Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.elsevier.com/locate/survophthal

Clinical pathologic reviews

The spectrum of postoperative scleral necrosis

Rishi R. Doshi, MD[a], *George J. Harocopos, MD*[b], *Ivan R. Schwab, MD, FACS*[c], *Emmett T. Cunningham Jr, MD, PhD, MPH*[a,d,*]

[a] *The Department of Ophthalmology, California Pacific Medical Center, San Francisco, California, USA*
[b] *The Departments of Ophthalmology & Visual Sciences and of Pathology & Immunology, Washington University, St. Louis, Missouri, USA*
[c] *The Department of Ophthalmology & Vision Science, University of California—Davis, Sacramento, California, USA*
[d] *The Department of Ophthalmology, Stanford University School of Medicine, Stanford, California, USA*

ARTICLE INFO

Article history:
Received 12 April 2012
Received in revised form
30 October 2012
Accepted 1 November 2012
Stefen Seregard and Milton Boniuk, Editors

keywords:
surgically induced
necrotizing scleritis
scleral necrosis
fungal scleritis
pterygium
beta-radiation

ABSTRACT

An otherwise healthy 62-year-old woman developed necrotizing scleritis 23 years following pterygium excision with adjunctive beta-radiation. Surgically induced necrotizing scleritis (SINS) was diagnosed, but the scleritis progressed despite anti-inflammatory therapy, and 10 weeks after presentation the patient developed a hypopyon and decreased vision. After cultures revealed no growth at 72 hours, immunosuppressive therapy was escalated, with a subsequent deterioration in the patient's clinical course. *Scedosporium* superinfection was eventually cultured and found on histological examination of the enucleated globe. In reported cases, infectious scleral necrosis occurs most commonly following pterygium (71.4%) and scleral buckling (97.2%) surgery. Hypopyon is uncommon (10.0%) in patients with postoperative scleral necrosis, but when present is a strong predictor of infection (odds ratio, 21.2; 95% confidence interval, 2.9–157.5). Rates of underlying autoimmune disease are generally low (0.0–12.5%) except following cataract and lens procedures, where the occurence of SINS heralds systemic illness in 42.9% of cases.

Necrotizing scleritis, a recognized manifestation of collagen vascular disease, may also occur after ocular surgery, an entity known as surgically induced necrotizing scleritis (SINS). Characterized histologically by granulomatous inflammation with fibrinoid necrosis within the sclera,[99] necrotizing scleritis is diagnosed clinically when ischemia or vascular nonperfusion accompanies scleritis with or without active inflammation. The sclera may appear porcelain white due to vascular closure or violaceous due to thinning with uveal show. Since the 1970s, SINS has been reported following various types of ocular surgery, but no standard definition exists. Importantly, the relationship of SINS to the type of surgery, adjunctive treatments, underlying autoimmune disease, and the presence or absence of postoperative infection is not well established. We report a patient who developed scleral necrosis many years following pterygium excision with adjunctive beta-radiation. Our patient subsequently developed a fungal superinfection that was heralded by the appearance of hypopyon. We review and analyse the existing literature on postoperative scleral necrosis.

* Corresponding author: Dr. Emmett T. Cunningham Jr, MD, PhD, MPH, West Coast Retina Medical Group, 185 Berry Street, Lobby 2, Suite 130, San Francisco, CA 94107, USA. Telephone #: 415 972 4600; FAX #: 415 975 0999.
E-mail address: emmett_cunningham@yahoo.com (E.T. Cunningham).
0039-6257/$ – see front matter
http://dx.doi.org/10.1016/j.survophthal.2012.11.002

1. Case report

1.1. Clinical history

A 62-year-old Asian woman presented with the acute onset of severe pain and redness in her right eye. She had had a splenectomy following a motor vehicle accident over 30 years ago. She had pterygium surgery with adjunctive strontium irradiation in the right eye 23 years ago. Visual acuity at presentation was 20/16-2 OD and 20/20 + 1 OS. Examination revealed diffuse anterior scleritis in the right eye, most notable superiorly and inferiorly, with a 4 × 4 mm area of avascular necrosis nasally (Fig. 1). No inflammatory cells were present in the anterior chamber or vitreous. Treatment was initiated with topical 1% prednisolone acetate, 0.5% moxifloxacin, and 1% atropine, as well as oral ibuprofen 400 mg daily. Two weeks later, symptoms had not improved, and oral prednisone, 60 mg daily, and levofloxacin, 500 mg daily, were added. Despite initial control on this regimen, symptoms recrudesced 3 weeks later, and the prednisone dose was increased to 80 mg daily. Corticosteroid-sparing treatment with methotrexate 15 mg weekly was initiated, and an extensive laboratory workup for underlying autoimmune conditions was unremarkable. A uveitis subspecialist recommended an increase in her methotrexate dosage to 20–25 mg daily and the addition of a TNF-alpha inhibitor to her immunosuppressive regimen; problems with insurance authorization delayed treatment, however.

