Mooren’s ulcer a rare cause of peripheral ulcerative keratitis

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Abstract

Authors report a rare cause of peripheral ulcerative keratitis. Mooren’s ulcer is a chronic, painful, relentless ulcerative peripheral keratitis that begins from periphery of cornea near limbus as narrow crescent corneal infiltrate which then progresses circumferentially and centrally. Mooren’s ulcer gets misdiagnosed as peripheral bacterial ulcer or another non-infective peripheral ulcerative keratitis.

Keywords: Mooren’s ulcer, Peripheral ulcerative keratitis, Autoimmune

Introduction

Mooren’s corneal ulcer was first described by Bowman in 1849 [1]. McKenzie described it as chronic serpiginous ulcer of cornea or ulcus rodens [2]. Authors here would like to report a rare cause of Peripheral Ulcerative Keratitis – Mooren’s ulcer in a 34 year old male patient with no systemic diseases. Patient typically presented with complaint of ocular pain and ulcer at peripheral cornea and characteristically had overhanging lip of the margin involved only anterior corneal layers in both eyes[3]. Mooren’s ulcer is an idiopathic condition with no other related systemic disease. It is an autoimmune disease in which antibodies are directed to corneal stroma [4]. Usually Mooren’s ulcer are misdiagnosed and wrongly treated, knowledge of which will help to differentiate and diagnose from other causes of peripheral ulcerative keratitis like infective keratitis, Pellucid and Terriens marginal degeneration etc and correct intervention can be done.

Case Report

A 34 years old male came to Dr D.Y. Patil Medical College, Hospital and Research Institute, Kolhapur Ophthalmology OPD with complaint of pain in both eyes since last 2 months.

Patient was apparently alright 2 months back when he started complaining of pain and foreign body sensation, photophobia in right eye which was associated with clear watery discharge. Patient then went to local doctor who prescribed oral analgesics and topical antibiotics. The symptoms were not resolved.

After 1 month he developed similar complaints in the left eye. Patient then continued using the same eye drops. After 2 months patient complained of severe pain in both eyes for which he came to our ophthalmology OPD. He gave no history for blunt trauma, chemical injury, skin rashes or vesicles over face or any other body part, or joint pains. He got operated for cataract surgery 3 years back in both eyes. Patient had no systemic diseases.

Following were the slit lamp examination findings:

- **Right eye** was pseudophakic with BCVA of 6/24, with normal lids, mild circum-corneal congestion, normal sclera & iris, anterior chamber was showing no reaction.
Cornea showed a peripheral ulcer inferiorly from 4 o’clock to 8 o’clock. Ulcer was progressive and crescentic in shape. It was extensively undermined with over hanging edge. It showed stromal melting. There was no clear zone between ulcer and the limbus. Thinned, scarred and vascularized cornea was seen inferiorly from where the ulcer must have initially progressed (Fig 1 and 2).

Left eye was also pseudophakic with BCVA of 6/18, normal lids, sclera, iris and anterior chamber with mild circumcorneal congestion.

Cornea showed a peripheral ulcer inferiorly from 5 o’clock to 9 o’clock. It was centrally more progressive and crescentic in shape. Ulcer was similarly undermined with over hanging edge with less stromal melting and thinning. There was no clear zone between ulcer and the limbus. (Fig 3 and 4)

Investigations were done which showed normal haemoglobin, RBC, WBC and ESR.

Patient showed negative serology for VDRL (suspecting syphilis), RA factor (rheumatoid arthritis), ANA (systemic lupus erythematosus), Hepatitis C. Liver and Renal Function Test reports were normal. For our patient conjunctival recession under local anesthesia was done, 2 clock hours on either side of the ulcer & 4mm posterior to corneoscleral limbus & post-operatively prednisolone acetate 1% every hourly and topical antibiotic were started to prevent secondary infection. On follow up ulcer showed healing with no progression but no improvement in vision due to corneal scarring.

- Fig-1: Right eye on slit lamp with diffuse illumination showing ulcer in inferior margin of cornea
- Fig-2: Right eye on slit lamp with parallelepiped illumination showing undermined ulcer with over hanging edge.
- Fig 3: Left eye on slit lamp examination with diffuse illumination showing ulcer in the inferior margin of cornea.
Fig 4: Left eye on slit lamp examination with parallelepiped illumination showing undermined ulcer with over hanging edge.

Discussion

Mooren’s ulcer is rare conditions mostly seen in adult with no systemic diseases but has also been observed in children [5,6]. Most commonly found in Nigerian young men [7].

Wood and Kaufman have classified ulcers [8].

- Type I is typical or benign Mooren’s ulcer, unilateral with mild to moderate symptoms, responding well to medical and surgical treatment occurring in older patients.

- Type II is atypical or malignant Mooren’s ulcer, bilateral in 75% of cases, with severe pain and more symptoms. It does not respond well to therapy and usually occurring in younger patients [9].

Mooren’s ulcer association with helminth, hepatitis C infection, herpes simplex, zoster, syphilis, tuberculosis, trauma, foreign body, chemical burns, and also after cataract, penetrating keratoplasty and lamellar keratoplasty like surgical procedure [10,11].

Exact pathophysiology of Mooren’s ulcer is unknown but pathological examination of involved area shows plasma cells, neutrophils, mast cells, eosinophils which support the autoimmune theory [12]. Schaap and his colleagues found circulating IgG & IgA antibodies in corneal and conjunctival epithelium cells using indirect immune fluorescent technique [13]. Martin and colleagues suggested that any infection, trauma or systemic disease can cause alteration of corneal antigen [14].

Mooren’s ulcer patient complains of pain which is associated with redness, watering, photophobia and diminution of vision due to corneal opacity, irregular astigmatism and iritis. Ulceration begins from the periphery that is from limbus then it spread towards the center of the cornea. There is involvement of the stroma of cornea which leads to overhanging of edges of the ulcer which is characteristic of Mooren’s ulcer. It may be associated with iritis, glaucoma, cataract rarely ulcer perforates [15]. Mooren’s ulcer can be identified by detailed medical history and various laboratory investigations like complete blood count, erythrocyte sedimentation rate, rheumatoid factor, complement fixation, antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, circulating immune complexes, liver function tests, renal function test, tests to find treponemal infection like VDRL test, urine analysis with microscopy, chest X-ray.

Topical steroids therapy is started with prednisolone acetate or phosphate & topical antibiotic to prevent secondary infection and cycloplegics are also added [16]. This topical therapy can be supplemented with systemic steroids like prednisolone is started if topical steroids are ineffective after 7-10 days, deep infiltrating ulcer [17]. Therapeutic bandaging contact lens will be useful as it prevents perforation.

Eyedrop of Interferon alpha are made by diluting injectable recombinant IFN-2 alpha with preservative-free balanced salt solution. Its concentration is one million international units per ml. It can be given as 2 hourly for first week. Systemic immunosuppressive can also be used, commonly used are cyclophosphamide, methotrexate, azathioprine. Recently cyclosporine, a drug is used, it helps to reduce helper T cell population and it stimulates suppressor T cell which retards the disease progression [18].

Conjunctival recession can also be done under topical or subconjunctival anesthesia. Conjunctival excision is done with bare sclera technique. Superficial lamellar keratectomy, partial penetrating keratoplasty can be done to hold the inflammatory process. Perforation is treated with tissue adhesives.
Case Report

Conjunctival recession with topical corticosteroid and antibiotic eye drops showed healing of the ulcer in our patient which is an age old technique and may even show failure to heal the ulcer. Further studies are required for reliability and effectiveness of new treatments to avoid surgical intervention.

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