Vaso-occlusive retinopathy by systemic lupus erythematosus associated with the antiphospholipid antibody syndrome

Retinopatia vaso-oclusiva por lúpus eritematoso sistêmico associada à síndrome do anticorpo antifosfolipídeo

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ABSTRACT

To report the case of a patient with vaso-occlusive retinopathy due to systemic lupus erythematosus (SLE) associated with antiphospholipid antibody syndrome (APAS), which started with signs and symptoms of autoimune hemolytic anemia accompanied by sudden monocular visual loss. Few cases of SLE manifestation primarily involving ocular changes have been reported in the Brazilian and international literature. Screening for antiphospholipid antibodies is of the greatest importance for patients with lupus retinopathy, so that immediate therapy with anticoagulants may be instituted in order to prevent vascular thrombosis, which worsens the visual prognosis.

Keywords: *Lupus erythematosus, systemic; Antiphospholipid syndrome; Antibodies; Anemia, hemolytic, autoimmune; Case reports*

RESUMO

Relatar um caso de paciente com Retinopatia vaso-oclusiva por Lúpus Eritematoso Sistêmico (LES) associado à Síndrome do Anticorpo Antifosfolipídeo (SAF), que se iniciou com um quadro de anemia hemolítica autoimune acompanhado por baixa visual súbita monocular. Poucos casos foram descritos na literatura nacional e mundial em que o LES se manifeste primeiramente com alterações oculares. O screening dos Anticorpos antifosfolípideos (APAs) é de suma importância para pacientes com retinopatia lúpica para que seja instituída a terapia imediata com anticoagulantes como forma de prevenir a trombose vascular, o que piora o prognóstico visual.

Descritores: Lúpus eritematoso sistêmico; Síndrome antifosfolipídica; Anticorpos; Anemia hemolítica autoimune; Relatos de casos

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Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune, multisystemic disease which may affect any part of the human body, including the eye. SLE affecting the eye may reflect the systemic activity of the disease, and thus take to the search of other body systems that might have been affected.[1]

The Antiphospholipid Antibody Syndrome (AAS) is an autoimmune disease defined by the presence of antiphospholipid antibodies (APA) along with a clinical manifestation of the disease: arterial/venous thromboses and recurrent fetal loss.[2] Despite the recurrent condition of arterial and/or venous thrombosis, other findings may be seen, such as: false VDRL and thrombocytopenia.[3] APA affect the segments of the blood vases over the body and may lead to hypercoagulability.[4] Ocular affection is most frequently manifested with the occurrence of retinal thrombosis, mainly among the youngsters.[5]

According to recent studies, AAS occurs in 34% 42% of SLE patients.[6] Retinal vascular occurrence will depend on the patient having or not APA associated to SLE.[6] As an example, retinal vascular occlusion is more frequent in APA patients (13.9%) than in those without APA (0.9%). Occurrences with extra ocular thrombosis in lupus patients with APA were 69.2% of cases compared to 22.8% without APA.[7]

SLE complications developing to a worse visual prognosis include occlusion of the central retinal artery occlusion (CRAO), central retinal vein occlusion (CRVO), retinal displacement (RD), vaso-occlusive retinopathy, and occurrence of the optic nerve with isquemic optic neuropathy and optic neuritis.[1]

The presence of APA is a risk factor for a worse ocular prognostic.[8]

The objective of the present study is to report the case of a patient with vaso-oclusive retinopathy by SLE associated to AAS with the first manifestation being sudden low vision of the left eye associated to symptoms characteristic of autoimmune hemolytic anemia. The patient was admitted to the Rheumatology Department of Hospital das Clínicas (HC) of Goiânia to investigate the cause of hemolytic anemia along with the investigation by Centro de Referência em Oftalmologia (CEROF) to evaluate sudden visual loss, with the correct diagnosis and anticoagulation being performed.

Case report

Female patient, 24 years old, white, from the State of Goiás, during her second day of hospitalization at the Rheumatology service of HC de Goiânia, requested the ophthalmology service of the hospital to assess her complaint of sudden low vision of the left eye since the first day of hospitalization. She was hospitalized due to a strong asthenia for a week, along with epigastric pain. She had isolated episodes of fever (37.6°C), besides dark-color urine and light feces. She was investigating a condition of autoimmune hemolytic anemia. She denied having thrombosis of other systems, and had never tried to get pregnant. She received a transfusion of 2U of red blood cells bag two months before, and presented jaundice 3+/4+. Her family history included one sister with AAS and amaurosis fugax for four years and one cousin with SLE. The physical examination of the abdome showed hepatosplenomegaly and at ectoscopy only yellowish sclera. Biomicroscopy and intraocular pressure of both eyes were normal. The fundoscopy of the right eye (RE) showed intra-retinal hemorrhages and associated roth spots, and the left eye (LE) showed pre and intra retinal hemorrhages in the 4 quadrants and peridiscal candle flame-shaped hemorrhage, in addition to diffuse roth spots. Corrected VA was RE 1.0 and LE 0.1. We requested colored (Figure 1) and fluorescent (Figure 2) retinography, OCT (OpticalCoherenceTomography) of the macula (Figure 3) as soon as she came to the service.

