The importance of ophthalmological examination in erythematous Lupus: A case report and literature review

Abstract

Objective: Underline the importance of ophthalmological clinical examination in the management of Lupus.

Case report: A 25-year-old female with no previous medical history presented 15 days ago with erythematous, non-itchy, scaly maculo-papular lesions of the face in the form of a butterfly's wing, evolving in a febrile context with altered general condition, constipation and memory problems. The diagnosis of systemic lupus erythematosus was made on the basis of the clinical symptoms, biological results and the biopsy. Treatment was based on a synthetic antimalarial agent. An ophthalmological opinion was sought with a view to the introduction of synthetic antimalarials.

Clinical examination revealed corrected visual acuity of 10/10 in the right eye and 10/10 in the left. Biomicroscopy of both eyes showed infiltration of the lower eyelids, and examination of the retina revealed diffuse cotton wool spots around the vessels, a sharply excavated papilla rated 0.4 on the right and 0.3 on the left, good-caliber vessels without vascular sheathing, and good macular reflexion. Fluorescein angiography of both eyes showed no signs of retinal ischaemia (at the posterior pole and retinal periphery) or diffusion at any time during the examination. The patient was treated with hydroxychloroquine, dermocorticoids and local care for the skin lesions. The first month was marked by an improvement in skin signs, with stabilisation of the cotton wool spotson the fundus. However, dry keratoconjunctivitis appeared, leading to the prescription of a wetting agent and a vitamin A-based healing agent.

Keywords : Systemic lupus erythematosus, cotton wool spots, retinal damage.

Introduction

Lupus is a chronic systemic autoimmune disease in which ophthalmic involvement occurs in 3.3% to 28.1% of cases during the course of the disease [1]. The most frequent ophthalmological manifestation is adry keratoconjunctivitis. Occlusive retinal vasculitis in lupus is rare, acute in onset, and linked in almost 70% of cases to the presence of anti-phospholipid antibodies. We report a rare case of early-onset retinal involvement in lupusand highlight the importance to carry out a thorough examination of any patient diagnosed with this disease.

Case Presentation

A 25-year-old female patient with no previous pathological history presented 15 days ago with non-itchy erythematous maculo-papular scaly lesions of the face in the shape of a butterfly's wing (Image 1), evolving in a febrile context with altered general condition, constipation and memory problems.

In view of her symptoms, the patient was admitted to internal medicine, where a laboratory work-up was performed. An inflammatory syndrome was observed, C-reactive protein: 25.3 , normocytic normochromic anaemia : Haemoglobin: 8.5 g/dl, Leukocytes = 2600, Neutrophils = 1400, Lymphocytes = 900. Anti-SSA antibodies: positive, Anti-nuclear antibodies: positive with a speckled nucleus. However, anti-native DNA, anti-SSB, anti-cardiolipin and lupus-type anticoagulant antibodies all came back negative. A skin biopsy was also performed, which came back in favour of lupus. A diagnosis of systemic lupus erythematosus was made, and treatment with synthetic anti-malarial drugs was adopted. An ophthalmological opinion was sought in view of the introduction of synthetic antimalarials.

Clinical examination revealed corrected visual acuity of 10/10 in the right eye and 10/10 in the left. Biomicroscopy of both eyes showed infiltration of the lower eyelids, clear conjunctiva, clear cornea with no fluorescein uptake, deep optically empty anterior chamber, good iris trophicity and colouration with no iris nodule, The lens and vitreous were transparent. Examination of the retina (Images 2 and 3) showed the presence of diffuse Cotton wool spots around the vessels, a papilla with a clear excavation edge rated at 0.4 on the right and 0.3 on the left, vessels of good caliber with no vascular sheathing, and a good macular reflexion.

Fluorescein angiography of both eyes showed no signs of retinal ischaemia (at the posterior pole and retinal periphery) or diffusion at any time during the examination,

An angiographic Optical coherence tomography (OCT A) (Image 4) centred on a lesion was performed and showed Cotton wool spots(hyper-reflective intra-retinal material and posterior shadow cone), and some ischemic territories surrounding the vessels.

