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Review Article

Ocular findings in systemic lupus erythematosus

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Abstract

Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease. Ocular complications occur in up to one-third of patients with SLE. The ocular findings may represent the initial manifestation of the disease and may lead to severe ocular morbidity and loss of vision. Early diagnosis and prompt management of patients with SLE are mandatory and require collaboration between the ophthalmologist and the rheumatologist.

Keywords: SLE, Autoimmune, Ocular complications

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Systemic lupus erythematosus (SLE) is a chronic multisystem autoimmune disease. The eye is frequently involved in SLE.[1] The disease may cause ocular involvement by several mechanisms including immune complex deposition in the basement membrane of endothelial cells of the small blood vessels.[1] Ocular complications have been reported in up to one-third of patients with SLE.[2] Ocular manifestations of SLE may be due to the disease or may be due to the complications of systemic or topical therapy.[3] Unlike other autoimmune diseases, which may have a predilection for either anterior or posterior segment of the eye, SLE may affect any structure of the eye and adnexa.[3] The ocular findings in SLE are important because they may be the initial manifestation of the disease.[3]

The diagnosis of SLE is clinical and is based on the presence of 4 of the 11 features listed by the American College of Rheumatology classification criteria.[4] The presence of four criteria makes the diagnosis of SLE, serially or simultaneously, during the course of the disease. The revised criteria include: (1) malar rash, (2) discoid rash, (3) skin photosensitivity, (4) oral ulcers, (5) nonerosive arthritis, (6) serositis, (7) renal involvement, (8) neurological disorder, (9) hematologic disorder, (10) immunologic disorder, and (11) positive antinuclear antibodies.[4] The presence of 4 of these 11 criteria confirms the diagnosis of SLE and yields a sensitivity of 85% and a specificity of 95% for SLE.[5] The main purpose of this paper was to present the clinical findings and complications of therapy in patients with SLE.

Ocular involvement in SLE is variable affecting various parts of the eye and the visual pathway and may be sight threatening. Table 1 shows the ocular manifestations of SLE.

Orbital and external eye disease

Orbital involvement is a rare manifestation of SLE. Clinical presentation may be in the form of proptosis, enophthalmos, orbital pain, blurred vision, chemosis, and restriction of extraocular motility.[6,7] Limitation of ocular motility may result from orbital vasculitis and/or orbital myositis.[8,9] Orbital vasculitis may lead to nonperfusion of the globe and extraocular muscles leading to restriction of extraocular movements. Orbital myositis secondary to SLE may be misdiagnosed as

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Table 1. Ocular involvement in systemic lupus erythematosus.

Structure	Clinical findings
Orbital and external eye disease	Discoid lupus-type rash over the eyelids Panniculitis Orbital masses Periorbital edema Orbital myositis Orbital vasculitis, acute orbital ischemia and infarction
Conjunctival involvement	Conjunctivitis
Corneal involvement	Dry eye syndrome Recurrent corneal erosions Peripheral corneal infiltrates Peripheral ulcerative keratitis Interstitial keratitis Endotheliitis Keratoconus
Sclera and Episclera	Scleritis Episcleritis
Uveal involvement	Anterior uveitis
Retinal involvement	Lupus retinopathy (cotton wool spots, intraretinal hemorrhages, and vascular tortuosity) Retinal hard exudates Retinal vasculitis Retinal artery and/or vein occlusion Arteriolar narrowing and arteriovenous crossing changes Macular pigmentary mottling Retinal scarring Macular infarction
Choroidal involvement	Central serous chorioretinopathy
Neuro-ophthalmic findings	*Optic nerve involvement* Optic neuritis Ischemic optic neuropathy Papilledema *Central nervous system vasculitis* Internuclear ophthalmoplegia Nystagmus Cranial nerve palsies Homonymous hemianopia

