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Necrotizing Scleritis without Inflammation (Scleromalacia Perforans): A Case Report

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Abstract **Case Report**

Scleromalacia perforans SP is a rare ocular manifestation of rheumatoid arthritis which can potentially lead to blindness and is a late consequence in the course of the disease; in deed it is most common in elderly female with long-term rheumatoid arthritis, but it was also observed with other systemic diseases. It presents as a blackish blue hue visible through a thin sclera. scleral thinning is slow in onset and painless without inflammation. Progression can be prevented if treatment is instituted early. There is no specific and efficient treatment. As it develops on autoimmune abnormalities immunosuppressive therapy is proposed. To preserve globe integrity, scleral patch grafting with subsequent immunosuppression is performed.

Keywords: Case Report, Scleromalacia Perforans, Steroids, Rheumatoid Arthritis, Ocular Complication, Scleritis.

INTRODUCTION

Rheumatoid arthritis (RA) is a systemic disease that can affect more than just the joints. It is a disorder of autoimmune origin causing chronic inflammation. And it also affects other organs of the body such as eyes. Scleritis is an ophtalmic manifestation; it is frequently divided into five subsets based on the clinical presentation: diffuse anterior; nodular anterior; necrotizing; scleromalacia perforans; and posterior scleritis. Scleromalacia perforans is a rare form of anterior scleritis which readily presents as a blackish blue hue visible through a thin sclera [1]. No significant redness or pain is present but it is represented by progressive thinning of the sclera; it is a rare form of necrotizing anterior scleritis [2].

CASE PRESENTATION

This 42-year-old married mother of two was treated for rheumatoid arthritis with oral corticosteroids (10 mg/day). She presented with a painless bluish lesion in the sclera of her left eye, which prompted an outpatient consultation. She also complained of pain in all small joints of her hands associated with morning stiffness without any deformation.

On general physical examination, the patient was vitally stable.

Examination in our training revealed preserved visual acuity at 9/10 both eyes, slit lamp examination revealed discrete inhomogeneous conjunctival hyperhemia and nodules in the sclera. There was scleral thinning in superior temporal quadrant with visible uveal tissue, with surrounding dilated scleral vessels in the absence of any apparent inflammation. Schirmer's test values in both eyes were less than 5 mm indicative of severe dry eyes.

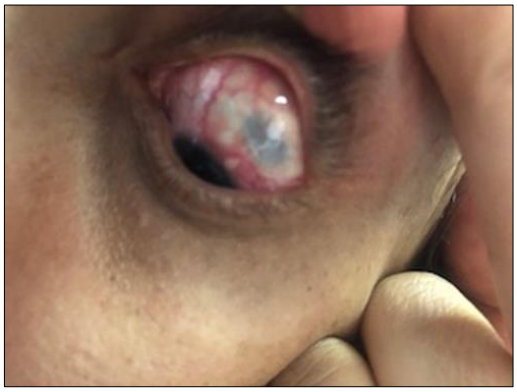

Figure 1: photograph of left eye showing presence of dilated scleral vessels with bluish discoloration of sclera at site of scleral thinning

The cornea is intact and clear, the anterior chamber calm, the iris color and trophicity normal and the lens clear. Examination of the posterior segment reveals a transparent vitreous and a posterior pole without anomalies.

On investigation she was found to have a strongly positive RA factor value of 170 IU/ml with a positive C-reactive protein (CRP) and anti-citrullinated cyclic polypeptide (CCP) of 30 IU/ml. The patient was not observant of her treatment.

She was started on methylprednisolone at a dose of 1 g intravenously administered once a day for 3 days, methotrexate 10 mg/week, analgesics, proton pump inhibitor. She received frequent topical tear substitutes, 8–10 times a day for dry eyes, topical non-steroidal anti-inflammatory drops and topical cyclosporine (0.1%) drops. A rheumatologist's consultation was sought to start systemic immunosuppressive therapy. The patient received intravenous cyclophosphamide and was maintained on oral cyclophosphamide. The dose of steroid was tapered to 10 mg then to 5 mg. She was asked to attend follow-up. Patient was advised regular follow up with the rheumatologist for systemic immunosuppression.

