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Case Report

Bilateral Submacular Serous Detachment in Systemic Lupus Erythematous

Miray Louis de Gonzague[1]*, Rafanomezantsoa Rindra[2], Domoina Hasina Laingonirina[3], Rakotomijoro Etienne[4] and Raobela Léa[2]

[1]*Department of Ophthalmology, University Hospital Tambohobe, Fianarantsoa, Madagascar*

[2]*Department of Ophthalmology, University Hospital Joseph Ravoahangy Andrianavalona, Antananarivo, Madagascar*

[3]*Department of Gastro-Enterology and Internal Medicine, University Hospital Andrainjato, Fianarantsoa, Madagascar*

[4]*Department of Infectious Disease, University Hospital Joseph Raseta Befelatanana, Antananarivo, Madagascar*

***Corresponding Author:** Miray Louis de Gonzague, Ophthalmologist, Ophthalmology service of *University* Hospital Tambohobe, Fianarantsoa Madagascar.

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Abstract

Systemic lupus erythematous (SLE) is a multisystem disorder of autoimmune etiology, predominantly affecting women. Systemic lupus erythematosus can involve many parts of the eye, including the eyelid, ocular adnexa, sclera, cornea, uvea, retina and optic nerve. we report- a case of 25-yera-old female patient presenting a blurred vision during SLE. Ophthalmological examination revealed multiple retinal lesions such as hemorrhage, exudates and cotton wool spots. The Optic coherence Tomography showed a bilateral submacular serous detachment. With systemic corticosteroid and intraocular injection of Avastin, there was an improvement of the symptoms. Systemic Lupus Erythematous is vision threatening. A systematic complete ophthalmological examination is crucial for all patients who present SLE even though the vision is still good to avoid a complicated form of ocular involvement.

Keywords: *Fianarantsoa; Ophthalmological Examination; Submacular Detachment; Systemic Lupus Erythematous; Vision Threatening*

Introduction

Systemic lupus erythematous (SLE) is a multisystem disorder of autoimmune etiology, predominantly affecting women [1]. Systemic lupus erythematosus can involve many parts of the eye, including the eyelid, ocular adnexa, sclera, cornea, uvea, retina and optic nerve [2]. Though ocular manifestations does not fall among the diagnostic criteria of SLE, ocular manifestations are a marker for overall systemic disease activity [3]. Lupus retinopathy is a potentially blinding ocular manifestation of SLE that needs a precocious diagnosis [4]. The purpose is to report a case of 25-year-old woman, who comes from Fianarantsoa, presenting a bilateral submacular detachment due to a systemic lupus erythematous (SLE).

Observation

The disease began spontaneously by polyarthralgia in July 2019, joins swelling in the context of fever 38 - 39°C, myalgia, frontoparietal cephaleus and general health alteration and weight loss. The patient reported using self-medication of doliprane 8 cp/day, indometacin 25 mg 6 capsules a day, tramadol 50 mg 2 tablets/day. Then there was an occurrence of retro auricular, edema, spread over the face and labial mucus began 2 weeks before her presentation to the internal service. Having seen her, a general practitioner sent him for further management at internal medicine of Tambohobe hospital Fianarantsoa in September 11, 2019.

The patient had a past history of hospitalisation at Befelatanana hospital in August 2019 for cephaleus and hyperthermia during which an urinary infection by *Streptococcus* D was found. Thus, she was given Clamoxyl capsules 1g twice a day during 10 days after antibiogramme. After, She was admitted to internal medicine service of university Hospital of Tambohobe Fianarantsoa on September 11, 2019. The chief complains of her admission were tiredness and bilateral blurred vision.

Physical examination found: Blood pressure: 118/80 mmhg, Heart beat: 82/minute, respiratory frequency: 18/minute, temperature = 38°C, SpO_2: 98%.

The patient was in a good health condition with glasgow of 15/15. She presented painless facial, eyelids and labial edema that is not painful to palpation (Figure 1). The buccal cavity presented an ulcerous inflammatory lesions. On her face, she had an itching skin rash consisted of red macules, with interval of healthy skin. The skin rash spread down to chest region, umbilical region and to her back afterward.

