Journal of Clinical & Experimental Ophthalmology

Kopacz et al., J Clinic Experiment Ophthalmol 2013, S2
DOI: 10.4172/2155-9570.S2-009

Review Article Open Access

Scleromalacia Perforans–What We Know and What We Can Do

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Abstract

Anterior necrotizing scleritis without inflammation, so called scleromalacia perforans, is a rare, severe eye disorder developing on autoimmune damage of episcleral and scleral performing vessels (hypersensitivity type III). The onset of the disease is insidious, progression is slow and no specific symptoms are observed until discoloration of the sclera (necrotic slough, bare choroid) is detected. Scleromalacia perforans is most common in women with long-term rheumatoid arthritis, but it was also observed with other systemic diseases. There is no specific and efficient treatment. As it develops on autoimmune abnormalities immunosuppressive therapy is proposed. To preserve globe integrity, scleral patch grafting (both tissues and synthetic materials) with subsequent immunosuppression is performed.

Keywords: Scleromalacia perforans; Rheumatoid nodules; Extra-articular arthritis; Immunosuppressive therapy

Classification, Epidemiology, Clinical Manifestations

The sclera is a dense connective tissue covering about five sixths of the eye [1,2]. Watson and Hayreh (based on the anatomic site of the inflammation and on the changes in scleral vasculature) proposed two main categories of the eye wall inflammation: episcleritis (simple and nodular) and scleritis (anterior and posterior) [2,3]. Analysis of the anterior scleritis subclassified it into diffuse, nodular and necrotizing (with inflammation-anterior necrotizing scleritis- or without inflammation-scleromalacia perforans).

Scleromalcia perforans (SP) is a rare (only 4% of scleritis) scleral disease, characterized by the progressive scleral thinning without inflammation [4]. The condition was previously described by Van der Hoeve: he noted that it was bilateral, began with yellow or greyish subconjunctival nodules and gradually developed into scleral necrosis with perforation and exposure of the uvea [1,5-7]. It is difficult to define the onset of the disease because of slow progression and total lack of symptoms. The change in scleral color is detected by patient's family, by patient looking in the mirror or ophthalmologist during routine examination. Progression of the disease reveals as necrotic slough without surrounding inflammation; absorption or slough separation leave bare uvea, covered only by thin layer of conjunctiva. Nevertheless perforations are uncommon without trauma [1,2,4]. The area of scleral lack is surrounded with pathological vessels anastomosing with each other or crossing the abnormal area to join with perilimbal vessels [1,4,5,8]. SP is commonly associated with severe, progressive, long standing rheumatoid arthritis with extra-articular manifestation, more frequent in women [1,2,6]. The condition was also described in other systemic vasculitic and collagen disorders (up to 66%): systemic lupus erythematosus, periarteritis nodosa, Wegener's granulomatosis, Behçet disease, limited scleroderma, Crohn's disease, graft-versus-host disease. SP was also observed in porphyria and herpes-zoster infection [2,8-15] (Figure 1).

Histopathology

Verhoeff and King published the first histological report of the condition: changes in the scleral nodules were similar to those of the subcutaneous nodules of the rheumatoid arthritis [6,16]. Chronic granulomatous changes with epithelioid cells surround central, necrotic masses (collagen and noncollagen fibers, cell debris). Young and Watson suggest three determinants of scleral destruction: activation of scleral fibrocytes and resorption of pericellular matrix, infiltration of the scleral stroma by inflammatory cells, prolonged local vaso-occlusion. In SP dense plaques of necrotic tissues are removed and it is associated with full thickness loss of conjunctiva and insufficiency of conjunctival epithelium to resurface the exposed area [1,6,8,9,17-19].

Fluorescein Angiography

Watson and Bovey analyzed vascular disorders in scleritis: results of fluorescein angiography shown differences between scleromalacia perforans and other scleritis [20]. In SP necrotic process appears to be the result of arteriolar–not venular like in anterior necrotizing scleritis-obliteration [1,20].