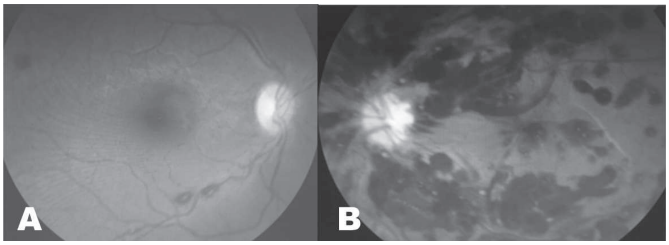

Figure 1: Retinography of the right eye (A) and left eye (B).

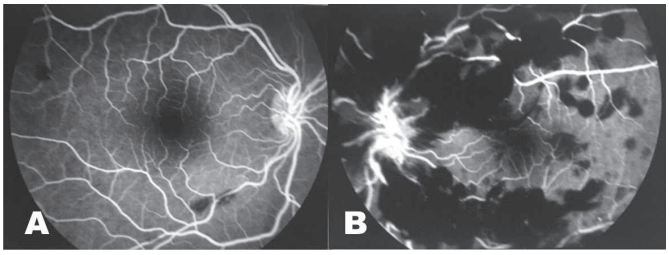

Figure 2: Fluorescent retinography of the right eye (A) and left eye (B)

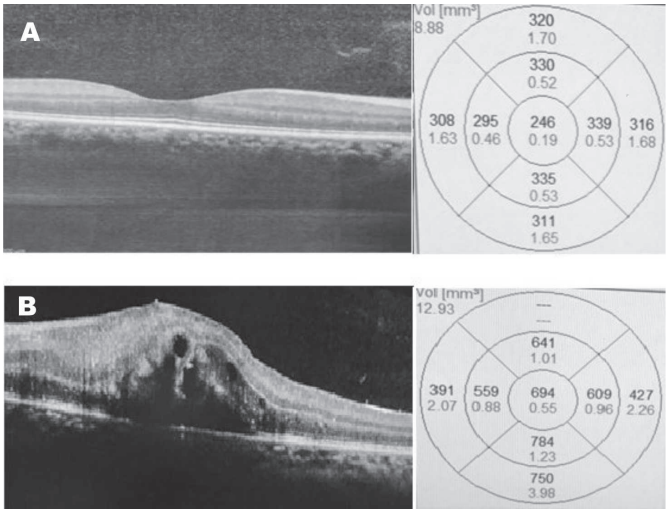

Figure 3: OCT of the macula of the right eye (no changes) and left eye with macular edema, entra-retinal cysts and hyperreflective points compatible with exsudates, respectively.

Laboratory exams carried out at the Rheumatology service showed anemia (Hb: 10.3 g/dL), thrombocytopenia (143,000), positive direct coombs, anticardiolipin (IgM = 70.01), lupus anticoagulant (1.6), VDRL: 1/64 and FtA-Abs negative (false positive), hematic cylinders in the urinary sediment. Besides that, after the fifth day of hospitalization the patient complained of arthralgia and alopecia. Thus, laboratory and clinical tests proved the presence of SLE associated to AAS. The patient was subjected

to pulse therapy since the first day of hospitalization with 1g of methylprednisolone/day for 3 days for hemolytic anemia, and then continued with prednisone (1g/kg) in immunosupressive dose. Besides, oral therapy with Warfarin 5 mg 1x/day was used as anticoagulation for AAS and hydroxychloroquine 400mg/day for SLE. After a week, as there was no improvement of the VA and macula edema seen in the OCT of the LE, 10 mg of subtenonian triamcinolone were precribed. Ten days after the subtenonian injection of triamcinolone, the VA of the LE changed to 0.2 (improvement of 2 lines in the Snellen table), and a new colored retinography was performed (Figure 4), showing changes only in the left eye with areas of subtenonian hemorrhage.

The condition was followed up, two months later the macula edema was completely cured in the OCT (Figure 5), and the VA of the LE evolved to 0.6.