Figure 1: *Facial and labial edema with skin rash.*

Systemic findings	Results
NFS	Red blood cell: 3,2T/L Hemoglobine: 100 g/L VGM 88 White blood cell: 3,5 G/L avec PN : 2,1 Platelet: 350 G/L
PCR	24 mg/L
Kidney findings	Serum Creatinine: 85 µmol/L with clearance of creatinine= 84,42ml/min Uremia: 4,25 mmol/L Proteinuria of 24H= 1,86 g
Blood ionogram	K^+: 5,1mmol/L Na^+: 144 mmol/L
Hemostasis findings	TQ: 12 sec TP: 100% INR: 1
Metabolic findings	Glycemia: 5,6 mmol/L Cholesterol Total: 3,0 mmol/L Triglycerides: 3,5 mmol/L Albuminemia: 23g/L
Serologies	AgHBs: Negative Ac Anti VHC: Negative Determine HIV: Negative
Antibodies	Antinuclear Auto-antibody positives with Index at 1280 (autoantibodies anti-DNA native and nuclear solubles anti-genes autoantibodies positives)

Table 1: *Biological findings results.*

Referring to the diagnostic criteria for SLE by American College of Rheumatology (ACR) [5], our patient presented more than 4 criteria such as malar rash, discoid rash, oral ulcers, non-erosive arthritis, serositis, renal disorder, neurological disorder, immunological disorder (anti-DNA antibody) and presence of antinuclear antibodies. So, a systemic erythematous lupus was diagnosed.

The patient was treated with diuretic (Furosemide 40 mg per day), and the bolus of injectable Methyl Prednisolone 1 g/day during 3 days, then shifted with orally taken steroid 1 mg/kg of prednisolone. The edema melt down only 15 days after medication (Figure 2).

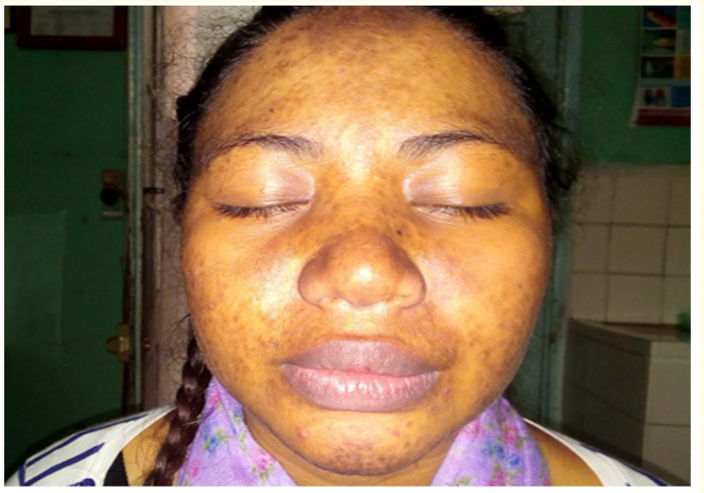

Figure 2: *Facial edema melted down, persistence of skin rash.*

The blurred vision persisted, thus she was sent to our ophthalmology service.

The visual acuity evaluated at hand movement. Anterior segment and intraocular pressure were normal in both eyes at slit lamp examination. Fundus examination showed bilateral tortuous retinal vessels, hemorrhagic and cotton wool spots, hard exudates especially around the macula, macular edema the optic discs were normal. The patient was sent to Antananarivo, the capital of Madagascar, located at about 400 miles from Fianarantsoa to undergo an Optic Coherence Tomography and Fluorescein Angiogram. Unluckily, the angiogram was out of service. The spectral-domain optical coherence tomography revealed massive submacular fluid and macular edema in both eyes (Figure 3a and 3b).

Regarding the evolution, there was an increase in the visual function at count finger three weeks after injection of the Avastin. The patient was considered to do the second injection of anti-VEGF but our patient disappeared.

Discussion

Systemic lupus erythematosus (SLE) is a multisystem disorder of autoimmune etiology, predominantly affecting women [1]. Systemic lupus erythematosus can involve many parts of the eye, including the eyelid, ocular adnexa, sclera, cornea, uvea, retina and optic nerve [2].

Ocular symptoms are correlated to systemic disease activity and can present as an initial manifestation of SLE. In our case, visual loss occurred tardively. The established treatment includes prompt systemic corticosteroids, steroid-sparing immunosuppressive drugs and biological agents. Local ocular therapies are options with promising efficacy. Of course, we treated the patient with systemic corticosteroid and an intraocular injection of anti VEGF (Avastin) that appeared to be helpful. The early recognition of disease and treatment provides reduction of visual morbidity and mortality. Early diagnosis is the key to successful treatment and better prognosis [6].

We found during a funduscopy examination the lesions sus as cotton wool spots, microaneurysms, hard exudates and dot haemorrhages. It is noticed that the classic retinal findings are similar to diabetic and hypertensive retinopathy [7,8].

