CASE REPORT

UDC: 616-002.52::617.735
DOI: 10.2298/VSP150605136R

Severe vaso-occlusive retinopathy associated with systemic lupus erythematosus

Teška vazookluzivna retinopatija udružena sa sistemskim eritematoznim lupusom

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Abstract

Introduction. Systemic lupus erythematosus (SLE) is a systemic idiopathic autoimmune inflammatory disease, with multiple organ involvement. Severe vaso-occlusive retinopathy is a rare, sight threatening lupus-related manifestation of the disease, which is more common in patients with coexisting antiphospholipid syndrome. **Case report.** We reported a 36-year-old female with severe vaso-occlusive retinopathy that manifested in the absence of antiphospholipid syndrome. In a 4-year follow-up, despite aggressive systemic corticosteroid and immunosuppressive therapy and panretinal laserphotocoagulation treatment, the disease progressed to retinal neovascularisation, neovascular vitreoretinopathy, neovascular glaucoma and, consecutively, severe visual loss. As the final option for preservation of visual function, *pars plana* vitrectomy with laserphotocoagulation was performed and had good results. Progression of ophthalmological findings indicated the progression of the systemic disease, as well as neurolupus. **Conclusion.** Severe vaso-occlusive retinopathy occurred as the ophthalmological manifestation of SLE in the absence of antiphospholipid syndrome, but correlated with neurolupus and led to visual deterioration despite the treatment.

Key words:
lupus erythematosus, systemic; retinal diseases; retinal neovascularization; comorbidity; vitrectomy.

Apstrakt

Uvod. Sistemski *lupus erythematosus* (SLE) predstavlja sistemsko idiopatsko autoimunsko inflamatorno oboljenje, koje može zahvatiti brojne organe. Teška forma vazookluzivne retinopatije je retka oftalmološka manifestacija ove bolesti koja može ugroziti vid, a koja se češće javlja kod bolesnika sa udruženim antifosfolipidnim sindromom. **Prikaz bolesnika.** U radu je prikazana 36-godišnja bolesnica sa teškom formom vazookluzivne retinopatije koja je nastala u odsustvu antifosfolipidnog sindroma. Tokom 4-godišnjeg praćenja, uprkos agresivnoj sistemskoj kortikosteroidnoj i imunosupresivnoj terapiji i panretinalnoj laserfotokoagulaciji, došlo je do progresije bolesti i nastanka retinalne neovaskularizacije, proliferativne vitreoretinopatije i neovaskularnog glaukoma i, posledično, do slabljenja vida. *Pars plana* vitrektomija sa laserfotokoagulacijom bila je poslednja mera u očuvanju vidne funkcije i imala je dobre rezultate. Progresija očne bolesti ukazivala je na progresiju sistemske bolesti i neuroloških manifestacija lupusa. **Zaključak.** Teška forma vazookluzivne retinopatije nastala je kao oftalmološka manifestacija SLE u odsustvu antifosfolipidnog sindroma, a njena progresija bila je povezana sa pogoršanjem neurolupusa i dovela je do teškog slabljenja vida uprkos tretmanu.

Ključne reči:
lupus, eritematozni, sistemski; mrežnjača, bolesti; mrežnjača, neovaskularizacija; komorbiditet; vitrektomija.

Introduction

Systemic lupus erythematosus (SLE) is an idiopathic autoimmune inflammatory disease that can affect multiple organs. The main features of the disease include the presence of circulating autoantibodies to one or more components of cell nuclei, generation of circulating immune complexes and activation of complement system resulting in tissue destruction [1]. Severe vaso-occlusive retinopathy is a rare lupus-related manifestation with poor visual prognosis, more common in patients with coexisting antiphospholipid syndrome [1–3].

We reported a 36-year-old female with SLE and severe vaso-occlusive retinopathy that occurred in the absence of antiphospholipid syndrome.

