

Sterile corneal perforation after cataract surgery in Sjögren's syndrome

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SUMMARY Painless, sterile, noninfiltrated corneal ulceration and perforation, which may occur after recent cataract surgery in patients with rheumatoid arthritis and Sjögren's syndrome, appears to be a distinct clinical entity. The cause is probably multifactorial. Contributing factors may be the underlying systemic disease process in rheumatoid arthritis and Sjögren's syndrome, the associated keratoconjunctivitis sicca, the surgical procedure, and the postoperative use of oral and topical corticosteroids. Two cases of this clinical entity are reported here. Preoperative recognition by cataract surgeons that corneas in patients with rheumatoid arthritis and Sjögren's syndrome have an increased risk for postoperative ulceration should prompt postoperative preventive measures.

Sterile corneal thinning, ulceration, and perforation in patients with rheumatoid arthritis is a well-defined entity.¹⁻⁷ These documented corneal abnormalities have not been associated with ocular surgery. I am aware of only one reported case of central, sterile corneal perforation after recent routine cataract surgery in a patient with rheumatoid arthritis and Sjögren's syndrome.⁸ I am reporting 2 additional patients to support my contention that postsurgical, painless, central, noninfiltrated, and sterile corneal ulceration and perforation in such patients is an important clinical entity worthy of definition. The multiple contributing factors which lead to this condition will be evaluated and recommendations offered for postoperative preventive measures.

Case reports

CASE 1

A 71-year-old white female with a history of severe rheumatoid arthritis since 1958 has been followed up by her ophthalmologist since 1966 for keratoconjunctivitis sicca, progressive cataracts, and bilateral senile macular degeneration. Therapy has been multiple artificial tear preparations. A diagnosis of Felty's syndrome was made in 1978, and treatment with oral prednisone was started. A splenectomy was performed on 31 October 1979.

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In July 1979 the best corrected visual acuity was 6/15 in the right eye and 6/24 in the left eye. There were hyperaemia and crusting of all lid margins. Biomicroscopic examination showed punctate epithelial erosions of the inferior one-third of each cornea, and both lenses showed moderate nuclear sclerosis. A Schirmer I test showed 1 mm of wetting in 5 minutes in each eye. Goldmann applanation intraocular pressures were 10 mm Hg in each eye. In September 1979 visual acuity had decreased to 6/30 in each eye because of progressive lenticular nuclear sclerosis.

On 31 March 1980 the patient was treated with oral acetazolamide and topical 2% pilocarpine hydrochloride for an attack of acute angle-closure glaucoma in the right eye. On 1 April 1980 the intraocular pressure was normal and the acetazolamide was stopped. Progression of the right cataract decreased the visual acuity to 6/60.

On 3 May 1980 the patient had another attack of acute angle-closure glaucoma in the right eye and was immediately admitted to hospital. Treatment with intravenous 20% mannitol was begun, and the glaucoma subsided within 12 hours. On 6 May 1980 an uncomplicated right intracapsular cataract extraction and a sector iridectomy were performed.

Postoperative treatment was atropine sulphate 1% ophthalmic solution every other day and Maxitrol (neomycin sulphate, polymyxin B sulphate, and dexamethasone) ophthalmic ointment twice a day. Artificial tear drops were also continued. The intraocular pressure remained normal, and at a post-

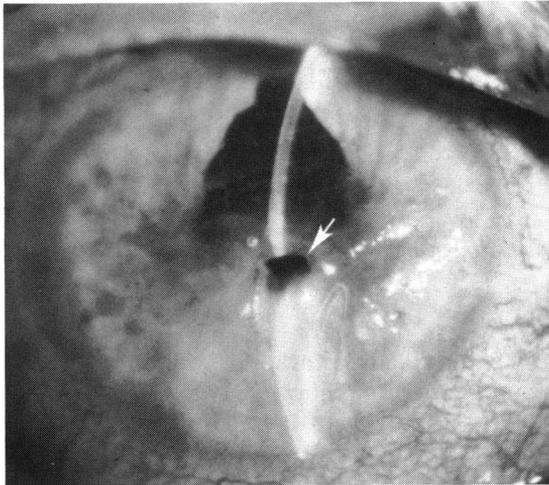


Fig. 1 The right cornea had a sterile paracentral perforation (arrow) through which the vitreous had prolapsed.

operative examination on 15 May 1980 the right cornea was normal. At the next routinely scheduled postoperative examination, on 28 May 1980, a painless, inferior, paracentral, right corneal perforation was observed. The patient was immediately referred to the Ophthalmology Clinic at North Carolina Memorial Hospital, Chapel Hill, North Carolina.