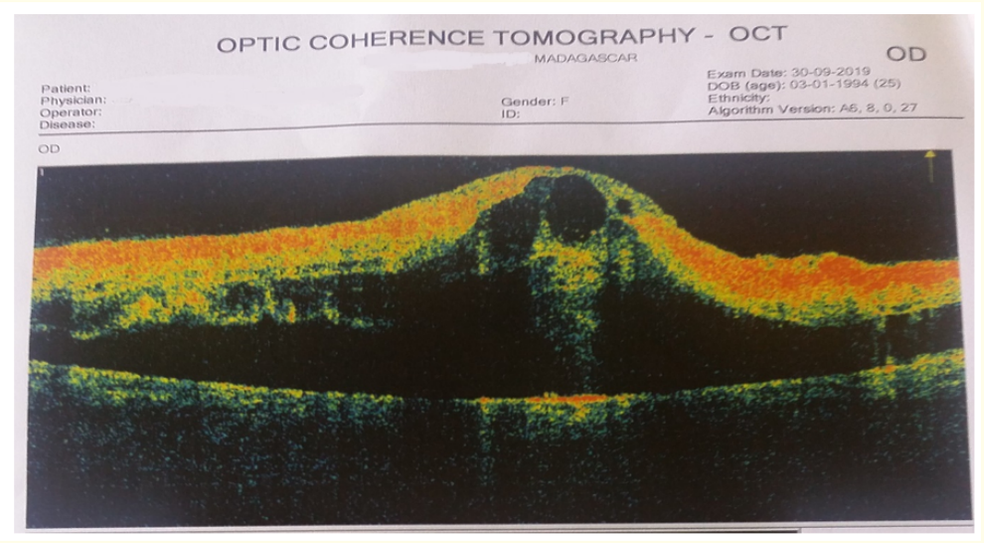

Figure 3a: *Optic Coherence Tomography of the right eye showing submacular fluid and macular edema.*

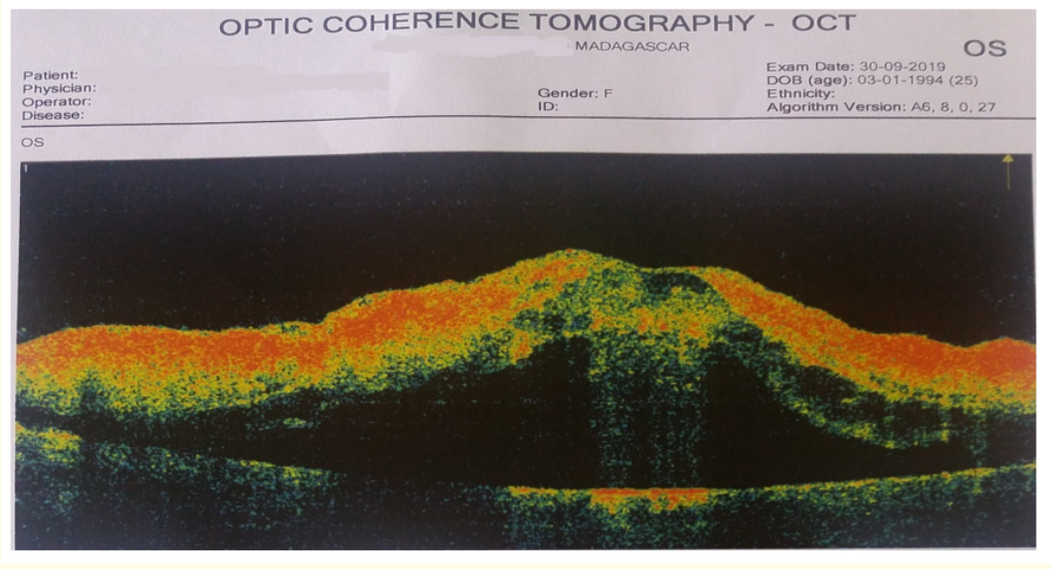

Figure 3b: *Optic Coherence Tomography of the left eye showing submacular fluid and macular edema.*

