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*Case Report*

# Diagnosis And Treatment For Bilateral Fuchs Superficial Marginal Keratitis.

**Shiri Harda,Yaser Hely Mohamd, Mo Kusno, Disuke Inue, Masaumi Umasu.**

*Dipartimento Salute Mentale e Dipendenze SERT Monza, ASST Brianza (MB), Italy.*

**Abstract**

Our investigation found two individuals with bilateral Fuchs' superficial marginal keratitis (FSMK) who had surgical treatment and anti-inflammatory medicines. The cases included bilateral photophopia, hazy vision, and pseudopterygium with normal intraocular pressure (IOP). Pseudopterygia appeared on the temporal and nasal sides, with a gray border between normal corneal epithelium and no lipid deposits. The The first instance had bilateral high mixed astigmatic error, whereas the second had dry eye symptoms.The laboratory tests revealed no anomalies, including systemic inflammatory illness.In the first instance, mitomycin C (MMC) was used, bilateral pseudopterygium excision was performed, and pedicled transplanting conjunctival flap tissue. The patient was monitored on a regular basis, and her eye findings showed no signs of deterioration or recurrence.Over the course of four years, the second case underwent conjunctival excision, amniotic membrane transplantation, and MMC treatment six times on the right eye and three times on the left, with recurrences after each procedure. Her two eyes' high IOP significantly reduced her visual acuity. The patient declined more surgery.

**Keywords** : *Fuchs' superficial marginal keratitis; pseudopteryguim; irregular astigmatism; corneal thinning.*

## INTRODUCTION

Von Arlt initially reported the unusual condition known as Fuchs' superficial marginal keratitis (FSMK) in 1885, and Fuchs provided a more thorough description of it in 1895 [1].The rare disorder known as FSMK is characterized by increasing corneal stromal thinning, marginal corneal infiltrates, and intermittent ocular inflammation. A fictitious pterygium could grow over the thinning of the cornea. In more severe situations, traumatic or even spontaneous perforation may result from severe stromal loss [2]. Usually beginning as a superficial marginal keratitis, this illness of uncertain cause progresses unevenly and nonuniformly over the cornea [3, 4]. The disease has only been described in a small number of accounts in the literature due to its rarity. We describe two patients whose clinical presentations support a diagnosis of bilateral Fuchs' superficial marginal keratitis, which was managed with anti-inflammatory medications and surgery. This research was authorized by Nagasaki University Hospital's Institutional Review Board (permission number 24021928), and informed consent was given by the study's patients.

## CASE REPORT

In April 2022, a 47-year-old Japanese woman with no underlying medical conditions who had binocular blurred vision and photophobia for the previous six years was sent to Nagasaki University Hospital by the ophthalmology clinic. At the initial examination, her right eye's best corrected visual acuity (BCVA) was 0.2 (decimal correction), with a refraction of +11.0 D sphere and −5.0 D axis. 170 cylinder) and 0.4 in the left eye (refraction was −5.0 D axis 170 cylinder, +10.0 D sphere), with both eyes' intraocular pressures (IOPs) being normal.Both eyes featured a gray line between the corneal normal epithelium without any lipid deposits, as well as circumferential corneal infiltrations and pseudopterygia that extended from the temporal and nasal sides .The front In the same region, segment optical coherence tomography (AS-OCT) showed modest corneal thinning and bilateral pseudopterygium. Binocular irregular astigmatism was discovered by corneal elevation topography. The laboratory tests revealed no anomalies, including systemic inflammatory illness. An FSMK diagnosis was made.

***Corresponding Author:** Disuke Inue, Dipartimento Salute Mentale e Dipendenze SERT Monza, ASST Brianza (MB), Italy.
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Bilateral removal of pseudopterygium, administration of mitomycin C (MMC), and transplantation of pedicled conjunctival flaps were carried out in June 2022. Topical steroids and antibiotics are used four times a day for one month following surgery. One month following surgery, topical antibiotics were stopped, but topical corticosteroids, however mild, continued to reduce conjunctival congestion. One month following surgery, cyclosporine was started orally to reduce postoperative inflammation and prevent recurrence. The dose is 175 mg/day, with a trough value of 70–100 ng/mL.

