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Ocular Manifestations of Systemic Lupus Erythematosus

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Systemic lupus erythematosus (SLE) is a chronic, systemic autoimmune disease, particularly prevalent in women, probably with a genetic predisposition with "triggering" from contact from an environmental stimulus, resulting in the production of pathogenic autoantibodies and immune complexes which produce the pathologic features of the disease. SLE has protean manifestations, and is difficult to diagnose in its early stages of evolution. The diagnosis can be definitively established if 4 of the 11 American College of Rheumatology criteria are met, serially or simultaneously. Although ocular inflammation may be a manifestation of SLE (indeed, may be the initial clinical obvious one), the ocular lesions **are not** included among the 11 criteria; we believe this is an oversight and believe, further, that inclusion of ocular inflammation among the diagnostic criteria for SLE would enable earlier establishment of the diagnosis and earlier therapeutic intervention in some instances. Corneal manifestations of SLE are confined primarily to ocular surface epitheliopathy secondary to keratoconjunctivitis sicca, and stromal keratitis (rare), particularly peripheral and segmental. In our 47 patients with SLE which we have analyzed, 16 had corneal complications, 62.5% secondary to keratoconjunctivitis sicca, and the rest secondary to the lupus disease activity itself. Episcleritis or scleritis may also occur as a consequence of SLE, and scleritis is a reasonably accurate guide to the presence of significant systemic activity in the SLE patient; it will resolve only with adequate control of the disease activity and will not respond to topical therapy. One of our 94 patients with episcleritis had SLE, and 7 of the 172 patients with scleritis of whom we have reported had SLE. Systemic nonsteroid anti-inflammatory drug therapy was necessary to eliminate the SLE episcleritis, unlike the usual situation in patients with idiopathic episcleritis. Systemic hydroxychloroquine was also effective. Retinal involvement is the most common ocular manifestation of SLE after keratoconjunctivitis sicca. Additionally, the presence of active SLE retinal vasculopathy is an extremely accurate guide to the presence of systemic disease activity, occult or overt. Additionally, life-table survival estimates have shown decreased survival in patients with SLE retinopathy, compared to SLE patients without retinopathy. Retinal lesions in SLE patients, therefore, are of critical importance, both visually and prognostically. Lupus retinopathy should alert the clinician to the likelihood of ocular and systemic vasculitis lesions meriting aggressive systemic therapy. Retinal vasculitis appears early in the development of lupus retinopathy, and evident on fluorescein angiography. Sub-clinical macular edema and intraretinal hemorrhages and cotton wool spots follow, and these latter changes may be indistinguishable from hypertensive retinopathy, and it may be impossible for the clinician to decide

whether the retinal lesions are secondary to hypertension or to SLE immune complex vasculitis. Aggressive therapy for the hypertension and the SLE systemic disease activity is associated with a dramatic decrease in retinal lesions in such patients and improves the patient's overall prognosis. The appearance of SLE retinopathy is associated with central nervous system lupus in particular. Additionally, 5% to 10% of patients with SLE retinopathy will develop large vessel disease and the presence of anti-phospholipid antibodies, anticardiolipin antibodies. Corticosteroid and Heparinization may be critical in treatment of this form of lupus retinopathy. Finally, choroidopathy can occur in patients with SLE, with resultant serous retinal detachment.