Original Paper

Received: December 29, 1983

Accepted: December 29, 1983

Ophthalmologica 1984;189:128-129

Unilateral Central Retinal Vein Occlusion in Systemic Lupus Erythematosus

L.	Laurent	Laroche
H.	Henry	Saraux

Department of Ophthalmology, Hôpital Saint-Antoine, Paris, France

Key Words

Central retinal vein occlusion
Systemic lupus erythematosus
Vasculitis

Abstract

A case of unilateral central vein occlusion occurring in a normotensive patient with systemic lupus erythematosus (SLE) is reported. This vasculitis is related to SLE. In spite of the observation of circulating immune complex and a positive response to the human basophil degranulation test in the presence of bovine soluble retinal antigen, the real pathogenesis of this unilateral vasculitis remains obscure.

Prof. H. Saraux, Department of Ophthalmology, Hôpital Saint-Antoine, 184, Rue du Faubourg Saint-Antoine, F-75571 Paris Cédex 12 (France)

Normotensive patients with systemic lupus erythematosus (SLE) may develop usually bilateral retinopathy which is not primarily a venous disease. However, a few cases of central vein occlusion in SLE have already been published [1], and Terhorst et al. [2] recently reported a case of unilateral arterial retinopathy. We report a case of unilateral central vein occlusion, occurring in a patient with SLE.

Case Report

A 53-year-old black woman, who had had florid SLE without cerebral involvement for 13 years, was referred to us due to visual haze as her general condition decreased. Four of the clinical diagnosis criteria of the American Rheumatism Association were present, typical biological abnormalities were found.

She received 20 mg of prednisone daily. Her visual acuity was RE 10/10 and LE 9/10. The left fundus showed papilledema, venous tortuosity, engorgement and sheathing. Many small diffuse intraretinal hemorrhages were present, and a few cotton-wool spots indicated coexistent arterial involvement. The right fundus was normal. Fluorescein angiography of the left eye did not demonstrated choroidal or arterial perfusion delay, but a marked venous filling delay. Disc and perimacular capillaries were swollen. There were a few scattered areas of obliterated capillary bed.

Late phase fluorescein angiograms disclosed a huge dye leakage from the veins, in areas with bright parietal staining (fig. 1). Right eye angiography was normal.

Her visual function improved quickly under treatment with prednisone (some dose) and heparin.

Discussion

Retinopathy, due to SLE itself, occurs more frequently in seriously ill patients with Unilateral Central Retinal Vein Occlusion in Systemic Lupus Erythematosus

Fig. 1. Late phase fluorescein angiogram showing diffuse leakage from the disc vessels and parietal venous dye staining.

vasculitis localization is not stable. Indeed, Aronson et al. [5] demonstrated immuno-globulins and complement deposits in the wall of the retinal vessels of patients dying of SLE. When skin is involved in SLE, biopsy specimens from areas with or without rash almost constantly show immunofluorescent staining. Then the normality of ophthalmo-scopic and angiographic right eye examination shows only the limits of these methods. The lack of ocular vasculitis in one eye does not mean the absence of immune complex deposits. The histopathological examination of the vasculitis-free eye would probably disclose immunofluorescent staining.

active systemic disease [3]. No correlation has been established between retinopathy and cerebral disease [4]. SLE is an autoimmune disease, simultaneously or successively involving many organs. Vasculitis is believed to be the most important process in the pathogenesis of the lesions because of vascular parietal immune complex deposition. In our patient's blood, C3 and C4 complement levels were within normal limits, but circulating immune complexes were present (IgG, anti- $\frac{1}{8}$ G, IgA, anti- $\frac{1}{8}$ A, IgM, anti-IgM). Moreover, a positive response to the human basophil degranulation test was found in the presence of bovine soluble retinal antigen. Such an IgE-mediated hyper-sensitivity was observed in experimental uveoretinitis induced by retinal S antigen in monkeys. In this disease a prominent bilateral vasculitis with venous involvement occurs. The real pathogenesis of our patient's retinal vascular disease remains obscure, though it is obviously related to SLE.

In a systemic disease resulting from immune complex deposition the 'only one eye'

References

- Silverman, M.; Lubeck, M.J.; Briney, W.G.: Central retinal vein occlusion complicating systemic lupus erythematosus. *Arthritis Rheum.* 21: 839 (1978).
- Terhorst, D.R.; Campo, R.V.; Abrams, G.W.: Unilateral systemic lupus erythematosus retinopathy. *Am. J. Ophthalm.* 95: 840 (1983).
- Gold, D.; Feiner, L.; Henkind, P.: Retinal arterial occlusive disease in systemic lupus erythematosus. *Archs Ophthalm.* Chicago 95: 1580 (1977).
- Lanham, J. G.; Barrie, T.; Kohner, E. M.; Hugues, R.V.: SLE retinopathy: evaluation by fluorescein angiography. *Ann. rheum. Dis.* 41: 473 (1982).
- Aronson, A. J.; Ordonez, N. G.; Diddie, K. R.; Ernest, J. T.: Immune complex deposition in the eye in systemic lupus erythematosus. *Archs intern. Med.* 139: 1312 (1979).