The patient received hydroxychloroquine at a dose of 400 mg per day, dermocorticoids and local care for the skin lesions. The evolution was marked by an improvement in the cutaneous signs, with stabilisation of the Cotton wool spotsin the fundus. A dry keratoconjunctivitis appeared, leading to the prescription of a wetting agent and a vitamin A-based healing agent.

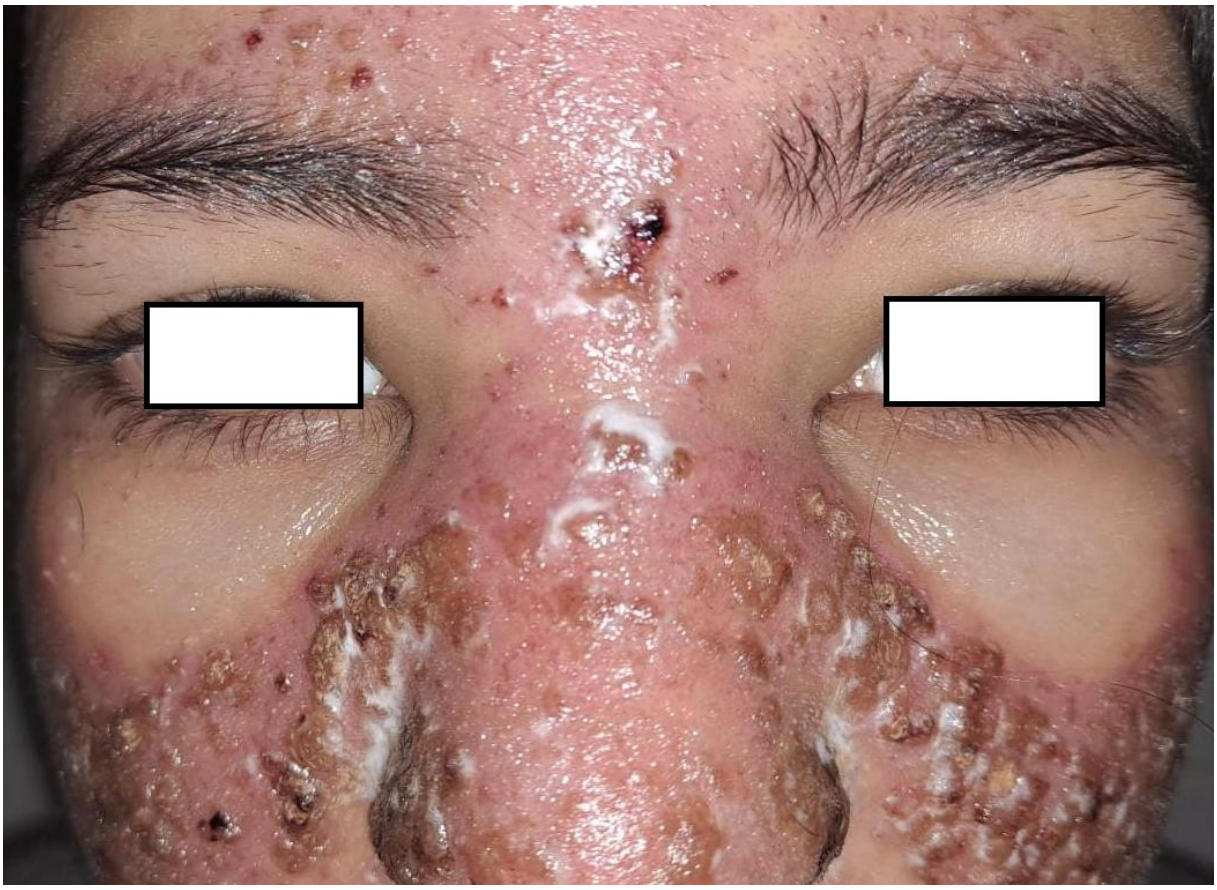

Image 1: non-itchy, erythematous, scaly maculo-papular lesions of the face in the shape of a butterfly wing, with infiltration of the eyelids.