DISCUSSION

RA is a systemic disease that can affect the eyes. The ophthalmic manifestations of RA include keratoconjunctivitis sicca, episcleritis, scleritis, peripheral ulcerative keratitis, and retinal vasculitis which are described in the literature [3, 4]. Scleromalacia perforans is a rare, bilateral condition, occurring predominantly among elderly females with a history of severe, progressive, long-standing rheumatoid arthritis with extra-articular manifestations [5]. Apart from rheumatoid arthritis, this condition is described in other vasculitic and collagen vascular diseases as well [6]. In contrast to necrotizing scleritis with inflammation, the eye is not painful. The condition is characterized by the appearance of yellow to grayish patches on the sclera that gradually develop a necrotic slough or sequestrum which eventually separates from the underlying sclera, leaving bare choroid, covered by a thin layer of fibrous tissue or conjunctiva.

An initial characteristic finding on slit-lamp examination is a reduction in the number and size of vessels in the episclera surrounding the sequestrum, giving porcelain-like appearance [7]. These vessels anastomose with each other and sometimes cross the abnormal area to join with perilimbal vessels. The necrotic process in scleromalacia perforans appears to be caused by arteriolar obliteration as evidenced by fluorescein angiography, as opposed to venular nonperfusion, which is more prominent in necrotizing scleritis.

The initial color changes in sclera from porcelain white to yellow are often noted by the patient while looking in the mirror, by the patient's family, rheumatologist, or on routine ocular exam. The areas of exposed choroid are appearing blue-black in color. A nodule pathologically similar to a rheumatoid nodule may form in the sclera [8].

While spontaneous perforation is rare, these eyes are quite susceptible to traumatic rupture; SP can also leads to visual loss, secondary to progression of astigmatism due to scleral, paralimbic and corneal changes; it may complicates with anterior uveitis, cataract (secondary to uveitis or steroid therapy) or glaucoma (secondary to ocular abnormalities or steroid therapy) which is described generally in late stages of the scleritis.

Generally, the associated rheumatoid arthritis is long-standing and seropositive. Patients may have associated nodules, vasculitis, or pleuropericarditis. They have a shortened life expectancy compared with other patients with rheumatoid arthritis. It is unusual for scleritis to be an initial manifestation of rheumatoid arthritis.

There is no specific and efficacious therapy; lubricants, especially preservative-free artificial tears, are used. Punctal occlusion to increase tear film volume is performed by an ophthalmologist. Topical nonsteroidal anti-inflammatory drugs (NSAIDs) and steroids are insufficient even if topical corticosteroids should not be prescribed because of the possibility of necrosis and perforation of the cornea.

For corneal and scleral thinning, systemic immunosuppression usually is effective. Ocular perforation requires surgical repair [9]. In literature we can find report on favourable clinical response to the topical sodium versenate used as an inhibitor of collagenolytic enzyme. To treat refractory cases topically used cyclosporine A is described. In cases with severe necrotizing scleritis immunosuppressive therapy, supplemented with steroids is suggested to interrupt destructive process. Cyclophosphamide is known as the most effective drug in patients with noninfectious necrotizing scleritis (oral dose 2-3 mg/kg/d). Other immunosuppressive drugs like methotrexate (7.5-20 mg weekly), azathioprine (starting dose 2,5 mg/kg/d), cyclosporine (2.5-5.0 mg/kg/d) and mycophenolate mofetil (2-3 g/d) are well described. There are some reports describing: tumor necrosis factor inhibitors–TNF1 (etanercept, infliximab), the interleukin-2 receptor blocker (daclizumab), the interleukin-1 receptor antagonist (anakinra), the antilymphocyte medicament (rituximab, alemtuzumab) in ocular diseases, including scleritis.

Surgical treatment of SP is necessary in those cases with exposed uvea to preserve the globe integrity. Patch grafting can be performed with the sclera (fresh or frozen globe or glycerin preserved scleral tissue), dermis, fascia lata, periosteum, aortic tissue, cartilage, cornea, pedicle-flaps of conjunctiva with Müller muscle or tarsus, synthetic material and eventually amniotic

membrane. In some cases topical or systemic immunosuppression is necessary to inhibit destruction of the graft [7].

Owing to its insidious onset and lack of apparent symptoms in the initial course of disease, the condition is often detected late. Hence, it is imperative to diagnose the condition early and to promptly treat to prevent vision or life threatening complications [10].

CONCLUSION

RA is associated with many extra-articular manifestations, which include ocular diseases such as scleromalacia perforans. caracterized with insidious onset, slow progression and lack of symptoms until the bare choroid is seen under the thin layer of the conjunctiva. There are a lot of reports about trials of treating it both with medicaments and surgery but no one is completely efficacious. It remains a challenge for further studies. Collaborative efforts between the ophthalmologists and rheumatologists are essential to effectively manage any ocular complications that may arise.

Competing Interests: The authors declare that they have no competing interests.

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