Ten weeks after initial presentation, the patient awoke with severe visual loss in her right eye, accompanied by an increase in pain. Examination revealed hand-motion vision in the right eye and a hypopyon (Fig. 2). A vitreous tap with injection of vancomycin and ceftazidime was performed, with no growth on culture of the vitreous fluid. Cyclophosphamide 50 mg daily was initiated, but despite these interventions, vision rapidly declined to light perception as the hypopyon increased in size (Fig. 3). The patient was given an intramuscular injection of adalimumab and hospitalized for intravenous immunosuppression with cyclophosphamide and methylprednisolone. A pre-admission conjunctival culture demonstrated mold, prompting the addition of oral voriconazole 200 mg twice daily for antifungal coverage. The base of the necrotic ulcer was scraped and eventually grew *Scedosporium* (unspeciated) on all culture plates. Hourly topical voriconazole was added to the patient's regimen, but her condition continued to deteriorate. Despite vitrectomy, surgical debridement, and placement of a scleral patch graft, the infection progressed, and the globe was eventually enucleated.

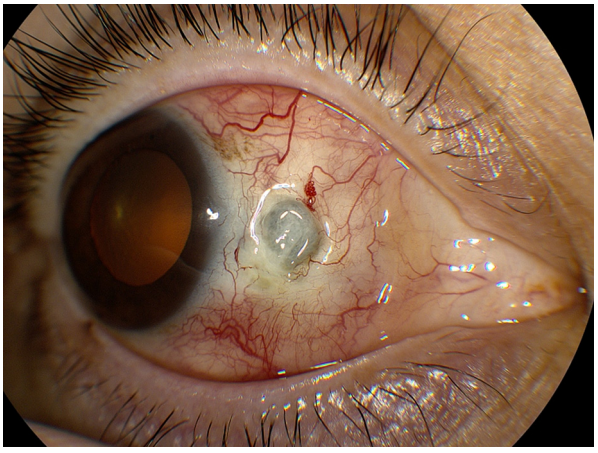

Fig. 1 – Diffuse anterior scleritis with a 4 × 4 mm area of avascular scleral necrosis and thinning nasally.

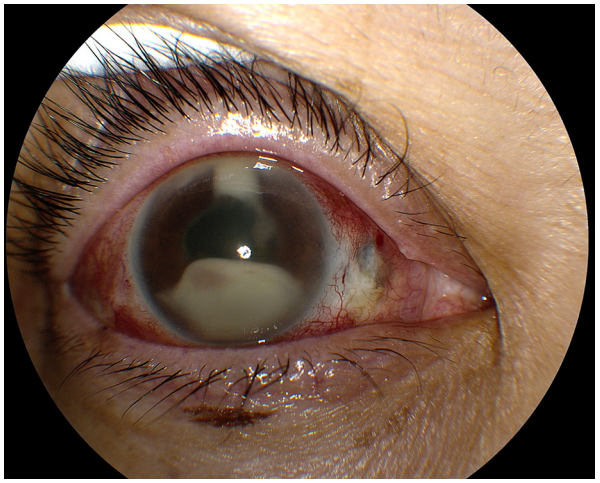

Fig. 2 – Hypopyon development 2.5 months following presentation of necrotizing scleritis.

1.2. Pathologic findings

Histology of the enucleated globe (Fig. 4) revealed granulomatous scleritis, which was most prominent nasally in the vicinity of the scleral patch graft. There was also perforated ulcerative sclerokeratitis on the opposite side at the limbus, with inflamed iris drawn toward the perforation site. A residual vitreous abscess was present, containing

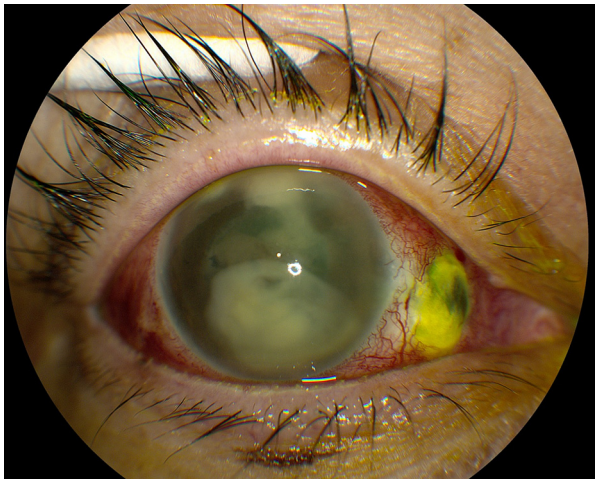

Fig. 3 – Increase in size of hypopyon with accompanying decline in vision to light perception, despite vitreous tap and injection of vancomycin and ceftazidime.