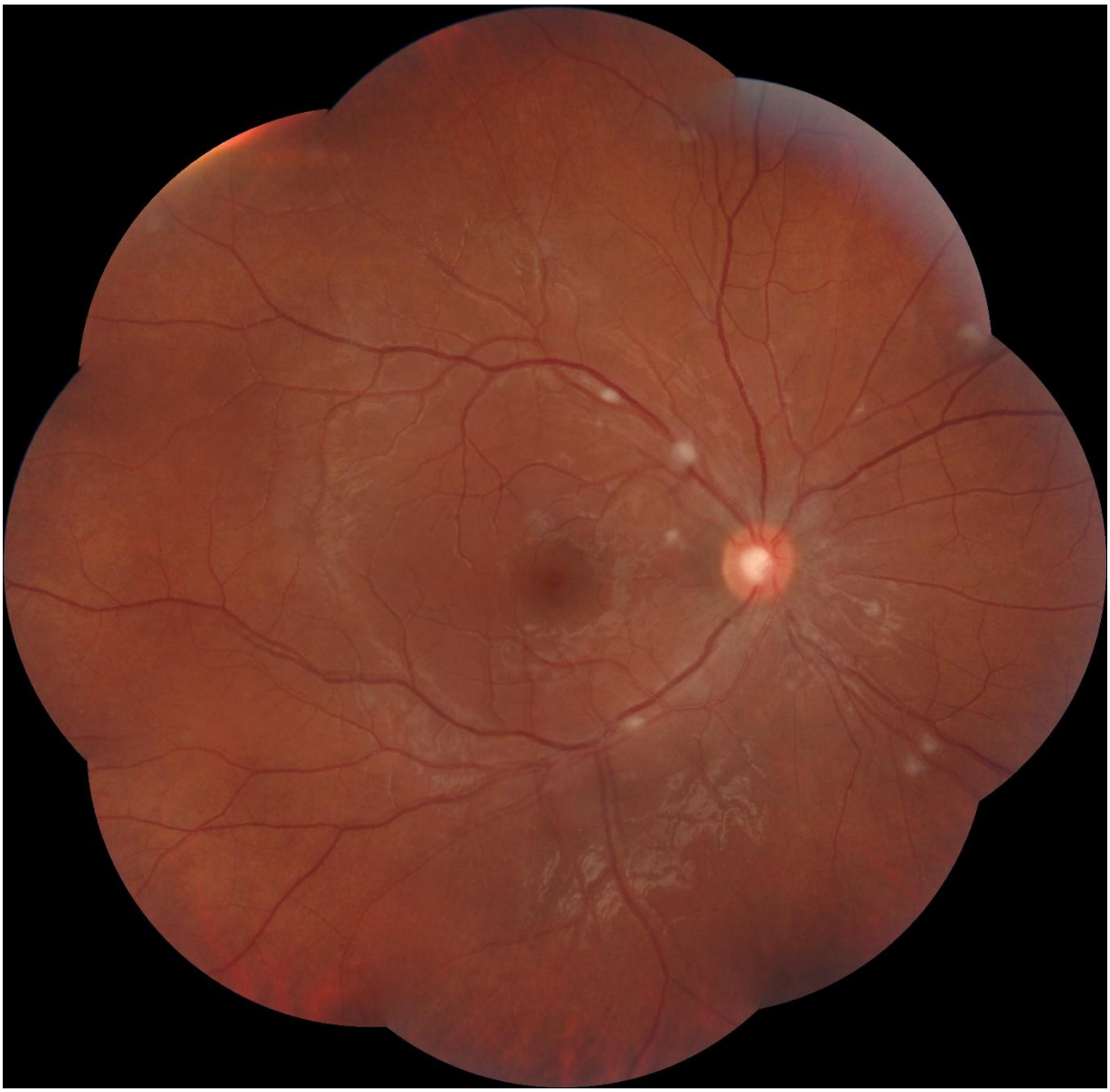

Image 2: Retinophotography of the patient's right eye showing a papilla with clear contours, excavation at 0.4, vessels of good caliber, presence of Cotton wool spots on the vascular paths, good macular reflexion.