Even if small intraretinal haemorrhages and cotton-wool spots are usually associated with a good visual prognosis [9]. The vision of our patient was poor. She might have had an undetected artery or venous severe- occlusion for fluorescein Angiogram was not performed and doesn't exist in ophthalmology service. Furthermore, the patient was seen tardively by the ophthalmologist then ocular lesions had an enough time to develop, but it could have been avoided. Lupus retinopathy is a potentially blinding ocular manifestation of SLE but curable [4]. So, Idriss., *et al.* reported a case of 45 year-old female SLE patient presenting a bilateral submacular and detachment choroidopathy during optic coherence Tomography and Fluorescein Angiogram. The visual acuity of their patient was improved with systemic cortico-steroid [10]. We couldn't confirm that our patient has choroidopathy because we didn't perform an Angiogram. Besides, the presence of

retinal lesion such as hemorrhage, exudate, and cotton wool are considered to reflect vascular damage [8]. It is said that The patients with retinopathy had higher levels of serum creatinine than the patients without retinopathy [8]. At the contrary, our patient had retinopathy but her serum creatinine was inferior to normal accounting for 85µml/L. In addition, It is understood that the causes of ocular manifestations of SLE include active lupus, antiphospholipid antibody syndrome (APS), and drugs used to treat SLE [8]. The self medication that our patient reported might have participated to the seriousness of her retinal damage.

Retinopathy and choroidopathy are vision-threatening disease of the posterior segment involving the retina and optic nerve can precede systemic features and may aid in early diagnosis and prompt treatment of patients with SLE [11,12]. Sobrin and Foster noted that Systemic Lupus Erythematosus Choroidopathy may precede a systemic flare by several months

Concerning the treatment, Sobrin and Foster reported a case of 36-year old woman who had a history of systemic lupus erythematosus (SLE). Similar to our case, their patient had been diagnosed with lupus choroidopathy two months prior to her visit and was being treated with CellCept and Prednisone. Her visual acuities were hand motions in the right eye and 20/20 in the left eye. Dilated fundus exam revealed multiple areas of subretinal fluid and retinal pigment epithelial (RPE) detachments in the macular and peripapillary areas of both eyes. She started on intravenous Cytoxan. Her visual acuity improved in the right eye to 20/50 but worsened in the left eye to 20/125. Their patient, after the failure of methylprednisolone and CellCept, underwent treatment with laser photocoagulation directly to the active RPE leaks and around leaks associated with a mechanical defect in the RPE in both eyes. After four total months of Cytoxan treatment and an additional laser treatment to the right eye, the vision of the patient improved to 20/25 in both eyes [13]. Moreover, Neuman and Foster pinpointed that Immunosuppression is the mainstay of therapy. Laser photocoagulation for proliferative retinopathy is also thought to be beneficial [3,14].

Honestly speaking, it was the first case we met. We treated our patient with pulse of prednisolone to heal systemic lupus and intraocular Avastin the only anti-VEGF available in Madagascar. Anti-VEGF was used in order to prevent neovasularisation from forming and to tread submacular fluid. After the first injection, there was a slight improvement of the visual acuity in both eyes and our patient didn't come back for follow-up. Despite immunotherapy is the backbone of treatment of Systemic Erythematous Lupus associated with retinopathy. Some authors suggested an intravitreal Bevacizumab that appears to be a powerful tool for vaso-occlusion, vasculitis and neovascularisation in patients with lupus [2,15].

Through these abovementioned affirmation we can conclude that SLE is vision threatening. Thus, a systematic complete ophthalmological examination is crucial for all patients who present SLE even though the vision is still good to avoid a complicated form of ocular involvement. More training about SLE should be given to the general practitioner, internist doctors and ophthalmologists so that they can manage properly the disease because the early recognition of disease and treatment provides reduction of visual morbidity and mortality. It is stated that Early diagnosis is the key to successful treatment and better prognosis [6].

An university hospital like Tambohobe Fianarantsoa should be equipped with ophthalmological instruments including an Optic Coherence Tomography, Angiogram, Laser so that the patients are taken in charge properly and to prevent the patient from going and for to Antananarivo to do ophthalmological findings. In addition, Malagasy government should supply the anti-VEGF drugs to all ophthalmology services in government hospitals in Madagascar. Greater action is needed to sensitize people to avoid self medication which is one of the public health concerns in Madagascar.

Conclusion

Systemic lupus erythematous (SLE) is a multisystem disorder of autoimmune etiology, predominantly affecting women. Lupus retinopathy is a potentially blinding ocular manifestation of SLE that needs a precocious diagnosis. A systematic complete ophthalmological examination associated with Optic computed Tomography and Fluorescein Angiography is crucial for all patients who present SLE even though the vision is still good to avoid a complicated form of ocular involvement especially choroidopathy which may precede systemic Lupus Erythematous manifestation. Immunotherapy accompanied with Laser are the mainstay of treatment of Systemic Lupus Erythematous Choroidopathy. More training about SLE should be given to the general practitioner, internist doctors and ophthalmologists so that they can manage properly the disease because the early recognition of disease and treatment provides reduction of visual morbidity and mortality.

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