On admission the oral medication included 650 mg of aspirin 3 times daily, 5 mg of prednisone daily, and 25 mg of indomethacin 3 times daily. The visual acuity in the right eye with a +10.00 spherical lens was counting fingers at 2 feet (60 cm) and in the left eye was correctable to 6/30. Biomicroscopic examination of both eyes showed absent tear menisci. The right cornea had a slightly inferior paracentral perforation 1.5 mm in diameter without any surrounding infiltrate (Fig. 1). The vitreous had prolapsed through the perforation, the anterior chamber was shallow, and the eye was soft. There were punctate epithelial erosions of the inferior one-third of the left cornea. The peripheral anterior chamber was narrow, and the lens showed moderate nuclear sclerosis. Goldman applanation intraocular pressure in the left eye was 15 mm hg. Indirect ophthalmoscopy revealed only hazy details of the right optic nerve, macula, and retinal vessels. A normal optic disc, peripapillary retinal atrophy, and pigmentary dispersion in the macula were seen in the left eye.

On 29 May 1980 a corneal patch graft with isobutylcyanoacrylate tissue adhesive and a closed eye, and partial anterior vitrectomy with the Ocutome instrument in the right eye, were successfully performed. Corneal scrapings performed at the time of surgery for anaerobic and aerobic bacterial and fungal cultures resulted in no growth.

CASE 2

A 74-year-old white female with a 15-year history of severe rheumatoid arthritis has been followed up by her ophthalmologist for recurrent keratoconjunctivitis and progressive cataracts since 1972. When she was first examined the best corrected visual acuity was 6/6 in the right eye and 6/7.5 in the left eye.

In September 1979, because of a complaint of decreased vision, the patient was examined. The best corrected visual acuity in the right eye was 6/30 and in the left eye was 6/15. Both lenses were cataractous and accounted for the decreased visual acuities. A Schirmer I test measured 3 mm of wetting in 5 minutes in each eye. Goldmann applanation intraocular pressures were 12 mmHg in the right eye and 14 mmHg in the left eye. A diagnosis of keratoconjunctivitis sicca was made, and treatment with frequent Hypotears (artificial tears) drops was begun.

In June 1980 the best corrected visual acuity was again 6/30 in the right eye and 6/15 in the left eye. The patient was advised to have cataract surgery, and on July 31 1980 an uncomplicated, right intracapsular cataract extraction and a peripheral iridectomy were performed.

Postoperative treatment was atropine sulphate 1% ophthalmic solution daily and Maxitrol ophthalmic solution 3 times daily. The Hypotears drops were continued. Examination on 12 September 1980 showed no thinning or ulceration of the right cornea. At the next routinely scheduled postoperative examination on 29 September 1980 a painless, inferior, paracentral, right corneal perforation was observed. The patient was immediately referred to the Ophthalmology Clinic at North Carolina Memorial Hospital, Chapel Hill, North Carolina.

On admission the oral medications included 650 mg of aspirin daily.

The visual acuity in the right eye with a +10.00 spherical lens was 6/120 and in the left eye was correctable to 6/30. Biomicroscopic examination of both eyes showed absent tear menisci. The right cornea had a slightly inferior paracentral perforation 1 mm in diameter. There was a small area of surrounding stromal oedema without any infiltrate, and the vitreous had prolapsed through the perforation. The anterior chamber was deep, and the eye was soft. There were punctate epithelial erosions and filaments of the inferior one-third on the left cornea, the anterior chamber was of normal depth, and the lens showed moderate nuclear sclerosis. Goldman applanation intraocular pressure in the left eye was 15 mmHg. Indirect ophthalmoscopy of each eye revealed a clear vitreous, normal optic nerves, peripapillary retinal atrophy, and pigmentary dispersion in the maculae.

On 30 September 1980 a corneal patchgraft with

isobutylcyanoacrylate tissue adhesive and a closed eye, and partial anterior vitrectomy with the Ocutome instrument in the right eye, were successfully performed. Corneal scrapings performed at the time of surgery for anaerobic and aerobic bacterial and fungal cultures resulted in no growth.

Discussion

The clinical entity of sterile, noninfiltrated, central and peripheral corneal thinning, ulceration, and perforation in patients with rheumatoid arthritis who have not undergone ocular surgery has been well defined.¹⁻⁷ Keratoconjunctivitis sicca or an alteration of the tear constituents has been suggested as one possible cause.^{2,6,7} However, keratoconjunctivitis sicca does not appear to be a necessary requirement to produce these corneal changes, since corneal thinning, ulceration, and perforation have been seen in patients with rheumatoid arthritis and no keratoconjunctivitis sicca.² Furthermore, Krachmer and Liabson did not see corneal perforations in patients who had only keratoconjunctivitis sicca.⁷ It may be postulated that the systemic immunological and pathophysiological abnormalities associated with rheumatoid arthritis sometimes cause sterile corneal ulceration in these patients.⁹