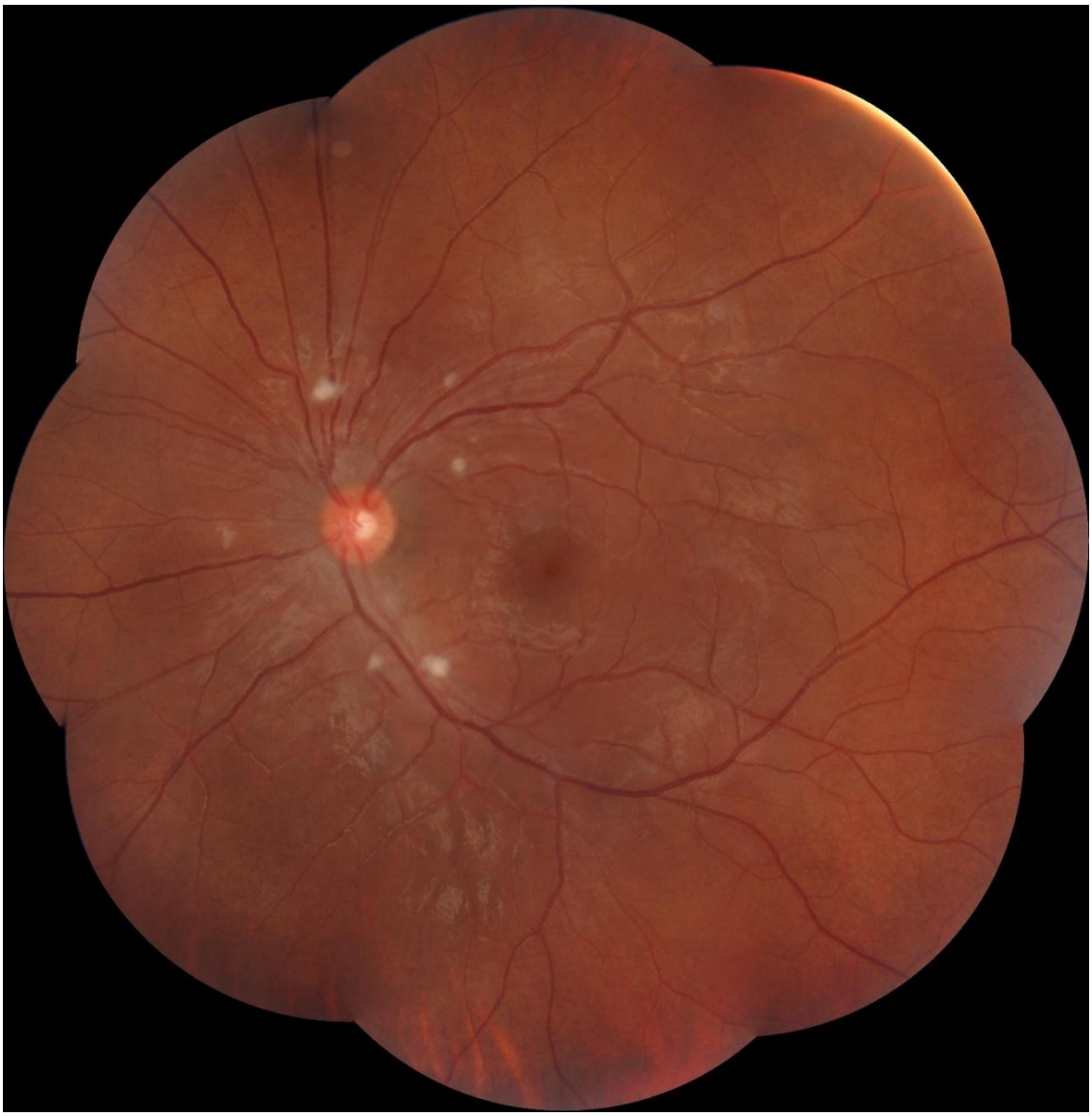

Image 3: Retinophotography of the patient's left eye showing a papilla with clear contours, excavation at 0.3, vessels of good caliber, presence of Cotton wool spots on the vascular paths, good macular reflexion.

