Retinal detachment as the first manifestation of systemic lupus erythematosus

Sirs,

Systemic lupus erythematosus (SLE) is a chronic systemic autoimmune disease that affects multiple organs. Ocular manifestations include dry eye syndrome and less commonly lupus retinopathy, scleritis and episcleritis (1). Lupus choroidopathy is a rare manifestation usually presenting when an established diagnosis of SLE has already been made. We present a case of bilateral choroidopathy as the first clinical manifestation of SLE.

A 65-year-old woman presented at our clinic complaining loss of vision in her right eye (RE). Her past medical history was significant for malar rash and photosensitivity. Visual acuity was 20/200 in the RE and 20/20 in the left eye (LE). Ophthalmologic examination showed a retinal detachment (RD) in RE, whereas it was unremarkable in LE. Fluorescein angiography (FA), indocyanine angiography (ICGA) and optical coherence tomography (OCT) showed choroidal inflammation and a concomitant exudative RD (Fig. 1) in RE. Anti-neutrophil cytoplasm antibody (ANCA), Quantiferon, Mantoux, Treponema pallidum particle agglutination assay (TPPA), fluorescent treponemal antibody absorption test (FTA-Abs), anti-double stranded anti-DNA test were negative, while antinuclear antibody (ANA) was positive. A diffuse lymphadenopathy was observed in the chest and abdominal CT scan. An axillary lymph node biopsy showed reactive lymphadenitis. At that time the patient voluntarily left the hospital without any treatment, but returned six months later with new symptoms in her LE, and an inferior exudative RD was diagnosed. In LE, FA and ICGA findings were consistent with choroidal inflammation (Fig. 1). The repeated anti-double stranded anti-DNA test was positive together with a positivity for anti-RO, anti-RNP, anti SM and low levels of C3 complement. Hypertensive choroidopathy was ruled out because of normal systemic blood pressure, diagnosis of SLE choroidopathy was made and treatment with systemic intravenous steroids and additional immunosuppressive drugs was started.

SLE is a life-threatening multisystem autoimmune disease with potential sight-threatening ocular manifestations.

Following the diagnostic criteria of the American College of Rheumatology (ACR) (2), in 2012 the SLE International Collaborating Clinics (SLICC) group produced a new classification system comprising 11 clinical and 6 immunological criteria. For a diagnosis of SLE, at least 4 criteria, including at least one clinical and one immunologic criterion, must be met (3).

In our patient the diagnosis of SLE was based on the presence of anti-DNA antibody, ANA positivity, anti RO, anti-RNP, anti SM, low complement (C3) together with photosensitivity and malar rash.

Choroidopathy is a rare lupus manifestation.

