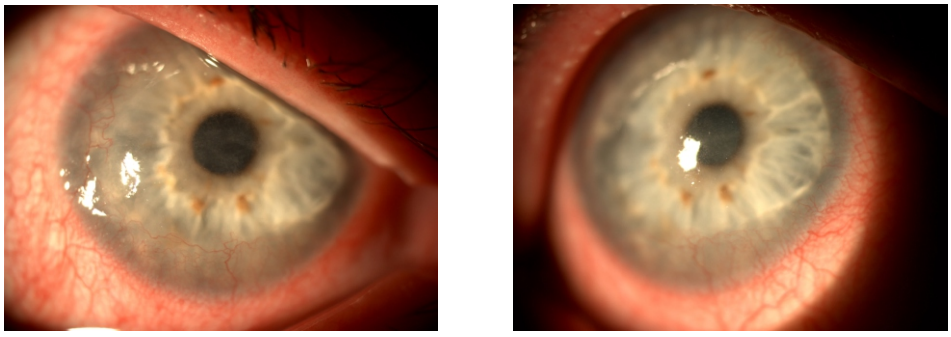

Figure 1. Peripheral corneal ulcer

Visual acuity for right eye was 0.09-0.1. Schirmer test was normal. Laboratory tests: Negative ocular microbiological result, elevated ANA, low level of IgA, high serological data for H. Pylori. After dermatological consultation, the diagnosis was ocular form of Acne rosacea. She was treated with corticosteroids – topically and parabulbar (p.b.), systemic Isoprinosine and antibiotic.

The ulcer did not progress further and a year later the new blood vessels almost disappeared and the best corrected visual acuity for this eye was 0.9-1.0 (Fig. 2).

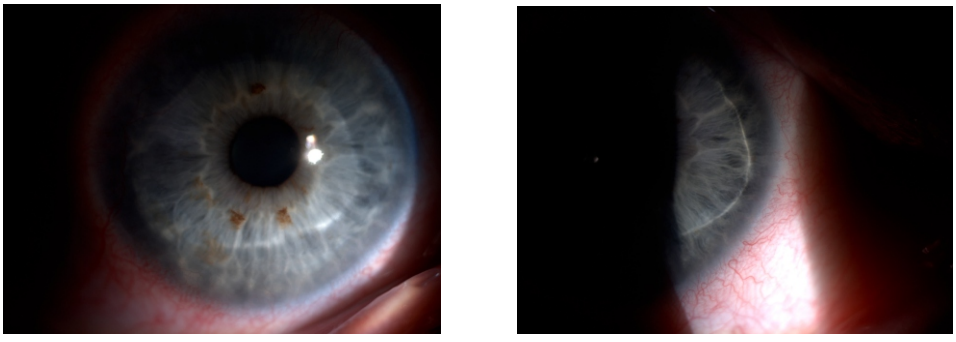

Figure 2. After corticosteroid treatment

A 76- year-old male presented with complaints of irritation, foreign body sensation and redness in his left eye. He had a history of RA for 12 years that had not been systemically treated.

Ocular examination showed circumferential stromal thinning, superficial new blood vessels and moderate injection of conjunctiva. Visual acuity for left eye was 0.5. Laboratory analysis showed high level of SRE (58) and Waller- Rose (72). Microbiological analysis of ulcer was non-contributory. Right eye was normal.

The therapy was initiated with topical and p.b. steroids, antibiotic, cycloplegics and artificial tears, but with no relief. The patient underwent a short treatment course with decreasing doses of a systemic corticosteroid and antibiotic Doxycycline 100 mg daily. Despite the treatment, the ulcer progressed centrally and an epithelial defect appeared in the right eye from 4 to 5 o'clock at the limbus, stained with fluorescein (Fig. 3; Fig. 4).