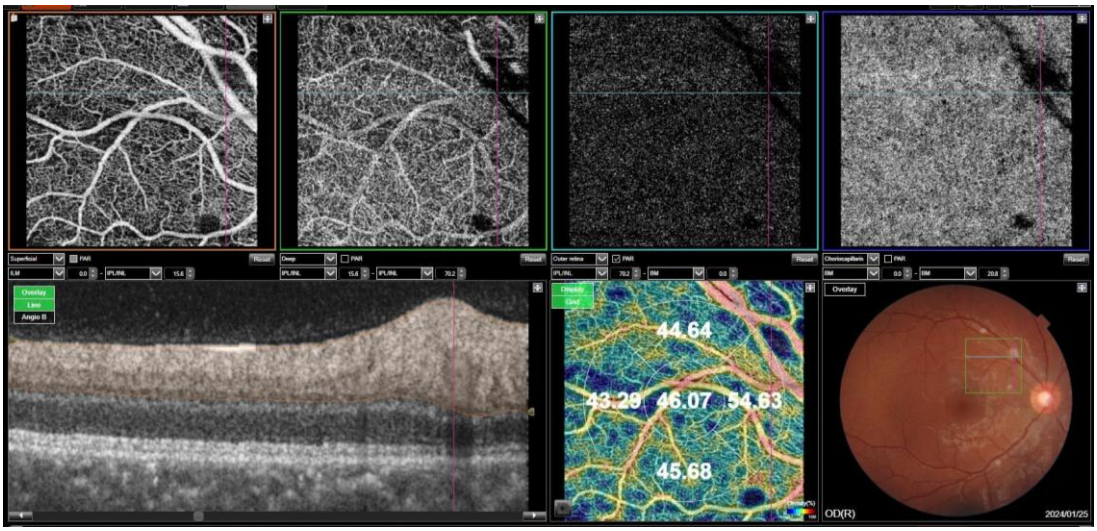

Image 4 :OCT angiography of the right eye centred on a cotton wool spots showing thickening of the inner layers of the retinaand some ischemic territories surrounding the vessels.

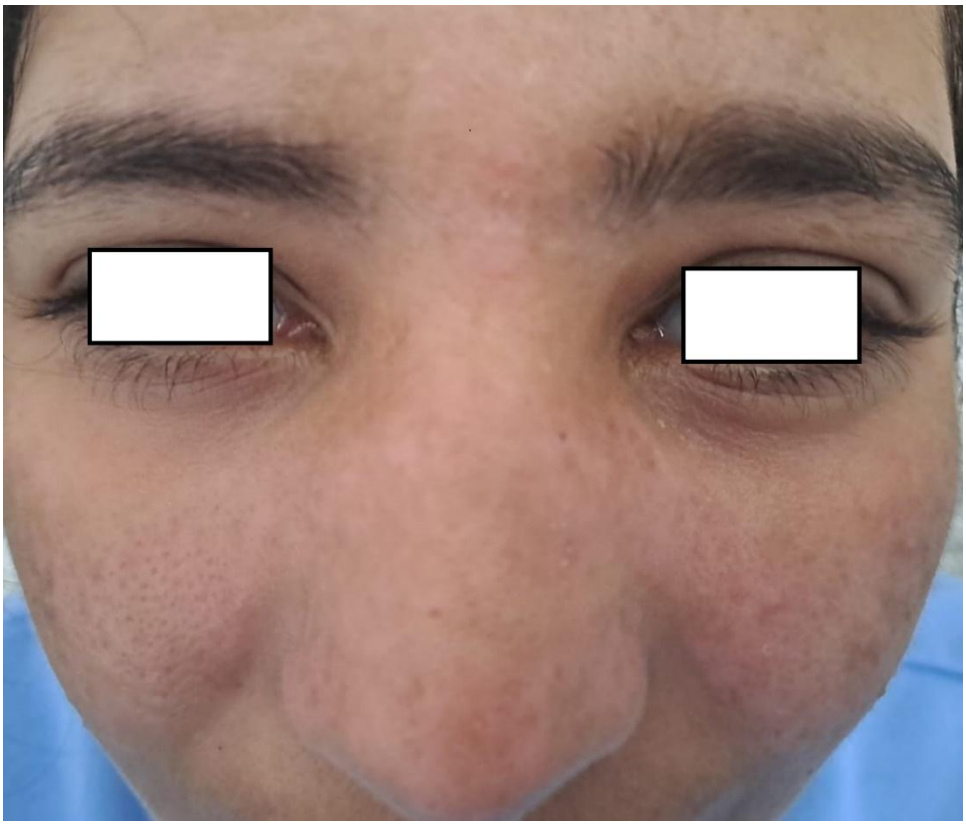

Image 5: Resolution of skin lesions after one month of treatment.

