A Case Report on Scleromalacia Perforans

Suneetha Nithyanandam[1], Shubashree Karat[1], Lizbeth Renny[2], Neelima Mathew[2*]

[1]Department of Ophthalmology, St. John's Medical College, Sarjapur Road, Bangalore 560034, India
[2]Department of Ophthalmology, St. John's Medical College, Rajiv Gandhi University of Health Sciences, Bangalore, India

ABSTRACT

Scleromalacia perforans is a bilateral severe form of necrotizing scleritis which is potentially serious and may lead to blindness. This condition is known to be associated with Rheumatoid arthritis in the majority of the patients. A delay in diagnosis and treatment can lead to serious ocular and systemic complications. We report a case of scleromalacia perforans with scleral perforation as the presenting sign of rheumatoid arthritis and its management and outcome.

KEYWORDS
Scleromalacia perforans, Rheumatoid arthritis, Scleral perforation, Necrotizing scleritis, Scleral perforation

* *Corresponding Author: Neelima Mathew, Department of Ophthalmology, St. John's Medical College, Sarjapur Road, Bangalore 560034, India;*
E-mail: neelimathew@gmail.com

How to Cite This Article:

Nithyanandam S, Karat S, Renny L, et al. A Case Report on Scleromalacia Perforans. J Evid Based Med Healthc 2025;12(1):133.

Received: 23-Oct-2023;
Manuscript No: JEBMH-23-118063;
Editor assigned: 26-Oct-2023;
PreQC No. JEBMH-23-118063 (PQ);
Reviewed: 09-Nov-2023;
QC No. JEBMH-23-118063;
Revised: 03-January-2025;
Manuscript No. JEBMH-23-118063 (R);
Published: 10-January-2025;
DOI: 10.18410/jebmh/2024/12/01/133

INTRODUCTION

Scleritis is the inflammation of episcleral and scleral tissues. It is categorized as anterior and posterior scleritis, with the anterior form being more prevalent. The rare, but most serious type of scleritis is necrotizing scleritis, a condition that is frequently linked to underlying systemic collagen vascular diseases, most common being rheumatoid arthritis.

Necrotizing scleritis with absent or minimal signs and symptoms of inflammation is known as scleromalacia perforans. Scleral perforation may result rarely in this variant. We report the presentation and management of a patient who presented with scleromalacia perforans and then diagnosed to have rheumatoid arthritis.

CASE PRESENTATION

A 47-year-old female presented with complaints of pain, redness, and swelling in OD for 4 months, for which she was being treated in another hospital with topical steroids. She presented to our hospital with a history of joint pains in both small and large joints and worsening ocular symptoms in OD.

Examination revealed a best-corrected visual acuity of 6/18 in OU. Slit lamp examination of the right eye revealed uveal tissue covered with thin layer of conjunctiva and thinned-out sclera surrounding the defect, measuring 7 mm vertically and 6 mm horizontally with dilated and tortuous episcleral vessels inferiorly, 3 mm from the superior limbus, (Figure 1A). Bluish discolouration of the sclera was noted nasally in the same eye and similar thinning noted in the left eye supero-temporally (Figure 1B). She had early posterior subcapsular cataract in OU, schirmer's value was 6mm in OU and intraocular pressure of 16 mmHg OU.

A diagnosis of scleromalacia perforans with right eye scleral perforation was made and the patient was worked up for systemic diseases. Complete blood count was within normal limits, CRP was 3.39, Anti-CCP was 390.80, RA factor was 316.04.

A diagnosis of rheumatoid arthritis was made. Patient received topical steroids, antibiotic, tear substitutes and cycloplegics. She was also started on systemic steroids and underwent scleral patch graft for OD the following day. The immediate postoperative period was uneventful (Figure 1C). On follow-up examination after 6 months, the graft was well taken and secure without any evidence of recurrence (Figure 1D).

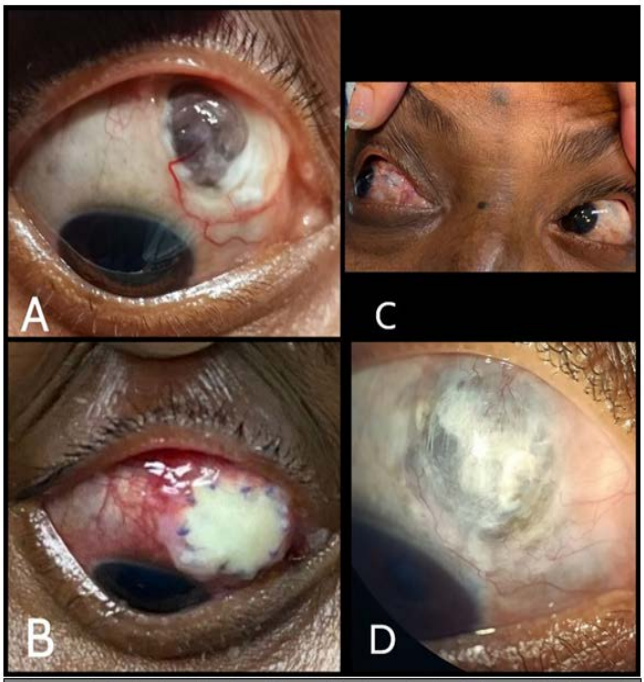

Figure 1. (A) Scleral Perforation with Uveal prolapse in the Right Eye; (B) Bilateral Scleral Thinning; (C) Post-operative Photograph of Same Patient after Scleral Patch Graft; (D) At 6 Months Follow-up