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Case report

In April 2008, a 36-year old female was admitted to the Clinic due to a sudden loss of vision in her right eye. Visual acuity in the right eye was 0.5/60 and in the left 0.3. The patient had been diagnosed with SLE 6 years before according to the international criteria proposed by the American College of Rheumatology [4], and was treated with corticosteroids and antimalarial agents (chloroquine 250 mg *per* day). Ophthalmological examination revealed perivascular sheathing, extensive cotton-wool spots, intraretinal haemorrhages, retinal oedema and wide areas of vascular occlusion on the periphery of both eyes (Figure 1). Fluorescein angiography revealed bilateral complete obliteration of macular capillaries and widespread occlusion of small retinal vessels (Figure 2). According to the advice of the rheumatologist, three-day-course pulse dose of methylprednisolone (1000 mg *iv*), followed by oral prednisolone 80 mg *per* day were introduced, as well as intravenous cyclophosphamide 500 mg administered in two-week regimen during the first two months and then continued once a month. After the treatment, visual acuity improved to 2/60 in the right eye and 0.5 in the left.

Further investigations were performed at the Institute of Rheumatology. Laboratory analyses showed elevated erythrocyte sedimentation rate, leucopenia, thrombocytopenia and normal erythrocyte count, haemoglobin rate and biochemical analyses. In immunoserology, elevated titres of antinuclear antibodies and circulating immune complexes were present, while other analyses (anticardiolipin antibodies and lupus anticoagulant) were negative. Renal function was normal. Objectively, the patient had iatrogenic Cushing syndrome with corticosteroid-induced obesity, hirsutism, osteoporosis and hypertension. On chest radiography, small pleural adhesion was detected in the right side. Due to the risk of development of neurolupus, magnetic resonance imaging (MRI) of endocranium was performed and punctiform ischemic lesions were detected in the *corona radiata*.

One year later, large areas of retinal ischemia persisted in both eyes with high risk of retinal neovascularization. Laser photocoagulation was performed in order to impede formation of new vessels. However, despite the aggressive treatment, one year later disc neovascularization with preretinal haemorrhage in the right eye and neovascularization elsewhere (temporal and nasal periphery) in the left eye developed. Panretinal photocoagulation continued (Figure 3) and after the treatment, visual acuity was 1/60 in the right eye and 0.3 in the left one and intraocular pressure was within normal limits with no neovascularisation in iridocorneal angle.

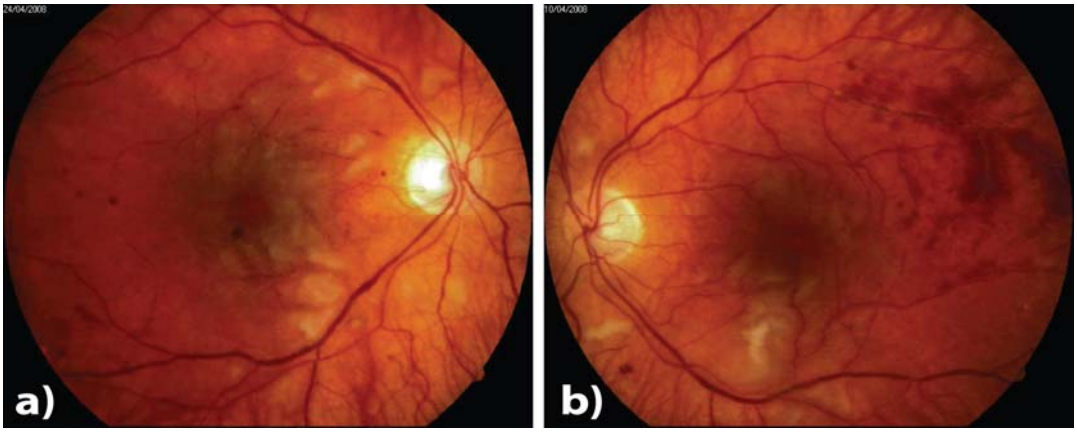

Fig. 1– a) Fundus colour photograph of the right eye showing vaso-occlusive retinopathy with cotton-wool spots and retinal oedema (April 2008), and b) Fundus colour photograph of the left eye showing vaso-occlusive retinopathy with intraretinal haemorrhages, cotton-wool spots and retinal oedema (April 2008).