Complications

Visual loss, secondary to progression of astigmatism (sclear and paralimbic/corneal changes), anterior uveitis, cataract (secondary to uveitis or steroid therapy) or glaucoma (secondary to ocular abnormalities or steroid therapy), was described generally in late stages of the scleritis (up to 60% cases) [1,2,4].

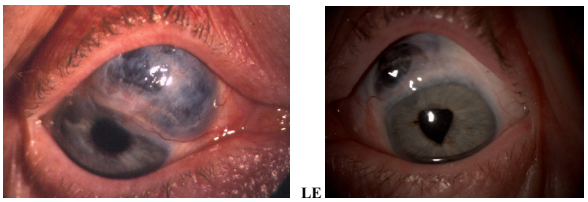

Figure 1: 73-years old patient with long-term rheumatoid arthritis and bilateral scleromalacia perforans (LE with thinning sclera next to scleral patch).

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Received January 20, 2013; **Accepted** March 01, 2013; **Published** March 08, 2013

Citation: Kopacz D, Maciejewicz P, Kopacz M (2013) Scleromalacia Perforans–What We Know and What We Can Do. J Clinic Experiment Ophthalmol S2: 009. doi:10.4172/2155-9570.S2-009

Therapy

There is no specific and efficacious therapy [21]. Topical nonsteroidal anti-inflammatory drugs (NSAIDs) and steroids are insufficient [1,2]. In literature we can find report on favourable clinical response to the topical sodium versenate used as an inhibitor of collagenolytic enzyme [12]. To treat refractory cases topically used cyclosporine A is described [2,22,23].

Irrespectively of the final diagnosis, autoimmune reaction is responsible for the vessels damage (type III hypersensitivity) [1,2,24]. That is why in cases with severe necrotizing scleritis immunosuppressive therapy, supplemented with steroids is suggested to interrupt destructive process [1,2,6,9,12-15,17,18,24]. 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 [1,2,8,11,13,17,23]. So called "biologics" are the new group of modifying immune response agents. 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. To detect and prevent side effects of immunosuppressive therapy, a collaboration with physicians trained in the early recognition and management of drug-induced complication is recommended (e.g. hematologist, rheumatologist, internal medicine specialist) [1,2,8,25-27].

Surgical treatment of SP is necessary in those cases with exposed uvea to preserve the globe integrity. Tectonic 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 (GoreTex®) and eventually amniotic membrane. In some cases topical or systemic immunosuppression is necessary to inhibit destruction [1,2,8,12,28-30] (Figure 2).

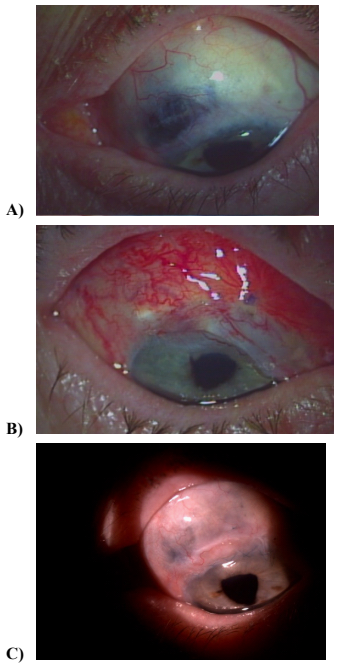

Figure 2: Tectonic patches. A) Sclera (progressive thinning of the sclera next to the patch) B) GoreTex® (RE from the figure1) C) Cornea (LE from the figure1).

Summary

Scleromalacia perforans is a severe disorder of the globe 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.

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This article was originally published in a special issue, **Ocular Diseases: Immunological and Molecular Mechanisms** handled by Editor(s). Dr. C S De Paiva, Cullen Eye Institute, Baylor College of Medicine, USA