bacterial orbital cellulitis. CT scan of the orbit or diagnostic ultrasound may reveal enlargement of the extraocular muscles in these cases.[6] Treatment of orbital disease is with systemic immunosuppressant drugs. Discoid lupus erythematosus (DLE) is a chronic cutaneous lupus erythematosus without internal organ involvement.[10] SLE and DLE can present with a discoid lupus-type rash over the eyelids. These discrete raised scaly lesions may be confused with chronic blepharitis. Treatment of these lesions is usually with systemic anti-inflammatory drugs.[1] Eyelid involvement was reported also in cases of cutaneous lupus erythematosus.[9] Lupus panniculitis is a rare skin condition, which predominantly affects the deep dermis and subcutaneous fat in young to middle-aged women. It may occur independently or in association with DLE or SLE. Clinically, these lesions appear as nodules or hardened subcutaneous plaques which are often adherent to the overlying skin. After healing, they may undergo atrophy and residual scarring. These nodules and plaques are usually located on the forehead, cheeks, extremities, and buttocks. They have occasionally been reported to affect the orbit.[11] The main treatment option for orbital and external eye disease is systemic hydroxychloroquine therapy. In cases of failure of antimalarials, immunosuppressive medications can be considered.[12] Several agents have been reported to be successfully used for DLE including azathioprine, dapsone, methotrexate, cyclophosphamide, thalidomide, retinoids, and interferon alpha-2. Corticosteroids are mainly used in patients with orbital inflammatory syndrome to control the severe inflammation or associated with hydroxychloroquine therapy at the beginning of the treatment.[12]

Ocular surface disease

Both the major and accessory lacrimal gland may be involved in patients with SLE.[13] Mononuclear cellular infiltrate of the lacrimal glands may lead to decrease in lacrimal fluid. Keratoconjunctivitis sicca is a common ocular feature of SLE. Keratoconjunctivitis sicca leads to upregulation of inflammatory cytokines causing chronic conjunctivitis[13] It tends to be mild but in rare cases it may lead to conjunctival scarring and shrinkage.[14] The prevalence of keratoconjunctivitis sicca among patients with SLE is approximately 25%.[2,15] The symptoms range from mild irritation, foreign body sensation and redness to severe pain due to corneal ulcer and filamentary keratitis. Significant visual impairment may occur due to corneal epithelial defects, corneal ulceration, vascularization and scarring. Treatment of dry eyes associated with SLE is usually through frequent instillation of lubricating eyedrops. Severe cases may require temporary or permanent punctual occlusion. Some cases may benefit from topical tacrolimus as it decreases the inflammatory cellular infiltrate of the lacrimal glands.[16]

Corneal involvement

In addition to dry eye syndrome SLE can be associated with other corneal manifestations such as recurrent corneal erosion, peripheral corneal infiltration, ulcerative Keratitis,[17] interstitial keratitis,[18] and endotheliitis.[19] Immune complexes may deposit in the basement membrane of the endothelial cells of the limbal blood vessels which may lead to release of chemotactic cytokines and may cause peripheral corneal infiltrates. Accordingly, corneal infiltrates can be treated with topical steroids with rapid response.[1] In addition, several autoimmune diseases such as SLE may be associated with keratoconus and this may point to the role of the immune system in the pathogenesis of keratoconus.[20]

Sclera and Episclera and Uveal involvement

Scleritis may occur in SLE, and may be the presenting feature of the disease.[3] The incidence of SLE in patients with scleritis is about 1%.[21,22] Scleritis in patients with SLE may present as anterior diffuse scleritis or anterior nodular scleritis. Necrotizing scleritis in patients with SLE is rare but may lead to significant scleral thinning.[22] Posterior scleritis is also rarely seen in patients with SLE.[23] Episcleritis may be also seen in SLE with milder symptoms and redness due to injection of the superficial blood vessels.[24,25]

Episcleritis is usually self-limiting disease which does not require treatment. Topical non-steroidal or steroidal eyedrops may be required in severe cases. On the other hand,