Discussion

Systemic lupus erythematosus (SLE) is an autoimmune disease in which retinal damage is an ophthalmological emergency because it affects visual prognosis, hence the need to intervene earlier in order to preserve visual function. The incidence of retinal damage in SLE ranges from 3.3% to 28.1% [1, 2], while the incidence of vasculitis is lower.

The pathogenesis of retinal damage is uncertain, but the effect of circulating immune complexes on retinal vessels has been suggested [3]. A complex interaction between genetic, hormonal and environmental factors is thought to be involved. The most common findings are diffuse and extensive arteriolar occlusions, capillary non-perfusion, severe retinal ischaemia and subsequent neovascularisation. In the majority of these cases, both eyes were affected, as in our patient. The pathogenesis of vaso-occlusive retinopathy in SLE is thought to involve an immune complex-mediated vasculitis. Histological studies have revealed fibrinoid changes with thrombosis in the vessel walls [4].

Lupus can affect all organs. The most recent diagnostic criteria have been formulated by the Systemic Lupus International Collaborating Clinics (SLICC) [21]. In the eye, all structures can be affected [20].

Damage to the microcirculation with occlusion of the precapillary arterioles, giving rise to the classic dysoric nodules or "cotton wool spots". Thus we emphasise the importance of OCT A, the only examination that can reveal microcirculation damage.The cause may be damage to the vascular walls, a change in blood viscosity or an embolism. They are characterised by the presence of cotton wool spots, as in our patient, and flaming haemorrhages, which generally have no effect on visual acuity. Other signs of lupus vasculitis: deep dry exudates; superficial retinal haemorrhage; microaneurysms; accentuation of arteriovenous crossings; narrowing of retinal arterioles; localised or generalised oedema; venous stasis; exudative detachments [5]. Involvement of the large arterial and venous trunks is rarer [6]. Retinal venous occlusion or occlusion of the central retinal artery may be observed. Retinal vasculitis is linked in almost 70% of cases to the presence of anti-phospholipid antibodies [7], which were negative in our patient.

Retinopathy generally correlates with SLE disease activity [4,8] and the presence of cotton wool spots is generally associated with a good prognosis for vision. In contrast, the severe vaso-occlusive retinopathy of lupus is a rare form of retinopathy, which tends to involve smaller retinal arterioles and has a much more profound effect on vision [10].the prevalence of vascular occlusion is thought to increase in the presence of anti-phospholipid antibodies [11] , but it can occur in the absence of these antibodies [4] . The natural evolution of ophthalmological damage could be towards occlusive vasculitis [9] with retinal ischaemia, with the risk of neovessels developing [12], leading to blindness [18, 19]. Even after treatment, remodelling of the retinal vasculature may occur. This type of vasculitis is rare and acute in onset, and may be accompanied by cerebral vasculitis, which can be life-threatening

[13]. Ocular imaging such as fluorescein angiography and macular OCT are necessary to monitor retinal damage [22].

Visceral damage, particularly to the eyes, in the course of lupus requires corticosteroid therapy (methyl prednisolone) in bolus form for three consecutive days, followed by a dose of prednisone 1mg/kg/day, then gradual and cautious tapering off. Early treatment improves prognosis. In severe forms, immunosuppressive drugs will be offered, as they are becoming increasingly important in the management of lupus [14]. Earlier treatment would also avoid the need for aggressive therapy, as in our patient's case. Retinal panphotocoagulation is necessary in cases of ischaemic retinal neovascularisation. Central retinal vein occlusion is a known complication of systemic lupus [15,16]. In oedematous forms of central retinal vein occlusion, intravitreal injections have proved effective [17].

Conclusion

The role of the ophthalmologist is necessary in the follow-up of a lupus patient, both for the disease itself and for monitoring retinal damage caused by treatment with synthetic antimalarials. Larger-scale studies will be needed to assess the efficacy of synthetic antimalarials in very early ophthalmological forms of the disease.

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

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