

## A RARE CASE OF UNILATERAL ANNULAR SCLERITIS PROGRESSING TO SCLERA – KERATO - UVEITIS

H. R. Padmini<sup>1</sup>, Anushree Kumar<sup>2</sup>

<sup>1</sup>Professor and HOD, Department of Ophthalmology, Adichunchanagiri Institute of Medical Sciences.

<sup>2</sup>Post Graduate, Department of Ophthalmology, Adichunchanagiri Institute of Medical Sciences.

### ABSTRACT

#### INTRODUCTION

We agree to the fact that ocular trauma may lead to non-infectious uveitis with scleritis. Here, we report the case of a patient who developed unilateral annular scleritis as the complication of anterior scleritis following ocular trauma. The causes of inflammation including Infection and systemic inflammatory diseases were excluded. Hence, our findings suggest that ocular trauma may act as a trigger for scleritis.

Annular scleritis is defined as a circum-corneal gelatinous brawny infiltration accompanying the sclerosing keratitis. This rare entity occurs usually after the age of sixty. The case here described was unilateral annular sclera-kerato-uveitis leading to diffuse scleritis.

#### KEYWORDS

Ocular trauma, Annular scleritis, Sclera-kerato-uveitis, Photophobia, Lacrimation, Blepharospasm, Perforation at the limbic area.

**HOW TO CITE THIS ARTICLE:** Padmini HR, Kumar A. A rare case of unilateral annular scleritis progressing to sclera – kerato - uveitis. J. Evid. Based Med. Healthc. 2016; 3(15), 570-572. DOI: 10.18410/jebmh/2016/129

**INTRODUCTION:** Scleritis being a debilitating inflammatory disease that affects the sclera, causing economic blindness. It can result from several autoimmune, systemic diseases, trauma and idiopathic causes. Interstitial keratitis is a non-ulcerating inflammation of the corneal stroma without the involvement of either epithelium or endothelium. Keratitis in scleritis can be either infiltrative or destructive. Infiltrative lesions can present as localised or diffuse stromal keratitis, sclerosing keratitis or as a deep keratitis. Sclerosing keratitis is known to be associated with 1. Rheumatoid arthritis, 2. Onchocerciasis, 3. Tuberculosis, 4. and other infective or auto immune diseases.

Annular scleritis (AS) is a potentially a vision-threatening inflammation of the sclera whose aetiology may include autoimmune and systemic conditions such as rheumatoid arthritis and tuberculosis and occasionally trauma as seen in our case. The signs and symptoms of AS include pain radiating immensely to periorbital region, tearing, tenderness, redness, painful sensitivity to light and decreased visual acuity. The most important ocular signs of scleritis are globe tenderness on palpation, sectorial or diffuse scleral erythema, thinning with bluish hue, oedema and possible nodules or necrosis. Scleritis is usually confined to one eye, but may affect both eyes and there may be possible corneal or intraocular inflammation. If left untreated, scleritis may cause perforation of the eyeball, leading to vision loss or extreme condition of endophthalmitis. Ocular and physical examinations

including blood tests to rule out underlying causes are important. Medications such as corticosteroids, non-steroidal anti-inflammatory drugs and possibly immune-suppressants and antibiotic cover are used in the management of AS. If care is not taken, AS can be misdiagnosed as keratitis. Such mis-diagnosis can be sight-threatening and therefore it is essential that primary eye care practitioners are cautious in all diagnosis of red eye conditions. Sclerosing keratitis is manifested by scleralisation of the deep peripheral cornea adjacent to area of scleritis. It is gray or grayish-yellow at onset and later becomes bluish white or dense white. Thus, a yellow or white "crystalline" opacity appears insidiously in the mid-stroma either in a diffuse or sectoral fashion. These opacities are characteristically described as "Candy Floss Opacities" or "Cotton Candy Opacities".<sup>(1)</sup> There is minimal vascularization, but is associated with corneal thinning. Individual opacities are often tongue shaped or triangular with base directed towards limbus. These opacities rarely resolve. There may be associated anterior uveitis whose severity depends on the individual case. Our case also progressed to such a condition of peripheral corneal thinning adjacent to scleritis leading to central corneal opacity mimicking Mooren's ulcer and also associated with intercalary uveitis.

**CASE REPORT:** A 55-year-old female patient, farmer by occupation presented to ophthalmology OPD of AIMS hospital/Bellur with a history of trauma left eye from the branch of a tree while working in the field. Patient had no past history of rheumatism, gout or tuberculosis. Previous ocular, medical, treatment and family histories were unremarkable. She complained of a left red eye preceding since one week, with photophobia, severe watering, severe

Submission 19-01-2016, Peer Review 04-02-2016,  
Acceptance 12-02-2016, Published 22-02-2016.

