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Case Report Open Access

Bilateral Choroidopathy, Nephritis and Hypertension in Systemic Lupus Erythematosus

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Abstract

A 18-year-old woman with systemic lupus erythematosus (SLE) presented with right sided migraine and blurred vision of the right eye. Ophthalmologic evaluation revealed multiple bilateral exudative retinal detachments, with increased choroidal thickness measured with optical coherence tomography (OCT). Acute renal dysfunction contraindicated fluorescein or indocyanine green angiography. The presence of choroidopathy was the first presentation of lupus nephritis. She was treated with corticosteroids and immunosuppressive agents with resolution of serous retinal detachments and complete remission of proteinuria and renal function. OCT may be a key exam for the early diagnosis of choroidopathy and implementation of appropriate therapeutic measures, necessary to prevent permanent damage.

Keywords: Systemic lupus erythematosus; Nephritis; Ocular; Chorioretinopathy

Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease of unknown etiology characterized by the production of autoantibodies and polymorphic manifestations of end-organ damage [1]. The disease can manifest in many forms and severity, ranging from mild cutaneous and joint involvement, to devastating ocular complications, to lethal renal, cardiac, and cerebral involvement [2]. Renal involvement, characterized by immune complex deposition, inflammation, and scarring of the glomeruli and interstitium, is the most common severe clinical manifestation of SLE [3]. Ocular involvement in SLE can involve almost all structures of the eye, most frequently resulting in keratoconjunctivitis sicca and retinopathy [2]. Among ophthalmic manifestations, posterior segment involvement, namely optic nerve, retina and choroidal involvement are particularly noteworthy, given their importance in visual prognosis and their correlation with central nervous system (CNS) and systemic disease activity [1,2]. Choroidopathy is rarely found and may be clinically silent [1]. It occurs essentially in women and is bilateral in 68% of cases [4]. The authors describe a case of acute choroidopathy as the first manifestation of lupus nephritis.

Case Report

A 18-year-old caucasian-woman who had been treated with hydroxychloroquine (HCQ) 200 mg/day for systemic lupus erythematosus (SLE) for 6 months, presented to the emergency department with blurred vision of the right eye (RE) and right-sided migraine. Systolic and diastolic blood pressures were 183 and 135 mmHg, respectively. Ophthalmic examination revealed a best corrected visual acuity (BCVA) of 20/50 in the RE and 20/20 in the left eye (LE). Intraocular pressure was 13 mmHg in the RE and 14 mmHg in the LE and anterior segment examination revealed bilateral subconjunctival hemorrhage. Fundoscopy revealed multiple bilateral exudative retinal detachments with foveal involvement only in the RE. Spectral domain optical coherence tomography (SD-OCT) with enhanced depth imaging software (EDI) was performed. OCT revealed subretinal fluid with increased subfoveal choroidal thickness, 323 µm in the RE and 366 µm in the LE (Figure 1). These findings were consistent with acute lupus choroidopathy.

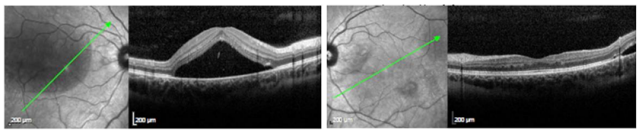

Figure 1: Spectal domain optical coherence tomography images before the treatment. In the right eye, there is a prominent subfoveal fluid accumulation; in the left eye, there is a small extra-foveal subretinal fluid pouch.

Laboratory tests revealed hypochromic microcytic anemia (Hb 11 g/L, MCV 73.6 fL, MCHC 25.3 g/L), leucopenia 2360 L, thrombocytopenia 135000/µL, hypocomplementemia (C'3 0.40 g/L and C'4 0.03 g/L (normal values 0.90-1.80 and 0.10-0.40 g/L, respectively), ANA positivity, anti-dsDNA positivity (ELISA: 400 IU/ml), and triple positivity for antiphospholipid antibodies; acute kidney

injury (blood urea nitrogen and creatinine of 112 and 1.85 mg/dl, respectively) with proteinuria 3.7 g/24h. The renal biopsy revealed Class IV G (A) lupus nephritis (Figure 2).

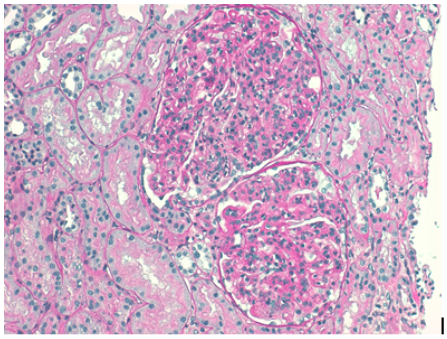

Figure 2: Renal Biopsy: PAS X200. Class IV lupus nephritis. This picture shows two glomeruli with intense endocapillary hypercellularity and subendothelial deposits.