The patient was monitored every two to three months, and up until her most recent appointment in December 2023, there had been no deterioration or recurrence in her ocular results. Her eyes' most recent follow-up photo is displayed in Figure2, and her most recent BCVA was 1.2 on the right. eye (refraction was 1.2 in the left eye, +4.0 D sphere, −2.0 D axis 150). A local ophthalmologist diagnosed a 28-year-old woman with dry eye and pseudopterygium after she had dryness and photophobia in both eyes for a few years.Topical steroids and lubricant-containing eye drops were used to treat her. When she was 29 in November 2011; in order to receive better assessment and care, she was sent to Nagasaki University Hospital. Her IOP was normal in both eyes, and her BCVA was 0.9 in the left eye and 0.6 in the right (refraction −1.25 D sphere). Both palpebral fissures' bulbar conjunctivas displayed hyperemia, and the conjunctival epithelium had circumferentially infiltrated the cornea.

She had no adhesions on her conjunctiva. Her nasal and temporal corneas showed conjunctival invasion, and the lower portions of both corneas showed punctate superficial keratopathy (Figure 3B,F). There were none significant anomalies in the topography of corneal elevation. Both eyes' fundus was normal, the lens was clear, and the anterior chambers were of normal depth. There were no indications of inflammation. Her underlying conditions included Sjögren's syndrome, allergic rhinitis, and obsessive compulsive disorder [for which she was taking selective serotonin reuptake inhibitors, or SSRIs]. Rheumatoid arthritis was not a condition she had. Although she received treatment for both of her punctal-plug eyes four to six times a day with eye drops (lubricants, topical steroids, and tranilast), the conjunctival invasion progressively got worse.

The patient was monitored every two months from 2011 to 2015. She had the Her right and left eyes underwent their initial conjunctival excision, MMC, and amniotic membrane transplants in June and July of 2015, respectively. Following surgery, she began taking oral cyclosporine and prednisolone. On the third day following right eye surgery, oral prednisolone (20 mg/day) was initiated, and four months later, it was reduced. Prednisolone taken orally was utilized in the maintaining the same routine throughout the second and third perioperative phases. Additionally, two weeks following the initial operation on the right eye, 150 mg of oral cyclosporine per day was began. The trough value of cyclosporine is between 70 and 100 ng/mL. Two months following surgery, the conjunctival invasion returned. Conjunctival resection was performed on the patient.+ MMC application + amniotic membrane transplantation + keratoepithelioplasty for the right eye's fourth procedure between the ages of 33 and 37 (2015–2019), six times in the right eye and three times in the left, and experienced recurrences following each procedure. Repeated peripheral corneal ulcers and increasing conjunctival invasion were seen in both eyes.

Her visual acuity had dropped to 0.07 in her left eye and 0.3 in her right eye as of February 2019. Furthermore, both eyes had high intraocular pressure (IOP) (right = 21 mmHg and left = 26 mmHg), and glaucoma eye drops were recommended. In November, during her final visit Her intraocular pressure was 15 mmHg in the right eye and 24 mmHg in the left with medication in 2023, when she was 41 years old, and her visual acuity had further declined to 0.01 in both eyes. At the time of the last visit, she was taking lubricants twice a day, glaucoma eye drops, tacrolimus twice a day, and tranilast four times a day. Figure 5 illustrates the patient's corneas at her most recent visit, which took place in November 2023. The patient refused to have any more surgeries and continued to take the same oral and ocular drops.

## DISCUSSION

A rare condition known as FSMK is typified by increasing marginal thinning of the corneal stroma and episodic ocular inflammation with marginal corneal infiltrates [2]. Young adults between the ages of two and four are most frequently impacted. It frequently has achronic course characterized by frequent episodes of red eye, discomfort, and tears, together with a marginal stromal thinning that has no accompanying lipid deposits and is usually uneven in depth and axial extension, with a fine intraepithelial gray line on its advancing edge [3, 5].The marginal thinning is often linked to a pseudopterygium and lacks a preferred limbal position [4,5]. In more severe forms, irregular astigmatism can impair visual acuity [6].

