CLINICAL IMAGE

Full visual acuity may be misleading in accompanying antiphospholipid syndrome: a case of bilateral fovea-sparing retinopathy in a patient with systemic lupus erythematosus

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We present the case of a 41-year-old Caucasian woman with established accompanying antiphospholipid syndrome (APS) in the course of systemic lupus erythematosus (SLE). The diagnosis of APS / SLE was based on clinical findings (neuropsychiatric: seizures and transient ischemic attack; renal: proteinuria and thrombotic microangiopathy on kidney biopsy) and immunological findings (presence of anti-double-stranded deoxyribonucleic acid antibody, anti-Smith antibody, lupus anticoagulant, as well as anti-beta-2 glycoprotein and anti-cardiolipin antibodies, low complement component 3). These findings were consistent with both the revised Sapporo APS Classification Criteria and the European League Against Rheumatism / American College of Rheumatology Classification Criteria for SLE.1,2 The patient was primarily treated with warfarin, glucocorticoids, and chloroquine.3,4 The treatment with the latter became eventually the reason for the ophthalmological consultation. Ophthalmological examination showed full visual acuity in both eyes (20/20), normal intraocular pressure, good color vision in the right eye, and improper in the left. Anterior segments of both eyes were unremarkable. The results of other ophthalmological tests are shown in FIGURE 1A–1H. An electrophysiological evaluation was contraindicated due to epilepsy.

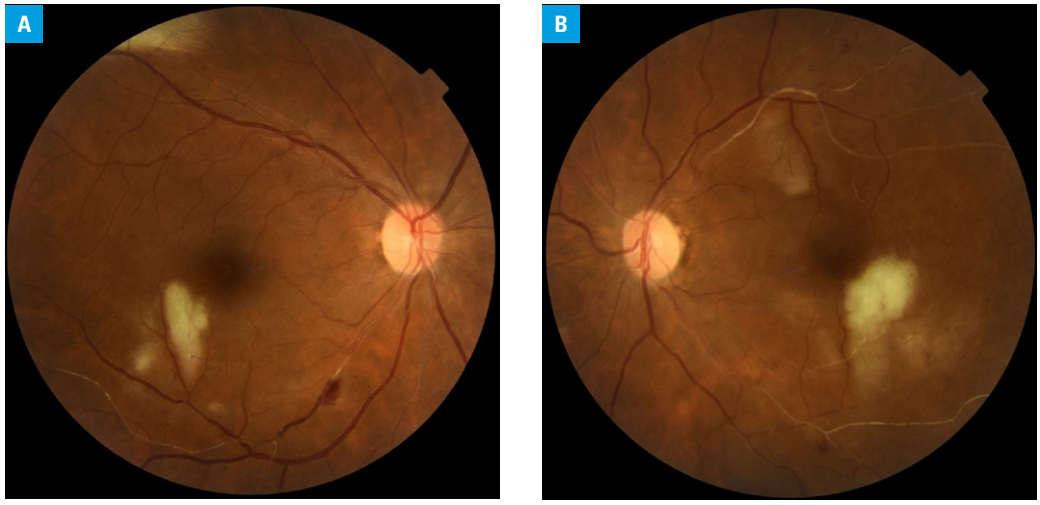

FIGURE 1 **A** – funduscopic findings: normal optic disc, occlusion of the inferotemporal branch of the central retinal artery ("ghost vessel"), intraretinal hemorrhages, occlusion of smaller peripheral vessels at 12 o'clock (not shown), and large cotton-wool spot inferotemporal from the fovea in the right eye; **B** – normal optic disc, occlusion of the supero- and inferotemporal branch of the central retinal artery ("ghost vessel"), intraretinal hemorrhages, occlusion of smaller peripheral vessels nasally from the optic disc (not shown), and large cotton-wool spots inferotemporal and superior from the fovea in the left eye

