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Case Report Open Access

Stellar Neuroretinitis Revealing Systemic Lupus Erythematosus without Antiphospholipid Syndrome

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Abstract

Introduction: Systemic Lupus Erythematosus (SLE) is an autoimmune systemic disease with multiple faces, secondary to auto-reacting antibodies targeting nuclear antigen. The optic nerve involvement is reported in less than 1% of SLE, dominated by optic neuritis and optic ischemic neuropathy. Neuroretinitis is defined as an inflammation of optic nerve and neural retina. We report a rare case of neuroretinitis as revealing form of SLE in young man.

Case report: 14 y old teenager boy, previously healthy, presented one month before his admission, rapidly progressive bilateral visual loss with no associated signs. Visual acuity evaluation revealed visual loss estimated to 2/10 right eye and 3/10 left eye with correction. Eye fundus objectified bilateral stellar macular with intermacular-optic disc exudates and moderate papillar pallor. The macular OCT found exudates in the plexiform layer of the retina. Other paraclinical test found bicytopenia with positive antinuclear and anti-DNA antibody; without antiphospholipid antibody. The patient underwent corticotherapy with favourable evolution.

Discussion: Neuroretinitis did not figure as usual cause of visual loss in SLE; moreover it has been very rarely reported as a revealing form of SLE. The exact pathogenesis behind neuroretinitis in SLE stays unknown. Sudden onset of unilateral painless loss of vision is the typical clinical presentation of neuroretinitis. Several etiologies may lead to neuroretinitis, dominated by infectious disease (bartonellosis, borreliosis, syphilis, herpes, hepatitis, HIV, CMV, Varicelle, EBV, Toxoplasmosis, Tuberculosis). Also neuroretinitis may be idiopathic. Concerning therapy, no clear guidelines are reported in neuroretinitis occurring in SLE. The visual prognosis is excellent with above 90% cases achieving a final visual acuity.

Conclusion: We learn from this case, that neuroretinitis may be the revealing form of SLE. This suggest the need of revision of SLE criterion, especially that neuroretinitis is a part of severe form.

Keywords: Neuroretinitis; SLE; HIV

Introduction

Systemic Lupus Erythematosus (SLE) is an autoimmune systemic disease with multiple faces, secondary to auto-reacting antibodies targeting nuclear antigen. The result is multiple tissues damaging including ocular ones. The optic nerve involvement is reported in less than 1% of SLE [1-3], dominated by optic neuritis and optic ischemic neuropathy. While retinopathy incidence in SLE varies from 7-26 % [4]. Neuroretinitis is rarely reported over the literature, especially as a revealing form. Neuroretinitis is defined as an inflammation of optic nerve and neural retina. The first case was originally described as "stellar maculopathy" by Leber in 1916, than corrected to "stellar neuroretinitis" by Gass in 1970, by proving that disc edema precedes macular exudates. We report a rare case of neuroretinitis as revealing form of SLE in young man.

Case Report

14 y old teenager boy, previously healthy, presented one month before his admission, rapidly progressive bilateral visual loss. There was no associated signs (headache, seizure, ocular pain, diplopia, eye redness), as there was no extra neuro-ophtalmologic symptoms (fever, articular, cutaneous, digestive, cardiac, respiratory). There was no history of exposure to pets, cats or birds. Visual acuity evaluation revealed visual loss estimated to 2/10 right eye and 3/10 left eye with correction. We noted no abnormal ocular motility. Slit lamp examination found normal anterior segment. Eye fundus objectified bilateral stellar macula with intermacular-optic disc exudates and moderate papillar pallor (Figure1).

There was no sign of uveitis. Hence we conclude to typical neuroretinitis aspect. Visual field and color vision evaluation was initially hard to make regarding the visual loss. The macular OCT found exudates in the plexiform layer of the retina (Figure 2). Visual field showed arciform lack in the inferior part of the field (Figure 3).

The teenager was normotensive. Neurologic examination was normal. Complete count blood objectified bicytopenia (leukopenia to 3500/mm^3 and anemia to 8.8 g/dl). There was biologic inflammatory syndrome with ESR to 50 mm first hour and CRP to 32 mg/l. Thus, infectious etiology was suspected and several serologies were established (HSV, CMV, EBV, HIV, HEPATITIS B and C, Syphilis, Borreliosis, Toxoplasmosis, Bartonellosis,Tuberculosis); and went all

negative. Immunologic tests found positive anti DNA antibodies and antinuclear antibodies with high titres, associated to low C3 and C4 serum complement. Antiphospholipid antibodies went negative. Thus, we had enough criteria for systemic lupus erythematosus SLE without antiphospholipid syndrome. Patient was treated by bolus methylprednisolone (1 g/d for 3 days) followed by oral prednisolone (1 mg/kg/d for 6 weeks then progressive degression to 10 mg/d). Evolution was favourable with recovering visual acuity passing to 9/10 right eye and 8/10 left eye without correction; and no dyschromatopsia. Eye fundus showed total exudates regression with normal papilla aspect (Figures 4 and 5).

