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OPHTHALMOLOGY
Case Report

Refractory Lupus Choroidopathy

Ana M. Suelves[1], David Fernandez[2] and Julia P Shulman[3]

[1]Department of Ophthalmology, New York Medical College, USA
[2]Department of Rheumatology, Hospital for Special Surgery, USA
[3]Department of Ophthalmology, New York Medical College, USA

***Corresponding Author:** Julia P. Shulman, New York Medical College, New York, USA.

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Abstract

We report a case of bilateral lupus Choroidopathy and central nervous system (CNS) vasculitis in a 43-year-old female with systemic lupus erythematous. The patient improved systemically on high dose steroids and mycophenolate. Choroiditis flared and escalation of therapy with Rituximab achieved CNS disease control. Choroiditis refractory to systemic therapy improved with micropulse laser therapy. Sequential indocyanine green angiography and enhanced depth imaging of optic coherence tomography allowed monitoring the response to therapy.

Keywords: *Lupus Choroidopathy; Choroiditis; uveitis; systemic lupus erythematous; Indocyanine green angiography; Spectral domain optical coherence tomography*

Abbreviations: (SLE): Systemic lupus erythematous; (CNS): Central nervous system; (ICG): Indocyanine green angiography; (OCT): Optical coherence tomography; (EDI): Enhanced depth imaging; (TM): Treated with micropulse; (od): Right eye; (os): Left eye; (RD): Retinal detachment;

Introduction

Systemic lupus erythematous (SLE) is an autoimmune disease characterized by production of Autoantibodies directed against ubiquitous self-antigens. Autoantibodies participate in the formation of immune complexes, which deposit in the walls of small vessels in multiple organs, including the choroid 1the renal glomerulus or joints; causing local inflammation and tissue damage [1].

Choroidopathy is an extremely rare manifestation of SLE, that correlates strongly with renal and central nervous system (CNS) involvement [2,3]. It presents with bilateral multiple serous retinal detachments. Even though, systemic immunosupression is the mainstay of therapy, laser therapy has showed to be useful in cases of recurrent serous detachments despite adequate systemic immunosupression [4].

We present a case of SLE-associated Choroidopathy, assessed by fluorescein, indocyanine green angiography (ICG) and spectral domain optical coherence tomography (OCT) with enhanced depth imaging (EDI) treated with micropulse laser photocoagulation.

Case report

A 43 year-old female presented with decreased vision and headaches for 2 weeks. She had a 15-year history of SLE with nephritis, arthritis, rash, positive antiphospolipid and antinuclear antibodies. She had completed 6 months of cyclophosphamide 6 years prior and was taking 10 mg of oral prednisone.

Her visual acuity was 20/50 in the right eye (od) and 20/100 in her left eye (os). Fundus exam showed bilateral retinal detachments (RD) in the posterior pole. ICG demonstrated early hyperfluorescence with focal clusters of hyperfluorescence in the late phases (Figure: 1.1). Fluorescein angiography revealed multiple pinpoint hyperfluorescence (Figure: 1.2). OCT showed bilateral serous RD with pigment epithelial detachments (Figure: 2). Brain magnetic resonance imaging revealed cerebral vasculitis.

She received high dose steroids intravenously and 2 grams of mycophenolate daily with improvement of vision and serous RD. Her choroiditis flared while tapering systemic steroids, so Rituximab therapy was initiated. Her systemic disease was quiescent, but had ongoing choroiditis despite appropriate systemic immunosupression. Micropulse laser over the areas of leakage on ICG achieved the resolution of serous detachments bilaterally (Figure: 3). Visual acuity improved to 20/25 od and 20/50 os at last follow up 6 months after first presentation. Table 1 showed central retinal thickness and choroidal thickness.

Discussion

Lupus patients typically present with retinal vasculitis, intraretinal hemorrhages and cotton-wool spots. Choroidopathy is an extremely rare manifestation of SLE, with less than 40 cases described in the literature [5,6]. It presents with multifocal neurosensory detachments and serous pigment epithelial detachments, that may mimic hypertensive retinopathy and multifocal central serous chorioretinopathy; also seen in lupus patients. It is imperative to diagnose this entity properly and differentiate it from other diseases, given the different therapeutic approach. In our patient, the positive response to steroids and systemic immunosupression support lupus Choroidopathy as the diagnosis.

The cause of lupus Choroidopathy remains poorly understood. Matsuo., *et al* hypothesized that the deposition of immune complexes in the choriocapillaris may trigger fluid leakage into the Subretinal space [7]. Clinically, the areas of serous detachments correlated in our patient with areas of choroidal hyperpermeability on ICG, supporting this disease mechanism.