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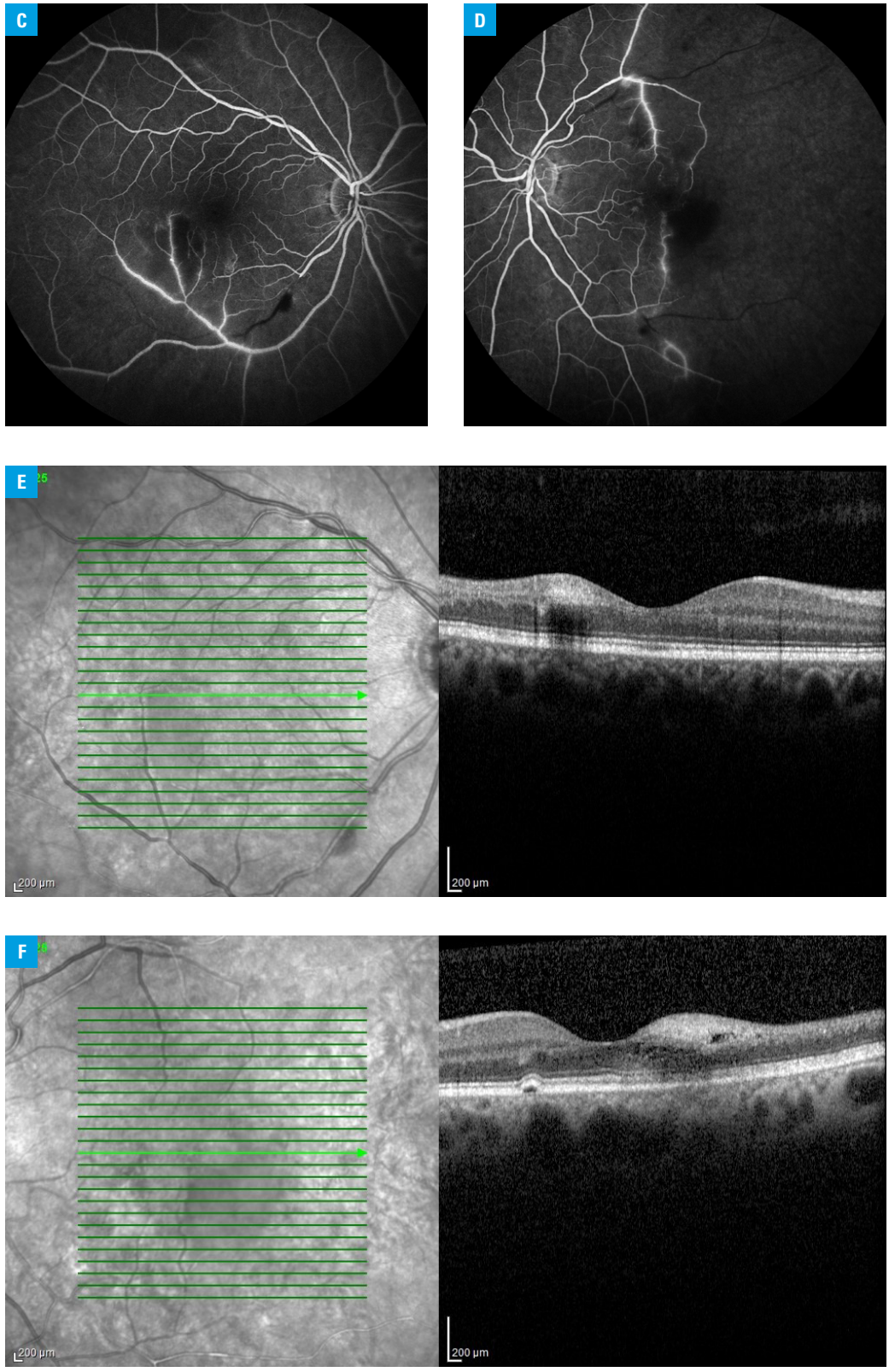
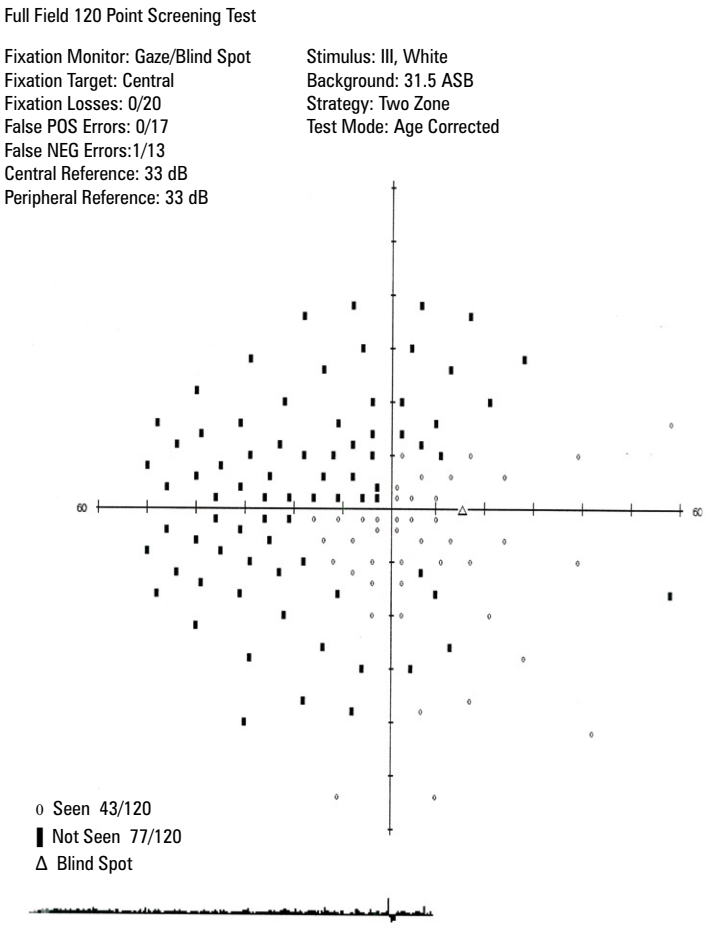