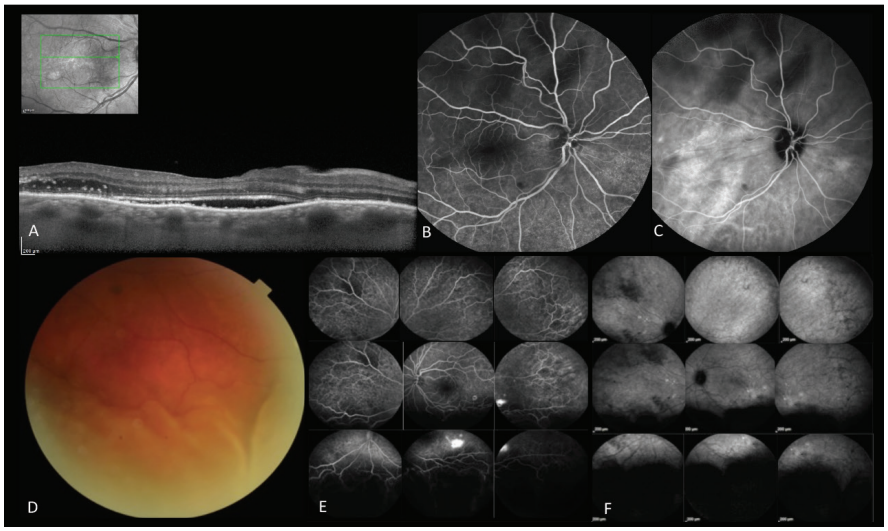

Fig. 1. Right eye. (**A**) OCT shows exudative retinal detachment (RD) associated (**B**) to choroidal folds visible as hypofluorescent and hypocyanescent areas at the posterior pole and in the mid-periphery of the retina in FA, and (**C**) choroidal hyperpermeability in ICG. Left eye presents (**D**) an exudative RD on colour fundus, (**E**) associated to chronic epitheliopathy showed as hypofluorescent areas in FA and (**F**) hypocyanescent areas in all phases of ICG. Hypercyanescent pinpoint spots appearing in late phases of ICG represent immune deposit at the level of choroidal stroma, Bruch membrane or retinal pigment epithelium basement membrane that contain immunoglobulins and leukocytes, which bind the ICG molecule.

To date, the literature yields approximately 40 cases (4). Lupus choroidopathy often occurs concomitantly with severe systemic manifestations of SLE (5). Nguyen *et al*. reported that 100% of their 28 patients had active systemic vascular disease at the onset of their choroidopathy (5), showing that choroidopathy is an indicator of severe underlying systemic disease. Conversely, in our case, when choroidopathy was diagnosed, there was no sign of nephropathy or central nervous system vasculitis. In the literature, another case of lupus choroidopathy with concomitant RD preceding the systemic manifestation of LES was described (6), but no imaging evidence of choroidal involvement was demonstrated. Since this article was published, new multimodal imaging has demonstrated choroidal vessel inflammation (4). In our case we were able to demonstrate choroidal involvement by ICGA. Although the pathogenesis of lupus choroidopathy is still unclear, it seems to be an inflammatory process related to immune complex deposition at the choroid, presence of autoantibodies against RPE and thrombotic microangiopathy resulting in hypoperfusion of the choroid, RPE damage, and exudative RD (4). For this reason, multimodal imaging (ICG and OCT) should be used by the ophthalmologist each time choroidal inflammation needs to be investigated, and choroidopathy should be considered by the rheumatologist as an active sign of systemic lupus, similarly to nephropathy or cerebral involvement. This provides an opportunity for an effective intervention before permanent life-altering or even lethal damage has occurred.

B. Iaccheri[1], *MD*
A. Cerquaglia[1], *MD*
E. Bartoloni[2], *MD*
C. Cagini[1], *MD*
T. Fiore[1], *MD*

[1]Department of Medicine, Ophthalmology Section, University of Perugia, S. Maria della Misericordia Hospital, Perugia; [2]Rheumatology Unit, Department of Medicine, University of Perugia, Perugia, Italy.

Please address correspondence to:
Barbara Iaccheri,
Dipartimento di Medicina,
Sezione di Oftalmologia,
Università di Perugia,
Ospedale S. Maria della Misericordia,
p.le Menghini 1,
Perugia 06129, Italy,
E-mail: barbaraiaccheri@gmail.com

Competing interests: none declared.

References

1. PAPAGIANNULI E, RHODES B, WALLACE GR, GORDON C, MURRAY PI, DENNISTON AK: Systemic lupus erythematosus: an update for ophtahlmologists. *Surv Ophthalmol* 2016; 61: 65-82.
2. HOCHBERG MC: Updating the American college of rheumatology revised criteria for the classification of systemic lupus erithematosus. *Arthritis Rheum* 1997; 40: 1725.
3. PETRI M, ORBAI AM, ALARCON GS *et al*.: Derivation and validation of the Systemic Lupus International Collaborating Clinics classification criteria for systemic lupus erythematosus. *Arthritis Rheum* 2012; 64: 2677-86.
4. HASANREISOGLU M, GULPINAR IKIZ GD, KUCUK H, VARAN O, OZDEK S: Acute lupus choroidopathy: multimodal imaging and differential diagnosis from central serous chorioretinopathy. *Int Ophthalmol* 2018; 38: 369-74.
5. NGUYEN QD1, UY HS, AKPEK EK, HARPER SL, ZACKS DN, FOSTER CS: Choroidopathy of systemic lupus erythematosus. *Lupus* 2000; 9: 288-98.
6. CARPENTER MT, O'BOYLE JE, ENZENAUER RW, ENZENAUER RJ, WATERHOUSE WJ: Choroiditis in systemic lupus erythematosus. *Am J Ophthalmol* 1994; 117: 535-6.