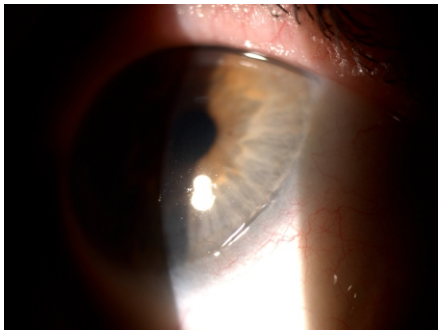

Figure 3. Right eye

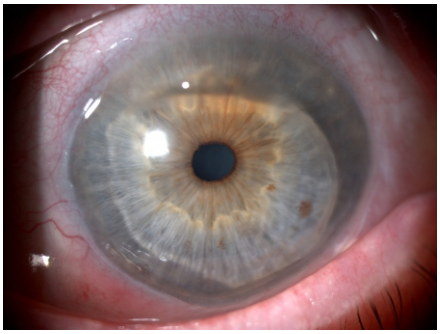

Figure 4. Left eye

The patient underwent a surgical procedure for his left eye – resection of conjunctival tissue adjacent to the cornea.

Three months later, the patient presented with the same complaints. Slit lamp examination revealed no progress of the ulcer, but infiltrates in the nasal part of the stroma were present (Fig. 5).

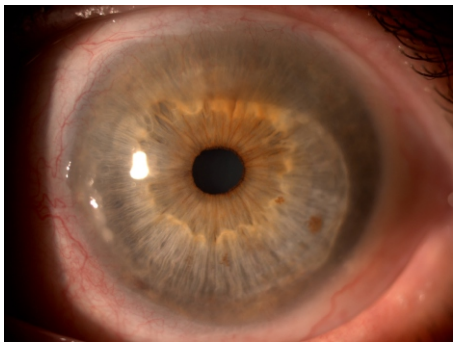

Figure 5. Three month after treatment

Topical therapy with steroids continued, but the systemic disease was not treated. A month later patient felt more comfortable and the infiltrates disappeared (Fig. 6).

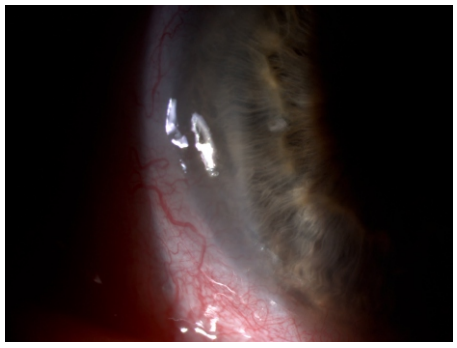

Figure 6. A month after corticosteroid treatment

Case 3

A 72-year-old female, diagnosed with corneal perforation, was referred to the clinic. Two weeks before, she had complaints of increasing irritation and, after an examination had been treated with Antibiotic (Floxal) for conjunctivitis. Treatment with a corticosteroisteroid was added 10 days later for two days. The patient had a long history of irritation in both eyes, without ocular examination and treatment. There was a history of pain in the joints but no systemic diseases. The examination revealed blepharospasm, mixed injection in her right eye, corneal perforation nasally on the limbus (3-4 o'clock) with iris prolapse. From 5 to 9 o'clock on the limbus, a crescent-shaped melting had caused thinning to 30% of the corneal thickness 2mm wide. A non-infiltrated leading edge did not stain with fluorescein, there was a mild iritis with drawn pupil at 3 o'clock. The examination of the lens revealed first signs of cataract.

The follow-up examination two weeks later revealed that the corneal graft was non-transparent, with superficial and deep new blood vessels, and epithelized surface (Fig. 13).

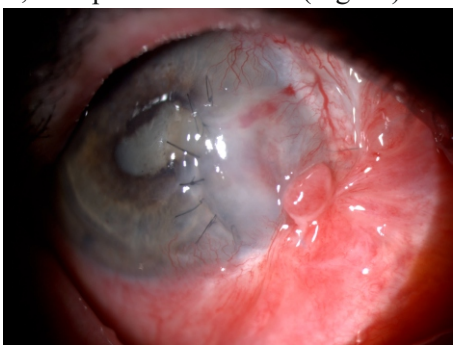

Figure 13. Five weeks after keratoplasty

The patient was re-evaluated two weeks later. The graft was well healed, non-oedematous and vascularised. The remainder of the cornea was clear, the melting on the limbus had not progressed. The pupil was irregularly shaped, narrow with synechia. The lens examination revealed intumescent cataract. Visual acuity was PPLC. The process had not advanced in the left eye (Fig. 14).