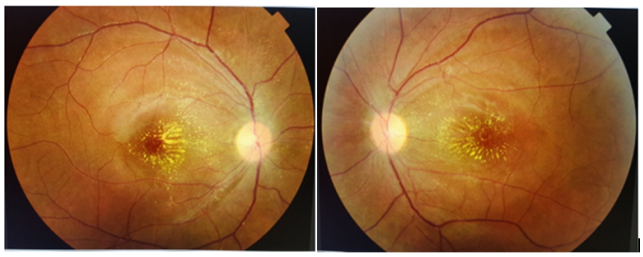

Figure 1: Eye fundus objectifying bilateral stellar macula with intermacular-optic disc exudates and moderate papillar pallor.

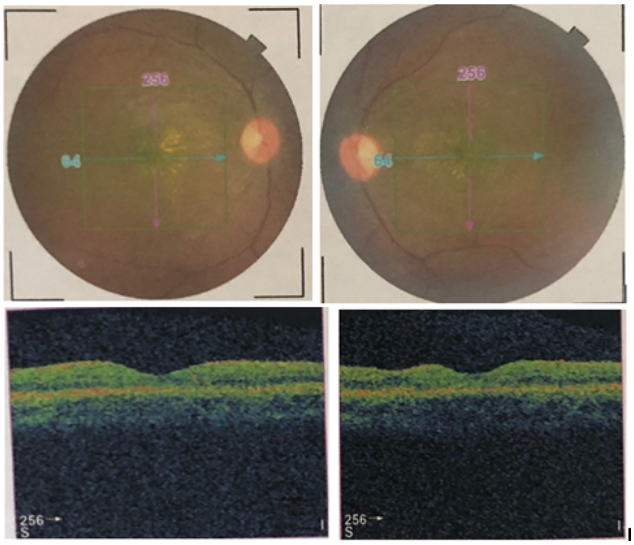

Figure 2: The macular OCT showing exudates in the plexiform layer of the left retina.

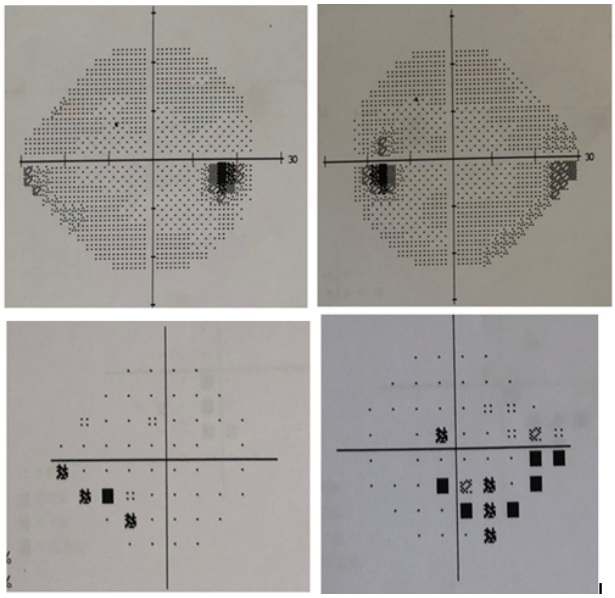

Figure 3: Visual field showing arciform lack in the inferior part of the field.

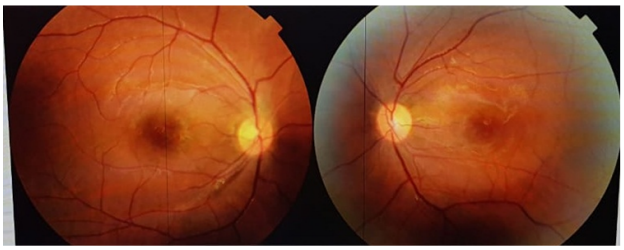

Figure 4: Eye fundus controle showing total regression of stellar macula following therapy.

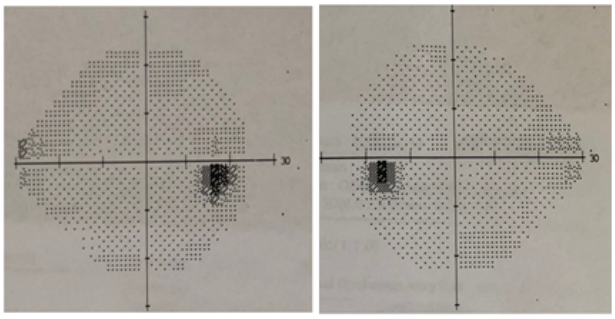

Figure 5: Visual field objectifying regression of the arciform lack.