The relationship of topical and/or corticosteroids to the corneal ulceration which can occur in patients with rheumatoid arthritis is an unclear but important issue. There has been a frequent but not invariable association between topical corticosteroid therapy and corneal ulceration. In the largest reported series evaluating the treatment of corneal ulceration and perforation in patients with rheumatoid arthritis and Sjögren's syndrome, Pfister and Murphey reported that 9 of 18 eyes had received topical corticosteroids prior to the development of the ulceration or perforation.⁸ Another study of patients with rheumatoid arthritis described 3 patients who developed corneal ulcerations after a topical corticosteroid was begun and one patient who developed a central corneal perforation while being treated with an oral corticosteroid.² Topical corticosteroids are thought to suppress the reparative corneal wound healing process by reducing new corneal collagen synthesis.¹⁰ Thus, if associated keratoconjunctivitis sicca and/or the systemic immunological and pathophysiological abnormalities found in patients with rheumatoid arthritis initiate corneal ulceration, the use of topical and/or oral corticosteroids may potentiate this existing ulceration and result in corneal perforation.

The systemic pathophysiology of rheumatoid arthritis, the frequently associated keratoconjunctivitis sicca, and the use of topical and/or oral corticosteroids all increase the risk of sterile, corneal

ulceration and perforation in patients with rheumatoid arthritis. Cataract surgery may provide several additional risk factors for the occurrence of these severe corneal complications in patients with rheumatoid arthritis. These risk factors are important to the cataract surgeon.

Sterile corneal ulceration and perforation after recent cataract surgery in patients with rheumatoid arthritis and Sjögren's syndrome does appear to be a distinct clinical entity. In the patient previously reported by Pfister and Murphey, who developed postoperative corneal ulceration and perforation, the site of involvement was the inferior, paracentral cornea, and the perforation occurred 6 weeks after surgery.⁸ There are several important similarities between this patient and the 2 patients reported in this paper. All 3 patients (1) had corneal perforations within 3 to 8 weeks after cataract surgery, (2) used postoperative topical corticosteroids, (3) had non-infiltrated corneas, (4) had perforations located in the inferior, paracentral cornea, and (5) had sterile corneal cultures for bacteria and fungi.

It should also be noted that these postoperative corneal perforations are painless and can occur without warning. The 2 patients reported in this paper had no postoperative corneal abnormalities noted at the preceding examinations (9 and 43 days after surgery respectively) prior to the perforation. At the next routinely scheduled examinations, 11 and 17 days later respectively, the corneal perforations were noted.

There are several potential postoperative risk factors that may promote and/or potentiate these painless, noninfiltrated, central corneal ulcerations. It may be postulated that surgery can aggravate the underlying systemic pathophysiological process associated with rheumatoid arthritis, which can then cause these corneas to ulcerate. Bloomfield *et al.* suggested that systemic vasculitis may be a possible cause of corneal abnormalities after cataract surgery.¹¹ They reported a patient with a diffuse vasculitis who developed painful, infiltrated, sterile, peripheral corneal ulcerations after cataract surgery. Immunopathological studies demonstrated immunoglobulins and complement in the walls of the conjunctival blood vessels. These corneal changes were similar to previously reported marginal corneal ulcerations after recent cataract surgery, and one of these patients had a positive serum latex test.^{12,13} Patients with rheumatoid arthritis can have a vasculitis.⁹ Bloomfield *et al.* further suggested that because patients with rheumatoid arthritis can have circulating immunocomplexes they may be at special risk for scleral and corneal melting after cataract surgery. Although the clinical description of Bloomfield *et al.*'s patient is different from Pfister and Murphey's patient and the 2 patients

in this report, the underlying pathophysiology for corneal ulcerations in these patients with rheumatoid arthritis may be similar.

Another postoperative risk factor appears to be topical corticosteroids which are routinely administered after cataract surgery. One of the patients reported in this paper was receiving not only a postoperative topical corticosteroid but also an oral corticosteroid for the treatment of her arthritis. In the past she also received an oral corticosteroid for the treatment of Felty's syndrome. Patients with rheumatoid arthritis are frequently receiving oral corticosteroids for the treatment of arthritis and disease-associated medical complications.

The effects of keratitis sicca may be potentiated postoperatively, since surgery may interfere with lid mobility, and postoperative corneal denervation may reduce blinking. Radtke *et al.* reported 5 patients with only preoperative keratoconjunctivitis sicca who had sterile, noninfiltrated, paracentral corneal ulceration after cataract surgery.¹⁴

Patients with rheumatoid arthritis and Sjögren's syndrome have corneas which can ulcerate and perforate without the added stress of cataract surgery. The recognition by cataract surgeons that the corneas of these patients have added risks for postoperative sterile ulceration and perforation should prompt preventive measures. The routine postoperative administration of topical and oral corticosteroids should be minimised or avoided, and intensive treatment of keratitis sicca should be instituted. Complete blinking can be encouraged, and incomplete lid closure should be corrected.

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