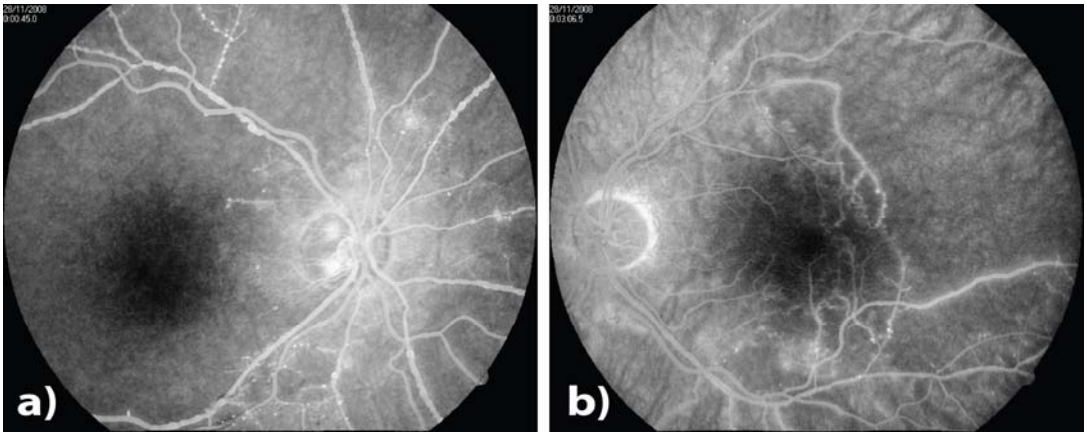

Fig. 2 – a) Fluorescein angiography findings of the right eye with diffuse avascular zones on the posterior pole and periphery of the retina (November 2008), and b) Fluorescein angiography findings of the left eye with wide avascular zones above and temporally of the macula (November 2008).

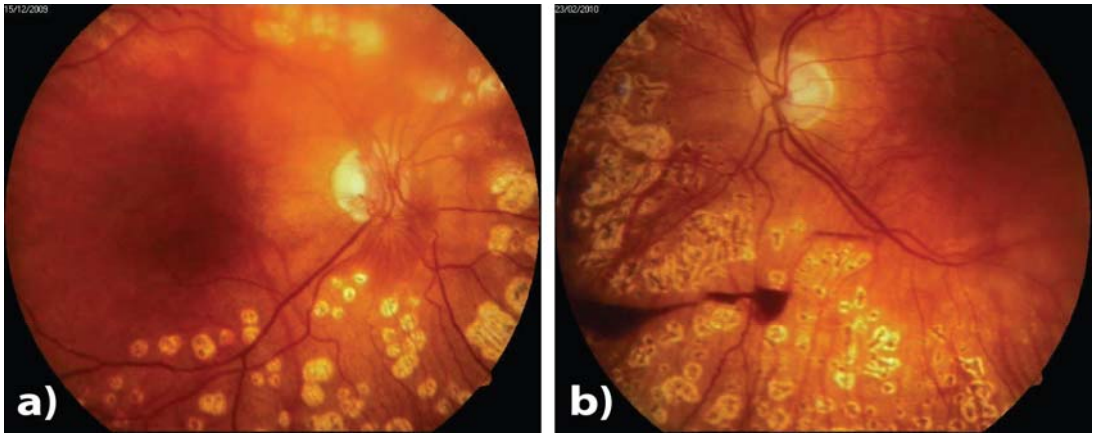

Fig. 3 – a) Fundus colour photograph of the right eye showing neovascularization at disc, intravitreal haemorrhage and scars after panretinal laserphotocoagulation (February 2010), and b) Fundus colour photograph of the left eye showing neovascularization elsewhere, preretinal haemorrhage and scars after laserphotocoagulation nasally and below the optic disc (February 2010).