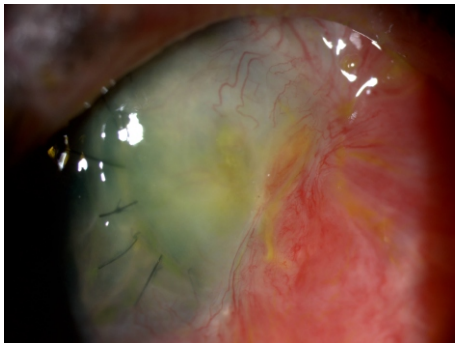

Figure 14. Seven weeks after keratoplasty

Discussion

PUK has a variety of clinical presentations. Patients typically describe non-specific foreign body sensation or pain, reduced visual acuity and watering eye. Slit lamp examination reveals non-infiltrating, crescent shape ulcer at the peripheral cornea, with adjacent injection of the conjunctiva. These findings are non-specific and may remain undistinguished from other inflammatory disease of the outer cornea.

Many systemic autoimmune diseases are associated with PUK. It has been described in patients with rheumatoid arthritis (RA), rosacea, Wegener's granulomatosis, systemic lupus erythematodus, polyarteritis nodosa, Sjogren's syndrome. In some disease, corneal involvement occurs after the systemic disease has been present for many years, whereas in others it may be the first manifestation. RA is the most common systemic disease associated with PUK, but the onset is late in the course of disease and indicates worsening of the systemic disease [4].

To make the right diagnosis, a thorough history, local examination and full general examination with diagnostic tests are needed. It is very important to rule out the local causes - infectious and non-infectious. Mooren's ulcer has to be ruled out too. It presents as a chronic unilateral or bilateral painful ulceration of the peripheral cornea, usually in the interpalpebral fissure and advances rapidly. It appears in otherwise healthy individuals without any associated systemic disease. The pain can be very severe [5].

Treatment is determined by the severity of intracorneal and extraocular pathological findings. It includes local treatment to prevent or reduce corneal damage, and systemic therapy to control the underlying disease. Corticosteroids are the cornerstone of therapy – systemic and topical, although the effect varies due to the delay of re-epithelization and eventual superinfection. In cases of severe, rapidly developing melting, and life-threatening systemic disease the management requires systemic immunosuppressive therapy.

Conservative treatment may be combined with surgical care. Surgical resection of conjunctival tissue adjacent to PUK has been promoted as a means to decrease the access of inflammatory cells and agents to the peripheral cornea. In 1972, Brown put the blame to the conjunctiva as a source of collagenase and proteoglycanase enzymes that caused the corneal ulceration. He also suggested excision of limbal conjunctiva as a possible treatment [6].

When corneal perforation is imminent or has already occurred, several surgical options exist, the main goal being to preserve the integrity of the eyeball. Tectonic procedures, including cyanacrylate glue, amniotic membrane, lamellar keratoplasty, penetrating keratoplasty, and corneoscleral patch grafts, are performed. Despite the improvement of graft survival with cytotoxic therapy, the outcomes of penetrating keratoplasty are disappointing. The most common complications following keratoplasty are suture loosening, epithelial keratopathy, corneal ulceration, recurrence of corneal melting

Her left eye showed mild chronic injection of the conjunctiva. Cornea was transparent, melting at 4-5 o'clock on the limbus, with superficial new blood vessels on that area. Lens examination revealed incipient cataract (Fig. 7; Fig. 8; Fig. 9). The achieved correction of visual acuity for the right eye was 0.15, and for left eye - 0.2. The Schirmer test performed revealed no deviations. Laboratory tests proved elevated rates of ESR – 21 and AST 600 (<200). The immunological status was normal. A consultation with a rheumatologist showed polyarthrosis.