DISCUSSION

Scleritis refers to a range of conditions characterized by inflammation of the sclera that may also affect the cornea, episclera, and underlying uvea with severe pain and redness of the eye.[1] It is one of the differential diagnoses for a patient with red eye. Scleritis can be divided into anterior and posterior subtypes, with the ora serrata acting as an arbitrary line dividing them. Anterior scleritis is further classified into necrotising and non-necrotizing scleritis, non-necrotising variant further subdivided into focal and diffuse types and necrotizing scleritis, further classified into with and without inflammation (scleromalacia perforans). Necrotizing scleritis can perforate the globe in the absence of prompt and appropriate treatment.[2]

Necrotizing scleritis is the most severe and destructive type of scleritis, and can occasionally result in globe perforation or even loss of the eye. If untreated, tissue loss develops, and a necrotic slough or sequestrum gradually forms over the perforation, leaving the choroid exposed or with a thin layer of conjunctiva covering it. A thin fibrous tissue may grow over the defect, if the defect is minor. Nevertheless, scleral grafting is preferred in conjunction with systemic immunosuppressives when the defect is large. The most common systemic disease associated with scleritis is rheumatoid arthritis. The most common variety of scleritis is diffuse anterior, however rheumatoid arthritis is known to be associated with other types of scleritis.

Wegener's granulomatosis, tuberculosis, systemic lupus erythematosus, juvenile rheumatoid arthritis, polyarteritis nodosa, relapsing polychondritis, psoriasis, gout, atopy, rosacea, syphilis, herpes simplex and zoster infections are known diseases associated with scleritis.[3]

Blood work-up to rule out these systemic disease include rheumatoid factor for RA, ANAs for SLE and associated disorders, ANCA for microscopic polyarteritis nodosa and Wegener's granulomatosis.[4,5]

Scleritis may cause serious consequences if not appropriately managed. Peripheral corneal thinning, stromal keratitis, and peripheral ulcerative keratitis are associated with scleritis. Extension of inflammation into the corneal stroma may cause sclerocornea.[6] Other ocular complications are exudative retinal detachment, keratitis, uveitis, glaucoma, and macular oedema. The appearance of uveitis and glaucoma indicate spread of inflammation to intraocular structures that could result in progressive vision loss.

Mainstay of treatment is systemic steroids at a dose of 1 mg/kg/day and tapered slowly. Topical steroids can be given to alleviate symptoms. Severe ocular lesions with and without systemic vasculitis require immunosuppressives. The first line of treatment is with methotrexate 7.5 mg once a week or 2 mg/kg/day of azathioprine is recommended if the condition is severe but rapidly progressing. Cyclosporin 5 mg/kg/day may be used as an alternative. Cyclophosphamide, at a dose of 2 mg/kg/day with high fluid intake and thorough monitoring, is the therapy of choice if the patient doesn't respond to these medications or has severe necrotizing scleritis. TNF-alpha is one of the key mediators of inflammation and triggers MMPs, which makes biologic TNF-blocking medications like infliximab and adalimumab desirable alternatives for treating scleritis.

Reinforcement with donor sclera, fascia lata, periosteum, or synthetic materials is necessary in cases of severe corneal thinning or perforation. Severe keratolysis or marginal corneal ulcer may call for corneal grafting, typically in the form of a lamellar patch transplant.[7]

A similar case report by Yangtze et al., emphasizes the need for prompt treatment with immunosuppressives to prevent ocular and systemic complications.[8]

CONCLUSION

In conclusion scleritis is a known ocular manifestation of collagen vascular diseases, however rarely it may be the first sign of a potentially severe systemic disease. Management of underlying systemic disease and prompt treatment of scleritis can mitigate the risk of vision loss. An early diagnosis and treatment of scleritis can not only protect the eye but also extend the patient's life.

REFERENCES

1. Valenzuela FA, Perez VL. Scleritis-Infectious Versus Inflammatory. US Ophthalmic Rev. 2016;9(2):92–96.
2. Majumder PD, Agrawaly R, McCluskeyz P, et al. Current approach for the diagnosis and management of noninfective scleritis. Asia Pac J Ophthalmol. 2021;10(2):212-223.
3. Watson PG. The nature and treatment of scleral inflammation. Trans Ophthal Soc UK. 1982;102:257-281.
4. Reichlin M. Antinuclear antibodies. Textbook of Rheumatology, 3rd Edition. W.B. Saunders Company Publisher. Philedelphia, Pennsylvania, US state. 1989:690-706.
5. Savage CS, Winearls CG, Jones S, et al. Prospective study of radioimmunoassay for antibodies against neutrophil cytoplasm in diagnosis of systemic vasculitis. Lancet. 1987;329(8547):1389-1393.
6. Watson PG, Hayreh SS. Scleritis and episcleritis. Br J Ophthalmol. 1976;60:163-191.
7. Foster CS, de la Maza MS. Clinical considerations of episcleritis and scleritis: The Massachusetts Eye and Ear Infirmary experience. The sclera. 1994:95-136.
8. Yangzes S, Sharma VK, Singh SR, et al. Scleromalacia perforans in rheumatoid arthritis. Int J Med. 2019;112(6):459-460.