Discussion

Ocular manifestations may occur in SLE. When the clinical presentation of SLE is visual loss, several ophtalomologic causes should be suspected (lens, vitreous, choroid, retina, optic nerve) (Table 1) [5].

Anterior segment	**Severe kerato-conjunctivitis sicca**
Lens	Cataract (Secondary to inflammation and/or corticosteroids)
Vitreous	Vitreous haemorrhage (Secondary to proliferative retinopathy)
Retina	Severe vaso-occlusive retinopathy
	Central retinal vein occlusion (CRVO)
	Branch retinal vein occlusion (BRVO)
	Central retinal arteriole occlusion (CRAO)
	Branch retinal arteriole occlusion (BRAO)
	Exudative retinal detachment
	Toxic maculopathy (secondary to anti-malarial treatment)
Choroid	Lupus choroidopathy
	Choroidal effusion
	Choroidal infarction
	Choroidal neovascular membranes
Neuro-ophthalmic	Optic neuritis
	Anterior ischaemic optic neuropathy
	Posterior ischaemic optic neuropathy
	Optic chiasmopathy
	Cortical infarcts

Table1: Causes of visual loss in SLE [5].

Neuroretinitis did not figure as usual cause of visual loss in SLE, moreover it has been very rarely reported as a revealing form of SLE [6,7]. Neuroretinitis is defined as an inflammation state of retina and optic nerve. Its physiopathology refers to optic disc edema that precedes by 1-3 weeks the appearance of stellar macula then resolving spontaneously 8-12 weeks later [8]. The leakage of optic disc vasculature lipoproteinaceous material and its accumulation in the outer retina layers are responsible of the optic disc swelling. This swelling subsides over weeks leaving behind radial deposits of lipoprotein in plexiform layer of retina [8]. Nevertheless, the exact pathogenesis behind neuroretinitis in SLE stays unknown. Sudden onset of unilateral painless loss of vision is the typical clinical presentation of neuroretinitis. Clinical examination objective decreased visual acuity with defect of visual field. Fundoscopy found optic disc swelling and hard stellar exsudates on macula [9]. Several etiologies may lead to neuroretinitis. They are dominated by infectious disease (bartonellosis, borreliosis, syphilis, herpes, hepatitis, HIV, CMV, Varicelle, EBV, Toxoplasmosis, Tuberculosis) (Table 2) [10-12].

Infections	**Autoimmune**
Bacteria: *Bartonella sp*. (*B. henselae, B. quintana, B. elizabethae, B. grahamii*), *Brucella, Mycobacterim tuberculosis, Rickettsia rickettsii* (Rocky Mountain spotted fever), *Salmonella* Protozoa: *Toxoplasma gondii* Spirochetes: *Borrelia burgdorferi* (Lyme disease), *Leptospira interrogans* (leptospirosis), *Treponema pallidum* (syphilis) Virus: Chikungunya, Cytomegalovirus, Coxsackie, Dengue, Epstein-Barr virus, Hepatitis B, Herpes Zoster Virus, Infuenza A, Measles, Mumps, Rubella, Varicella, West Nile virus Nematode: *Toxocara* Fungus: Coccidioidomycosis, Histoplasmosis	Sarcoidosis Ulcerative Colitis Polyarteritis nodosa Tubulointerstitial nephritis and uveitis (TINU) Systemic lupus erythematosus Antiphospholipid syndrome

Table 2: Causes of neuroretinitis [10-12].

Thus, multiple paraclinical tests should be performed to eliminate these infections. The second etiology is the autoimmune disorders; including SLE [6,13,14]. Also neuroretinitis may be idiopathic.

Referring to the 1997 update of the 1982 American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus; 4 items are necessary for diagnosis, with at least 1 clinical criterion and at least 1 immunological criterion [16]. However neuroretinitis did not figure in the classic clinical syndrome of SLE even if it is frequently associated to severe form of SLE [15].

Concerning therapy, no clear guidelines are reported in neuroretinitis occurring in SLE. It is noticed that in idiopathic neuroretinitis; even without treatment; the visual prognosis is excellent with above 90% cases achieving a final visual acuity [8,16,17].

Conclusion

We learn from this case, that neuroretinitis may be the revealing form of SLE. Nevertheless, it did not figure in clinical criteria, and is considered as unique entity; different from optic neuritis and lupus retinopathy. Its physiopathology remains understood. This suggest the need of revision of SLE criterion, especially that neuroretinitis is a part of severe form.

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