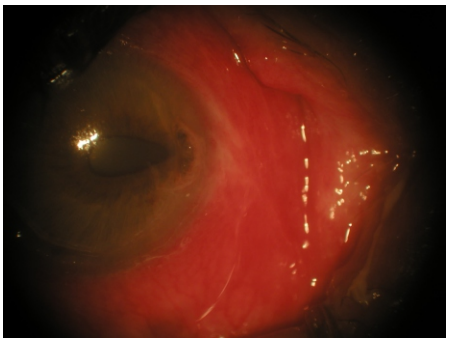

Figure 7. Right eye

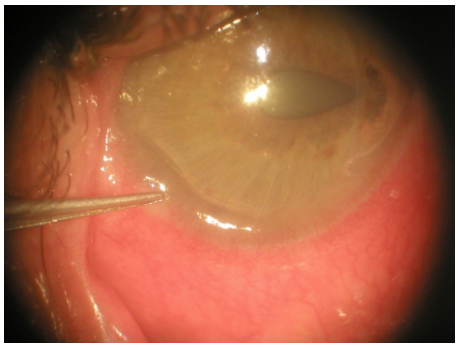

Figure 8. Right eye

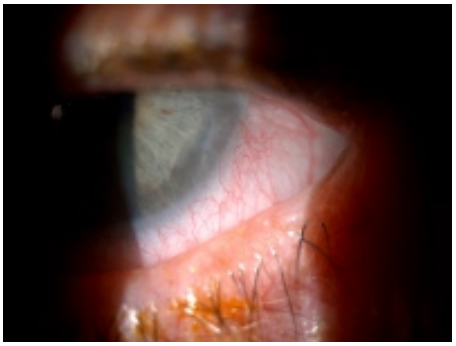

Figure 9. Left eye

A prompt surgical management of direct closure of the wound and reposition of the iris was performed. As a result, the integrity of the eye was restored though with certain deformation of the cornea (Fig.10).

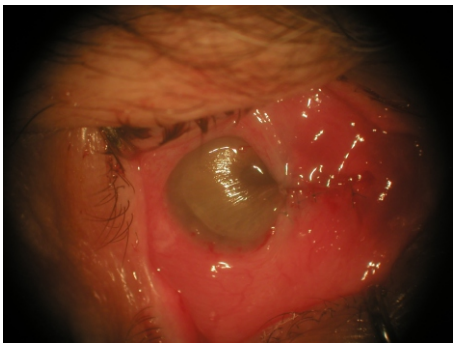

Figure 10. Direct closure of the wound of right eye

A week later, the patient underwent penetrating keratoplasty with reconstruction of the iris – suturing the iris and forming a pupil. The post-operative treatment for the right eye included a topical antibiotic, artificial tears, and corticosteroids (a slow-release corticosteroid p.b. – two times – a week after keratoplasty and 15 days later). A corticosteroid p.b. was administered for the left eye two times. Two weeks after keratoplasty, the graft was transparent and well-adapted. The anterior chamber was hermetic and uneven (Fig.11).

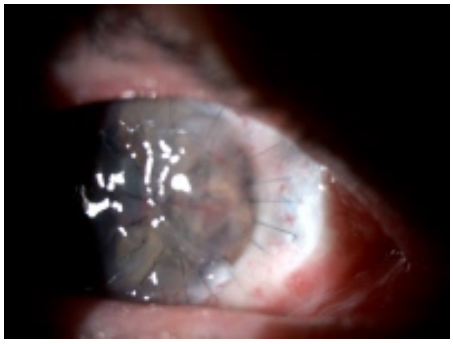

Figure 11. Two weeks after keratoplasty

Twenty days after keratoplasty, the graft was oedematous hemi-transparent, with several loose sutures. The sutures were removed, risking dehiscention (Fig. 12).

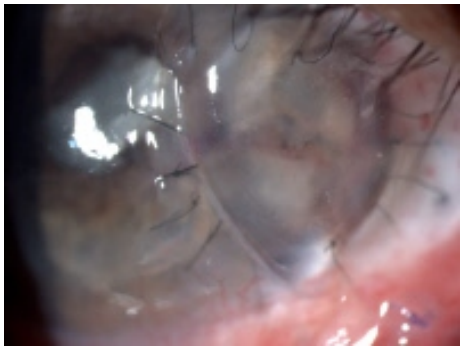

Figure 12. Twenty days after keratoplasty

or deep stromal defects. Maeno *et al*. showed that although the number of penetrating keratoplasties performed for ulcerative keratitis has remained low, it demonstrates the highest likelihood of requiring a regraft [7, 8, 9].

Conclusion

The melting of the cornea is a rare disease. The diagnosis is based on thorough clinical and immunological analysis. Often, it may be self-limited but it may progress to perforation of the eyeball. Successful management requires local conservative and surgical treatment, and the administration of systemic disease treatment in association with physician of another specialty who has expertise in immunosuppressive therapy.

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