FSMK was determined to be the most likely diagnosis due to the remarkable resemblance in clinical symptoms between our patients and the previously reported FSMK cases.Terrien's marginal degeneration (TMD), Mooren's ulcer, autoimmune peripheral ulcerative keratitis, and limbal stem cell deficiency (LSCD) are among the conditions that are included in the differential diagnosis of FSMK. Since several blood tests for systemic inflammation and collagen disease came back negative, autoimmune peripheral ulcerative keratitis was ruled out. Painful, increasing corneal peripheral ulceration

(epithelial defect stained with Mooren's ulcer) was the initial sign of the condition.fluorescein), which is linked to severe edema and inflammation of the limba, was not the same as our patients.

Both eyes are affected by the uncommon, slowly developing corneal disorders TMD and FSMK, which have no recognized cause. A number of clinical symptoms are similar across them. Peripheral corneal thinning, which usually starts in the upper half of the cornea but can occasionally start in thelower portion [7]. Early biomicroscopic analysis of FSMK typically shows peripheral corneal thinning in the lower nasal segments.The primary pathological symptom in both conditions is peripheral corneal thinning, although the lesions in the two eyes are typically asymmetrical [7]. A rare illness called FSMK is similar to TMD in a number of ways.to the degree that they might be regarded as distinct symptoms of the same degenerative marginal corneal disease, the cause of which is unknown [7].

Young and middle-aged individuals are the main demographics affected by TMD and FSMK. Both of them entail paralimbal stromal thinning, which usually manifests asymmetrically but bilaterally. Although any condition can cause pseudo-pterygium, it is more traditionally connected to FSMK. Furthermore, either traumatic or spontaneous corneal perforation may result from either disease [5,8]. Some scientists have proposed that both diseases are symptoms of the same disease because of these similarities [5,7].

In contrast to TMD, FSMK affects the limbus anywhere (with no preference for the superior cornea), has concomitant epithelial detects, has a gray line that delineates the epithelium, and does not possess lipid buildup [3, 5]. LSCD can be distinguished from FSMK by its underlying etiology, such as chemical or mechanical trauma or other known factors that contribute to LSCD [8]. Conjunctivization is the result of FSMK's gradual destruction of corneal limbal stem cells.It is impossible to rule out the possibility that this uncommon illness is the cause of some LSCDs with an unclear etiology. Until additional cases are documented, it will be impossible to ascertain whether FSMK plays a role in LSCDs.

One risk factor for both spontaneous and traumatic perforation is progressive stromal thinning [3,9]. Determining the extent of thinning due to concomitant pseudopseudopterygium may be challenging. Because perforation can happen by accident during surgical intervention, caution must be used [9]. It has been observed that the illness can persist even after Lamellar keratoplasty, even though it might be outside the graft's edge [2,10]. Because it will enable the differentiation of the pseudopterygia tissue from the underlying thinning corneal tissue, AS-OCT could be a very useful preoperative technique. While topography can quantify the total thickness of corneal tissue and pseudopterygia, AS-OCT will provide distinction based on cross-sectional appearance. According to Cheung et al., the nasal portion of the cornea in their FSMK instance was the thinnest, accounting for 34% of the thickness of thecentralcornea.Nonetheless, they proposed that the hole may potentially occur in the superonasal quadrant, which is the narrowest region. Remarkably, the mid-periphery showed the most thinning in the AS-OCT pictures [10]. AS-OCT allowed for a more accurate measurement of corneal thickness. precisely in the afflicted region, which is crucial for selecting therapeutic approaches [7].

A curved gray ring distinguishes the center cornea from the marginal superficial limbal corneal deterioration and ulcers [3]. Until the condition progresses and visual acuity declines, the central cornea is usually clean.This is a chronic condition that causes processive circumferential peripheral corneal thinning, which can result in perforation, and periods of remission and relapse [9]. Areas of corneal thinning are typically where pseudopseudopterygia grow [4].

Ellis documented the disease's histology and examined two examples of bilateral FSMK. In the substantia propria of the cornea of one eye, epithelioid large cells were seen. The area with the highest concentration of acute inflammatory cells was beneath the ulcerated regions [11]. Pterygia-like degeneration of the corneal limbal and conjunctival regions was seen [11].