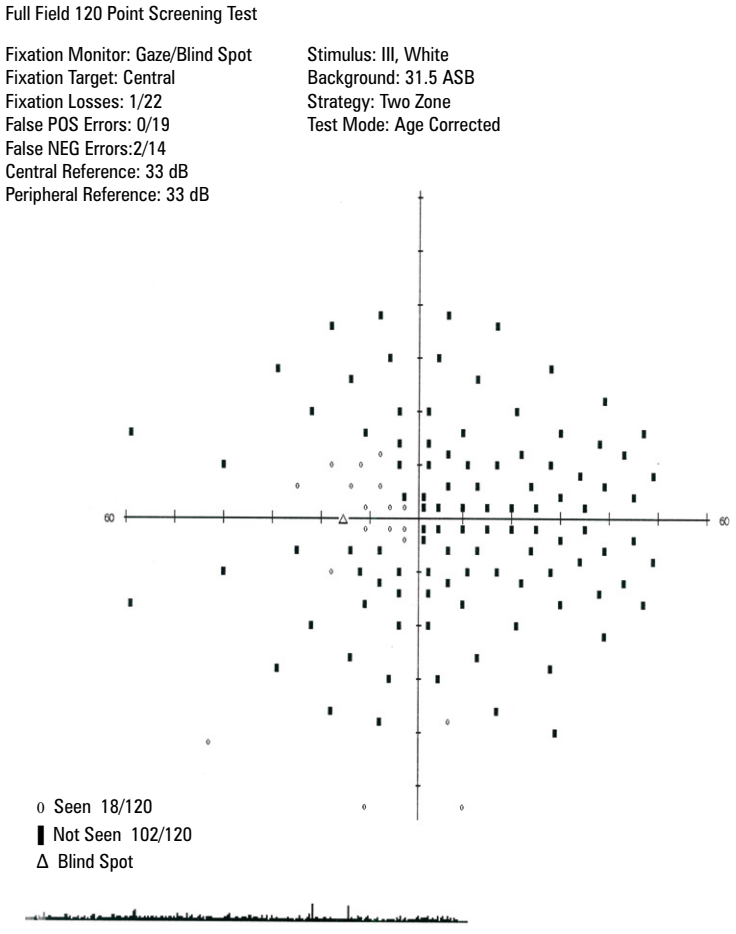
FIGURE 1 **C** – fluorescein angiography: occlusion of the inferotemporal branch of the central retinal artery with capillary nonperfusion and fovea sparing, and staining of the inferotemporal branch of the central retinal vein at 6 o'clock in the right eye; **D** – occlusion of the supero- and inferotemporal branch of the central retinal artery with prominent capillary nonperfusion and fovea sparing, and staining of the temporal branches of the central retinal vein at 12 and 6 o'clock in the left eye; **E** – optical coherence tomography: normal foveal profiles and thickening of the nerve fiber layers corresponding to the cotton-wool spots in the right eye; **F** – normal foveal profiles and thickening of the nerve fiber layers corresponding to the cotton-wool spots in the left eye

FIGURE 1 **G** – full-field 120 perimetry: multiple scotomas with fovea sparing, less prominent in the right eye; **H** – multiple scotomas with fovea sparing, more prominent in the left eye

G



H



Despite retaining full visual acuity, the patient presented with bilateral active severe retinopathy. The severe retinal damage was supposedly a manifestation of accompanying APS and vasculitis associated with SLE.5 Mycophenolate mofetil was initiated, warfarin and chloroquine continued, and the dose of glucocorticoids was gradually tapered down until withdrawal. Partial renal remission was achieved (a decrease in serum creatinine level and proteinuria, normal urine sediment). As the preexisting ischemia of the temporal quadrants was found in the left eye, the patient was scheduled for retinal photocoagulation.

In conclusion, a regular ophthalmological follow-up is warranted in patients with APS / SLE treated with chloroquine or glucocorticoids, as these drugs may produce side effects in the eye. Thromboembolic and / or vasculitic retinal damage, although still underrecognized, should be considered a sight-threatening condition.

Even with full visual acuity, a patient with APS / SLE may suffer from severe visual function loss and require prompt treatment by both rheumatologist and ophthalmologist.

ARTICLE INFORMATION

CONFLICT OF INTEREST None declared.

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