Progression of the eye disease was noted three years later, with sudden visual loss in the single functional left eye. Visual acuity deteriorated to light perception bilaterally, due to dense vitreous haemorrhages. Also, neovascularisation in iridocorneal angle was noted in the right eye with intraocular pressure in 40 + mmHg range, which could not be controlled even with maximum medicamentous treatment. Vitreoretinal surgery (*pars plana* vitrectomy, endolaser and phaco) was performed in the left eye, because it was estimated that it has more visual potential. After the surgery visual function improved to 0.1 and was stabile for 18 months. During the follow up, repeated analyses for antiphospholipid antibodies were negative.

Discussion

SLE is an autoimmune inflammatory disease characterized by the numerous autoimmune phenomena and lesions in multiple organ systems [5]. Although eye findings are not listed as the criteria for SLE diagnosis, the thrombotic and inflammatory processes associated with SLE, can affect any part of the eye and result in manifestations such as *keratoconjunctivitis sicca*, scleritis, uveitis and retinal involvement [6]. Retinopathy in SLE is the second most common (after *keratoconjunctivitis sicca*) ophthalmological manifestation and usually is presented with cotton-wool spots, with or without intraretinal haemorrhages [2, 5, 6]. Depending on the study group, SLE retinopathy was reported to be present in 3.3–28.1% of all SLE patients, with the incidence rising at the advanced stages of the systemic disease [7, 8]. A severe variant of retinal vaso-occlusive disease is less common and may include central or branch retinal vascular occlusion or diffuse vaso-occlusive retinal disease [9–11] and the latter occurred in the presented case. Visual impairment is usually secondary to ischemic retinopathy and higher incidences were reported in association with antiphospholipid syndrome (APC) [1, 5]. However in the presented case, there were no evidences of this state.

Similar to the presented case, Mendrinos et al. [9] reported a case of bilateral combined central retinal artery and vein occlusion associated with normal levels of antiphospholipid antibodies and magnetic resonance imaging (MRI) confirmed central nervous system (CNS) vasculitis. The patient was treated with intravenous, followed by oral prednisone and cyclophosphamide, but unfortunately, despite all interventions, extensive vaso-obliteration led to permanent vision impairment. Also, Ho et al. [11] reported a case of bilateral severe SLE vaso-occlusive retinopathy in the absence of antihpospholipid syndrome that led to devastating vision deterioration. In addition, Zou et al. [10] reported a case of bilateral central retinal artery occlusion (CRAO) as the primary manifestation of with normal levels of antiphospholipid antibodies. Visual acuity deteriorated despite the systemic corticosteroid and immunosuppressive treatment [11].

Severe diffuse bilateral vaso-occlusive retinopathy as a complication of SLE, with the normal concentration of antiphospholipid antibodies is a rare complication in SLE. This type of retinopathy may be explained by the immune complex deposition, microthrombosis and endothelial damage in small blood vessels in the retina and choroid [2]. In the presented case, permanent loss of vision developed despite the treatment (systemic corticosteroid, immunosuppressive therapy and panretinal laserphotocoagulation). The patient's poor visual acuity along with bilateral optic nerve pallor could be attributed to posterior ischaemic optic neuropathy. Vitreoretinal surgery was the final option that impeded the retinal proliferative disease and improved the visual function. Furthermore, patient had vaso-occlusive retinopathy associated with similar changes in CNS and therefore had to receive the most aggressive systemic treatment, since it was shown that patients with neurolupus have the poorest prognosis in respect to survival rates [6].

Conclusion

Vaso-occlusive retinopathy is a rare lupus-associated complication and its presence is highly suggestive for SLE activity. It is usually, although not exclusively, a manifestation of antiphospholipid syndrome. Treatment should be

prompt with close communication with rheumatologists and aimed to prevent further thrombosis and complications arising from neovascularisation.

Conflict of interest

The authors declare no conflict of interest.

R E F E R E N C E S

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Received on June 5, 2015.
Revised on July 13, 2015.
Accepted on July 29, 2015.
Online First May, 2016.