Over a few of years, the pseudopterygium progressively spreads over the cornea while avoiding the core cornea. In two of three instances, Bierly et al. reported notable thinning beneath the pseudopterygium. As a result, during pseudopterygium, the cornea was perforated.After forceful trauma in one instance and excision in another. These issues emphasize the need for extra care when caring for these patients [3]. Over a few of years, the pseudopterygium progressively spreads over the cornea while avoiding the core cornea. In two of three instances, Bierly et al. reported notable thinning beneath the pseudopterygium. As a result, during pseudopterygium, the cornea was perforated.After forceful trauma in one instance and excision in another. These issues emphasize the need for extra care when caring for these patients [3].Treatment for FSMK is based on its stage. Lubricants [4], topical steroids [4,5], oral doxycycline [5], and vitamin C have all been shown to alleviate symptoms during the acute period [5,6]. Instillations are effective in controlling the active inflammatory process. A perilimbal conjunctivotomy, which involves separating the conjunctiva around the whole circumference of the eye, was done if symptoms of excruciating eye pain, severe corneal syndrome, and the failure of conservative treatment continued.the damaged eye's limbus [7].

It's possible that long-term topical corticosteroid therapy helped down the disease's progression. Early application of such therapy may help slow the progression of the disease, even if long-term anti-inflammatory drug treatment for

FSMK and TMD is typically not used unless there are clinical symptoms of ocular inflammation [5].

Scleral contact lenses are an option for people with developed reverse astigmatism, and they can greatly enhance visual acuity. Rarely, perforation is observed in patients with TMD and FSMK who have had little to no trauma; the likelihood of this is around 15%, in addition to corneal edema, the development of corneoscleral or intracorneal cysts, and the partial or complete separation of Descemet's membrane [7].

When it comes to corneal microperforations, cyanoacrylate is temporarily helpful. Penetrating keratoplasty [3], lamellar keratoplasty [2], amniotic membrane repair [4], superficial keratectomy with conjunctival autograft [4,5], and other procedures are used in a few case reports.Graft of the corneoscleral lamellar patch [6]. Keenan et al. emphasized the challenge of managing FSMK-related peripheral corneal thinning that progresses gradually. Despite using vitamin C and doxycycline for treatment, they noticed increasing thinning [5]. Due to in vitro research demonstrating that doxycycline inhibits matrix metalloproteinase, this therapy strategy has potential advantages.Collagen production is improved by ascorbate and activity [12,13].

In FSMK, pseudorrhea may worsen to the point that it endangers visual acuity. As long as patients are thoroughly monitored in the early postoperative phase to detect corneal infiltrates, surgical excision can be both safe and beneficial in improving eyesight [7].In order to try to delay the formation of recurrent pseudopterygia using the same mechanism that inhibits recurrence following the removal of typical pterygia, recommended surgical treatment may involve combining superficial keratectomy with conjunctival autograft or amniotic membrane transplantation [4,14].According to Kotecha and Raber, this medication seems to reduce marginal keratitis flare-ups. Considering the potential contribution of the conjunctiva to inflammation, a conjunctival autograft combined with a lamellar patch graft has been suggested as well [4].

Even several surgical procedures and long-term oral cyclosporin use were unable to treat severe cases, such as our second patient. But in the first instance, surgery was done before the conjunctival invasion became extensive. Cyclosporin used orally was given right after surgery, and for one and a half years after the operation, there was no discernible decline in the corneal condition. Our examples illustrated the two extremes of the illness and how they react to therapy, making it challenging to forecast how well a treatment will work. Strict inflammation control and surgical intervention from the beginning of the disease may be able to slow the progression of FSMK.

## CONCLUSIONS

In two of our cases, we found pseudopterygia with refractory peripheral corneal infiltration in both eyes, which raised the possibility of FSMK. Even though FSMK is uncommon, it should be taken into account if both eyes exhibit pseudoopterygia, corneal thinning, and peripheral corneal infiltration. Despite the difficulty of treating this illness,Prompt surgical and anti-inflammatory measures could aid in the disease's management.

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### Institutional Review Board Statement

The Institutional Review Board of Nagasaki University Hospital granted approval for this study, and its approval number is 24021928).

### Informed Consent Statement

Since no personal information was disclosed and the clinical data was gathered from medical records, the Institutional Review Board at Nagasaki University Hospital did not require informed consent.

### Data Availability Statement

Upon reasonable request, the corresponding author will provide the datasets used in this study.

### Conflicts of Interest

No conflicts of interest are disclosed by the writers.

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