Corresponding Author:

Dr. Anushree Kumar,

Post Graduate, Department of Ophthalmology,  
Adichunchanagiri Institute of Medical sciences.

E-mail: anushree2203@gmail.com

DOI: 10.18410/jebmh/2016/129

pain radiating to temporal side disturbing her normal sleep rhythm and diminution of visual acuity of left eye.

Ocular examination revealed blepharospasm and redness of the left eye accompanied with circum-corneal congestion and chemosis with infiltration of the corneal periphery presenting from past 1 week. The best corrected visual activity was 6/9 in the Right eye, while the Left eye presented with painful perception of light. Further Slit lamp examination of the left eye showed annular scleritis with swollen anterior sclera and markedly peripheral congested vessels. Adjacent corneal stroma was oedematous, with infiltrate and haemorrhage from deep vessels giving a picture of ulcerative keratitis. Aqueous contained small number of cells seen in 1\*1mm slit in all quadrants. Intraocular pressure as measured with rebound tonometry was normal for both eyes. Overall patient presented with good general health. No intraocular foreign body could be viewed. Right ocular examination was unremarkable in all aspects. The diagnosis of a rare unilateral annular scleritis progressing to sclera-kerato-uveitis for left eye was made.

Due to suspicion of an infectious aetiology, treatment was instituted with topical moxifloxacin with dexamethasone hourly and natamycin six times daily. Oral ciprofloxacin and ketoconazole, as well as ibuprofen and diamox 250mg BD were also started. No micro-organisms were isolated from the left conjunctival swab. C-reactive protein, rheumatoid factor, anti-nuclear, anti-double stranded DNA and anti-neutrophil cytoplasmic antibodies, and syphilis serology were either normal or negative. In view of negative microbiological investigations normal chest X-ray and systemic examination, and high endemicity of tuberculosis in the patient's homeland, her reaction was considered unrelated to the scleritis. Clinical improvement was apparent within 48 hours, with marked reduction of scleral oedema and congestion but increased rapidly later leading to thinning of peripheral cornea with sclerosing keratitis predicted to have unusual corneal perforation with iris prolapse and loss of vision.<sup>(2)</sup>



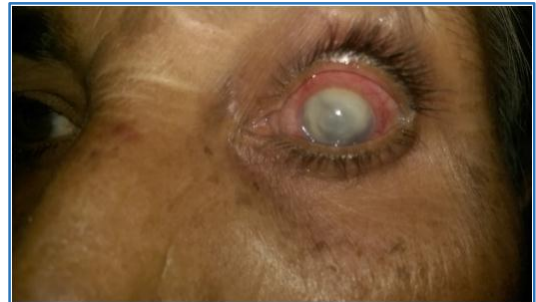
**Fig. 1: Showing left eye Anterior Annular Scleritis**



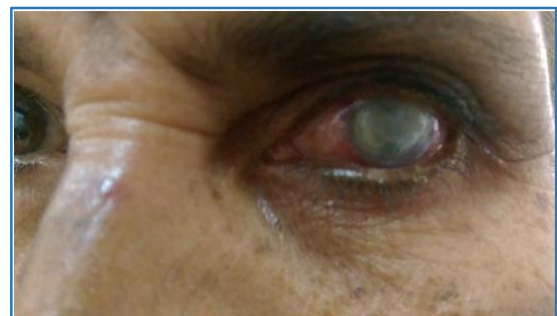
**Fig. 2: Slit lamp examination showing cells and flare in anterior chamber characteristic of uveitic reaction seen in all quadrants**



**Fig.3: Picture depicting staining of cornea after implication of fluorescein dye**



**Fig. 4: Unilateral anterior annular scleritis going for sclera-kerato-uveitis, comparing both eye**



**Fig. 5: Sclera-kerato-uveitis mimicking Mooren's ulcer**

**DISCUSSION:** If sclera gets inflamed, oedematous and infiltrated with inflammatory cells it leads to a very severe inflammatory condition called Scleritis. The presenting symptoms commonly seen are pain and redness. It is most commonly seen in the fourth to sixth decade, with a high incidence in the fifth decade. In 40-45% of the cases it is bilateral. Subsets of T-helper lymphocytes, which have recently been described are known as Th-17, and known to be the key factors in the pathogenesis of scleritis. Although scleritis is not a common disease, it is usually associated with autoimmune diseases such as rheumatoid arthritis, systemic lupus erythematosus, ankylosing spondylitis, giant cell arteritis, polyarteritis nodosa, Wegener's granulomatosis, and gout. Different organisms such as Pseudomonas, aspergillus, Mycobacteria and mixed organisms can cause infectious scleritis. Primary fungal scleral infection in a non-immunocompromised patient is extremely rare but can be suspected when there is a history of scleral traumatic injury, pterygium surgery,

glaucoma surgery, retinal detachment surgery, cataract surgery especially in diabetics 1,2 with/without topical steroid therapy, or in association with systemic fungal infection. The onset of fungal scleritis may start early after trauma. Recent studies have shown that scleritis has also been associated with erythema elevatum diutinum, a chronic and rare dermatosis that is considered to be a variant of leukocytoclastic vasculitis. Other systemic conditions such as tuberculosis, herpes zoster ophthalmicus and syphilis have also been reported to be associated with scleritis and sometimes the cause is unknown. It can also be attained through disorders of menstruation. It is for this reason that scleritis occurs more frequently in women aged 30 to 60 years and is rare in children.