In addition, the hemorrhage areas of both eyes disappeared. Anticoagulation was kept with warfarin 5mg/day, hydroxychloroquine 400 mg/day and dose regression of prednisone to 10 mg/day to estibilize the disease from the systemic point of view.

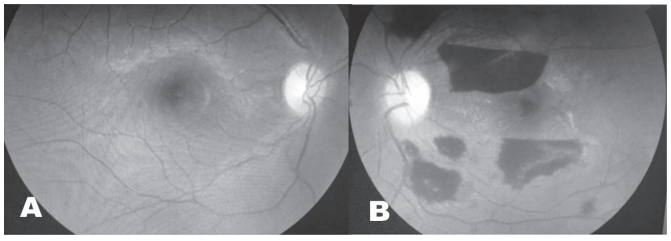

Figure 4: Retinographies of the right (A) and left (B) eyes 10 days after the subtenonian injection of triamcinolone.

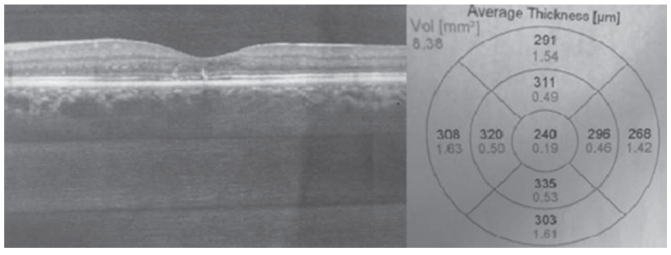

Figure 5: OCT of the macula of the left eye (with the cure of the condition).

Discussion

The incidence of SLE varies from 1.8 to 20 or more cases per 100,000 individuals per year. Of these patients, from 80% to 90% are childbearing-age women of about 30 years of age, as in the case described.[9]

SLE may affect various structures of the eyes and annexes, with no preference for the anterior or posterior segment.[1] The ocular occurrence of SLE, despite less frequent, may be the first manifestation of the disease.[10] The most frequent retinal findings are: cottony exudates, retinal hemorrhages and vascular tortuosity.[11,12]

In the presence of antiphospholipid antibodies, there is a greater tendency to hypercoability of systemic blood vases and the onset of retinal thrombosis[5], as described in the patient studied. Such antibodies are also associated to a greater severity of retinopathy and vascular occlusion in the lupus patient.[2] Besides, Ermakovaet al. associated amaurosis fugax and essential hypertension to episodes of vascular occlusion in SLE.[7]

In the presence of antiphospholipid antibodies, anticoagulation with warfarin is important in the secondary prevention of new episodes of thrombosis, in addition to allowing a better visual prognosis. Thus, it must be done in long term.[13] However, aspirin and immunosuppressive agents do not yet have scientific evidence for such prophylaxis.[14]

Thus, in the case described, we preferred to institute the therapy only with warfarin 5mg per day, maintaining the medication even after the improvement of the patient's clinical and ocular condition as a way of avoiding recurrences.

Bajwa et al. report that in case of retinal vasculitis it is also necessary to use infusion of methylprednisolone as an emergency treatment. And also, there are patients in need of therapy with a daily dose of oral corticosteroids. And in case the oral corticosteroid therapy lasts a long time, immunosuppressive agents such as azathioprine and cyclophosphamide must be used.[15]

Subtenonian deposition corticosteroids (triamcinolone acetate) are widely used for the treatment of ocular inflammatory conditions which are refractory to topical and systemic corticoid treatment.[16]

The use of the subtenonian pathway leads to increased intraocular concentration of the drug by transcleral absortion, allowing the reduction of the systemic therapy and minimizing the side effects of the prolonged corticotherapy.[17]

The main indications for this route of administration of corticosteroids are the low visual acuity associated to chronic intravitreal inflammation and/or the presence of cystoid macular edema.[18] The improvement of visual acuity after this type of treatment varies from 65 to 85%,[17] and is an effective treatment for low visual acuity secondary to retinal vasculitis.[18]

In this case, subtenonian triamcinolone was used as the VA was stable to improve the macular edema, intra-retinal cysts, and above all the VA of the LE.

We conclude that the case report on the development of the clinical case and visual prognosis with the adequate therapy in patients with vaso-occlusive retinopathy for Systemic Lupus Erythematosus associated to the Antiphospholipid Antibody Syndrome helps improve the treatment of similar cases in the ophthalmology services in the country. Thus, it is extremely valuable to carry out studies on the subject, since the visual prognosis in patients with antiphospholipid antibodies associated to lupus retinopathy is still low.

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