

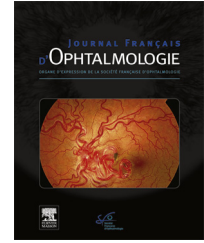


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LETTER TO THE EDITOR

Severe vaso-occlusive chorioretinopathy in a patient with catastrophic antiphospholipid antibody syndrome secondary to systemic lupus erythematosus



Vascularite occlusive chez une patiente avec un syndrome catastrophique des antiphospholipides secondaire au lupus érythémateux systémique

Introduction

Severe vaso-occlusive chorioido-retinopathy is a rare manifestation of catastrophic antiphospholipid antibody syndrome (CAPS) secondary to systemic lupus erythematosus (SLE) [1]. We present the case of a 20-year-old woman who had neurological, ocular, dermatologic and haematological manifestations of CAPS secondary to SLE.

Case report

A 20-year-old Caucasian woman was admitted initially to our hospital for epileptic seizure. She presented a Raynaud's phenomenon at the right hand, a livedo of the knees, a bilateral malar erythema and a holosystolic murmur. The neurological status was stable with no seizure; nevertheless, she had a poor attention, right hand paresis and an acute bilateral visual loss. Laboratory studies disclosed leucopenia, an increased erythrocyte sedimentation rate and a high proteinuria (3.4 g/24 h). Furthermore, the tests for anti-cardiolipin antibodies (IgG: 60 UI/ml) and lupus anticoagulant antibodies were positive (IgG: 34 UI/ml).

Moreover, the transthoracic echocardiogram showed the presence of small vegetations at the mitral valve, diagnosed as Libman-Sacks endocarditis [2]. The brain magnetic resonance imaging showed bilateral ischemic lesions in frontal and occipital subcortical areas. Hence, SLE and CAPS were diagnosed according to the revised American College of

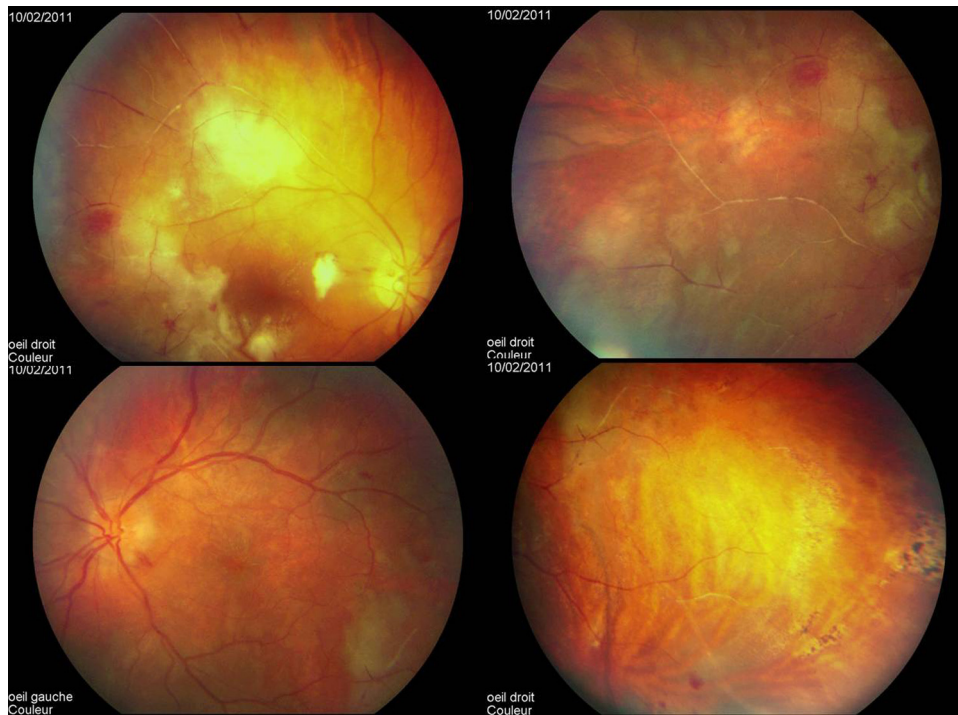


Figure 1. Color fundus photographs showed retinal vascular disease defined by the presence of cotton-wool spots, retinal hemorrhages, retinal vessels narrowing, retinal vessels cuffing and venous sausageing with alternating stenosis.