There are 2 patterns of ICG described in lupus Choroidopathy patients. First, IGC shows focal areas of early hypofluorescence of stromal choroidal vessels with fuzziness of large choroidal vessels in late phases. Second well-defined clustered pinpoint spots of choroidal hyperfluorescence seen in intermediate or late phases. It is unknown if the different ICG patterns represent different pathophysiologic mechanisms, however ICG is important in suspected lupus choroiditis, as it can be present even with a normal fluorescein angiogram [8].

Evaluation of the choroid with EDI-OCT was found to be an excellent tool to assess the response to treatment and may allow detect subclinical recurrences in posterior uveitis [9]. In our patient, increased choroidal thickness corresponded to active disease and Subretinal fluid. This may be a useful modality in lupus patients with Choroidopathy to monitor disease activity or in case of shellfish allergy where ICG is contraindicated.

The cornerstone of lupus Choroidopathy treatment is systemic immunosupression that achieves disease control in 82% of patients. Although, choroiditis might be challenging to treat, requiring both systemic therapy and ocular treatment with focal laser over the areas of active choroiditis [10]. Cho., *et al*. [4] reported for the first time the efficacy of focal laser and photodynamic therapy as adjuvant therapy for treating refractory choroiditis. The micropulse photocoagulation technique divides the laser emission into a "train" of repetitive pulses, which allows heat dissipation and reduces retinal damage. It has been showed to be as effective as conventional laser for treating diabetic macular edema [11] and chronic central serous chorioretinopathy [12]. To the best of our knowledge, the use of micropulse laser has not been reported in treatment of lupus Choroidopathy.

We present a case of lupus Choroidopathy with refractory bilateral serous detachments despite appropriate systemic immunosupression that responded to micro pulse laser therapy. Multimodal imaging allowed for monitoring of disease activity. Our case highlights the strong association of choroiditis, nephropathy and cerebral SLE-related vasculitis.

	Right eye		Left eye	
	Central retinal thickness (µm)	Choroidal thickness (µm)	Central retinal thickness (µm)	Choroidal thickness (µm)
Presentation	745	607	294	525
1 month	716	502	300	547
6 months	342	495	260	478

Table 1: *Central retinal thickness and subfoveal choroidal thickness.*

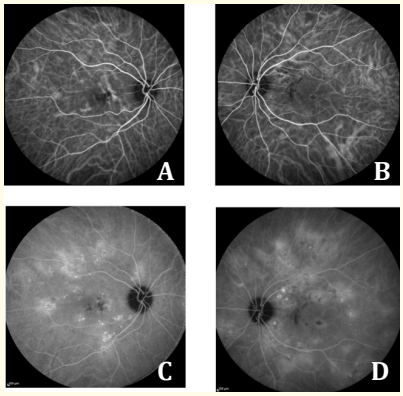

Figure 1.1: *ICG images shows early hyperfluorence (A, B) with multiple foci of choroidal hyperfluoresce undetected on FA or clinical exam (late-recirculation phase) (C,D).*

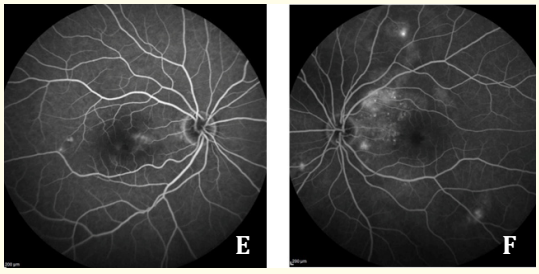

Figure 1.2: *Fluorescein angiography in midstage showed multiple areas of delayed choroidal filling with pinpoint hyperfluorescence with pooling corresponding to the areas of neurosensory detachment (E,F).*

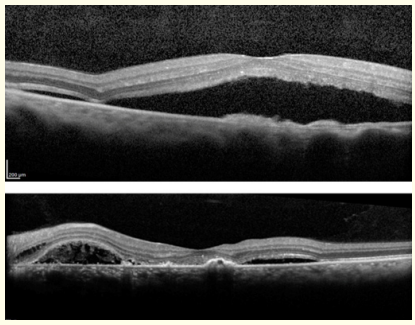

Figure 2: *Optical coherence tomography at baseline showed multiple bilateral neurosensory retinal detachment and pigment epithelial detachment with areas of hyperreflectivity in subretinal space suggestive of chronic exudation.*

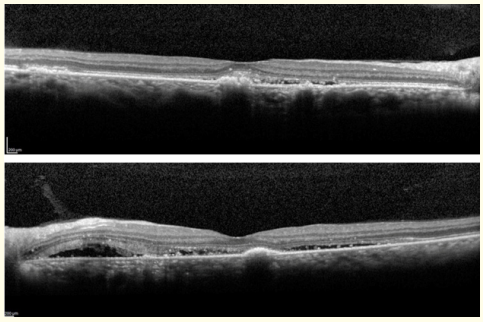

Figure 3: *OCT showed resolution of serous retinal detachment at 6 months of follow up.*

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