Scleritis is classified depending on the site of pathology and severity of inflammation into anterior and posterior. There anterior scleritis are further divided into: diffuse, nodular, necrotizing scleritis with inflammation and necrotizing without inflammation (scleromalacia perforans). The inflammation of diffuse scleritis is generalized, involving some area of the whole anterior segment. Nodular scleritis causes the affected area of the sclera to be confined to small nodules and necrotizing scleritis is the most severe. Posterior scleritis is a rare and potentially blinding condition, characterized by flattening of the posterior aspect of the globe, thickening of the choroid and sclera, and retro bulbar oedema presenting with poor or double vision, severe pain.<sup>(3)</sup> Traumatic and infectious anterior scleritis clinically presents with persistent redness, ulceration, severe anterior chamber reaction, abscess and/or nodule formation, and typically not responding to routine antibiotics. Therefore, patients presenting with scleritis especially after surgery in diabetics or in geographic areas with hot and humid climates should alert the ophthalmologist for fungal infection. The microbiological and histopathological diagnosis of fungus scleritis is often difficult as the organisms are usually deep in the sclera, few in number, and are rarely revealed by scraping. The gold standard initial therapy of non-infectious scleritis includes a corticosteroid such as prednisolone and the use of non-steroidal anti-inflammatory agents for pain relief. In more extreme cases of scleritis, chemotherapy (such as immunosuppressive therapy with such drugs as cyclophosphamide or azathioprine) may be used to treat the disease. Oral Itraconazole and topical Amphotericin B and natamycin as a medical therapy for fungal scleritis along with topical antibiotic cover.<sup>(4)</sup> Surgical outcome in debilitating sclerotic and keratitic cases can be achieved by scleral debridement, dura matter patch graft as it is

advantageous over cryotherapy. Neuro-imaging tests such as computed tomography (CT) scan, magnetic resonance imaging (MRI) or ultrasonography of the eye may be ordered in severe posterior cases along with complete haemogram investigations.<sup>(5)</sup>

Wolfgang1 0 et al emphasized failure of microbiological detection in traumatic infectious scleritis in 25 patients in his study.<sup>(6)</sup> Thus, we conclude that ocular trauma was likely a precipitating factor in this case of scleritis. The temporal relationship and inflammation at the site of trauma strongly suggest the association. Sympathetic ophthalmia is a dreaded complication of penetrating eye injury, and recently, even trivial blunt trauma has been recognised as a precipitant of uveitis.<sup>(7)</sup> In our case also after detecting the uniocular annular scleritis intense antibiotic and antifungal treatment was instilled but after 15 days patient came back with the progressed stage of sclero-kerato-uveitis with peripheral corneal thinning going for severe complication of ocular perforation and endophthalmitis.

**CONCLUSION:** A case of annular scleritis with complications of sclera-kerato-uveitis following ocular trauma to the left eye in a woman aged 55 years which led to perforation followed by its further complications discussed in detail.

## REFERENCES

1. Gopal Kishan M, Sheetal Baldava, Syed Musaab Mohiuddin. Sclerosing keratitis in reactivated systemic tuberculosis. *Journal of Evidence based Medicine and Healthcare* 2015;2(16):2438-2441.
2. Smith JR, Chee SP. Nodular anterior scleritis associated with ocular trauma. *Singapore Med J* 2001;42(4):180-181.
3. Ratchitskaya A, Mandelcorn ED, Albin TA. An update on the cause and treatment of scleritis. *Curr Opin Ophthalmol* 2010;21(6):463-467.
4. Mashige KP. Anterior diffuse scleritis diagnosed as conjunctivitis. *S Afr Optom* 2012;71(1):51-54.
5. Rosenbaum JT, Tammaro J, Robertson JE. Uveitis precipitated by nonpenetrating ocular trauma. *Am J Ophthalmol* 1991;112(4):392-395.
6. Tu EY. Interstitial keratitis in principles and practice of ophthalmology 3e, Jakobienced 2007.
7. Osman E, Hassanyh S, Al-Muammar A, et al. Post-traumatic Aspergillus scleritis. *Saudi Journal of Ophthalmology* 2005;19(1).