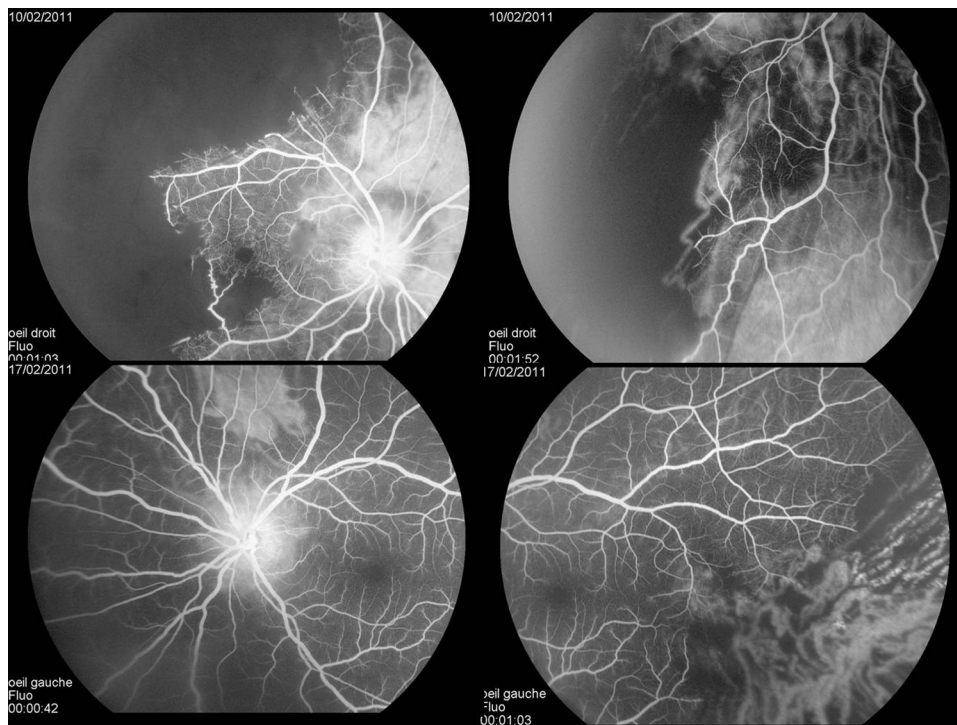


Figure 2. Retinal angiography disclosed severe retinal vaso-occlusive phenomena.

Rheumatology classification criteria [3]. A cortical blindness secondary to occipital ischemia was suspected in this patient with CAPS.

Ophthalmologic evaluation showed that her best corrected visual acuity (VA) was hand motion in the right eye and counting fingers at 30 cm in the left eye. The anterior segment and vitreous were clear. Fundus examination showed retinal vascular disease defined by the presence of cotton-wool spots, retinal hemorrhages, intraretinal edema and retinal vessels narrowing (Fig. 1). Fluorescein retinal angiography (FA) disclosed severe retinal vaso-occlusive phenomenon (Fig. 2). The indocyanine green (ICG) angiography showed a severe choroidal vaso-occlusive phenomenon. The optical coherence tomography (OCT) disclosed an ischemic edema of the retinal fibers layer (Fig. 3).

Hence, the visual loss was attributed, at least partly to the retinal vaso-occlusive phenomena secondary to SLE and CAPS.

At first, she was treated by high dose (500 mg a day) intravenous methylprednisolone pulses for three days followed by an antimalarial drug hydroxychloroquine 200 mg two times daily. Hence, we noted an important improvement of her neurological status. On second step due to the important chroidal and retinal ischemia, we started a rapid four sessions of scatter panretinal photocoagulation to prevent retinal neovascularization. The follow-up in our ophthalmologic department enabled us to note her VA increasing and avoid any complications of retinal ischemia. At the 1-month follow-up examination, her VA was 20/63 in the right eye and 20/100 in the left eye. Fundus, FA and ICG angiography showed improvement of the retinal perfusion (Figs. 4 and 5).

The OCT disclosed an ischemic atrophic lesion of the macula (Fig. 6). The six-month follow-up showed a stable VA and we did not disclose any ophthalmologic complication.

Discussion

SLE is a chronic inflammatory autoimmune disease that affects multiple organ systems and is characterized by prominent autoantibody production.

Ocular findings in SLE include anterior segment signs such as keratoconjunctivitis sicca, episcleritis and scleritis. Posterior segment manifestations of SLE include retinal microvasculopathy such as cotton-wool spots and retinal hemorrhages [4].

The antiphospholipid antibody syndrome (APS) is a thrombophilic disorder characterized by the association between recurrent arterial or venous thromboses, recurrent foetal losses and the presence of circulating antiphospholipid antibodies [5]. In rare cases, APS leads to rapid organ failure due to generalised thrombosis; this is termed CAPS and is associated with a high risk of death.

Our case of CAPS associated retinopathy, presented with all the features of lupus retinopathy, which is characterized by retinal capillary vasculitis associated with local microinfarction [6]. Such an involvement has been previously reported to reflect systemic vascular damage associated with the central nervous system (CNS) disease in SLE [7].

Since the ocular complications of SLE are generally associated with active disease elsewhere in the body, control

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