Given the risk of renal function deterioration retinal angiography was not performed. She was treated with pulses of methylprednisolone 1 g/day for 3 days and rituximab 1 g, followed by mycophenolate mofetil 1 g/day and prednisolone 1 mg/kg/day. After two weeks, the second dose of rituximab was administered and mycophenolate mofetil was increased to 3 g/day. During the first week of hospitalization, labetolol infusion and multiple other antihypertensive drugs were required to control the blood pressure. Progressive recovery of visual acuity was documented over the following weeks and, 2 months after admission, BCVA was 20/20 in both eyes. Ophthalmic examination and SD-OCT revealed complete resolution of serous retinal detachments (Figure 3). Subfoveal choroidal thickness reduced to 298 µm in the RE and 287 µm in the LE. Over the next 8 months, it was possible to discontinue steroids and taper mycophenolate mofetil to 2 g per day, respectively, while maintaining HCQ 400 mg/day, with complete remission of proteinuria, normalization of blood pressure, renal function, and progressive decrease of disease activity (Figure 4).

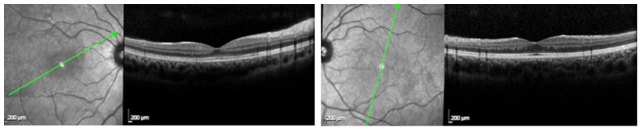

Figure 3: Spectal domain optical coherence tomography images after the treatment, showing complete resolution of retinal detachments.

Discussion

Ocular manifestations can sometimes be the first presentation of SLE, especially when there is organ involvement such as nephropathy (lupus nephritis), CNS vasculitis and uncontrolled hypertension [4-6]. Ophthalmic manifestations can be detected in one-third of SLE patients and may be present at the onset of the disease or manifest during its course and are usually indicative of disease activity [7]. Lupus choroidopathy is a rare ocular manifestation, with less than 40 cases described in the scientific literature until 2012 [6]. The presence of choroidopathy is an indicator of disease activity and may announce the appearance of SLE nephropathy [8]. Due to their rarity, ocular manifestations have not been included in the diagnostic criteria scoring system for establishing clinical diagnosis of SLE [9]. However, the finding of choroidal alterations, even in asymptomatic patients could represent a promising early indicator which is sensitive to ocular involvement and thus "indirectly" to renal involvement [1]. The introduction of these instruments for ocular and, eventually, renal involvement should be considered useful for prognostic purposes in the approach of these patients [1]. It has been suggested that all patients diagnosed with SLE should undergo a complete ophthalmologic evaluation, including OCT and eventually fluorescein and indocyanine green angiography to exclude ocular involvement [7]. In patients with lupus choroidopathy, fundus fluorescein angiography presents delayed choroidal filling or areas of choroidal nonperfusion in the early stages, followed by focal cluster pinpoints of hyperfluorescent areas with pooling, corresponding to the areas of exudative retinal detachment [2].

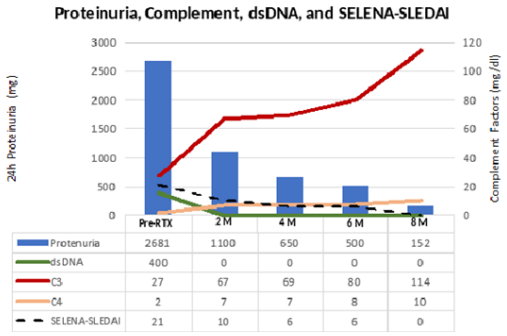

Figure 4: Disease course: Shown is a summary of the patient's disease course before rituximab and after rituximab with regression of proteinuria (blue bar), normalization of complement factors (red and orange solid lines) and anti-dsDNA autoantibody levels (green line) and significant reduction of disease activity until remission as seen in the SELENA-SLEDAI (black dash line).

Indocyanine green angiography is more sensitive to choroidal involvement and typically presents with focal, transient early-phase hypofluorescence secondary to perfusion delay followed by late-phase diffuse hyperfluorescence due to vascular hyperpermeability. More subtle findings include distortion of the large choroidal vessels and also pinpoint clusters of choroidal hyperfluorescence in the intermediate phase [9]. Angiography, however, may be of limited use in these patients given the frequent impairment in renal function. On the other hand, OCT is a non-invasive, non-contrast, fast and objective technology that has been proven useful in the diagnosis and follow-up of these patients [10]. Typically, it reveals multiple serous retinal elevations, with or without intraretinal cystoid macular edema and retinal pigment epithelium irregularities [11]. Additionally, newer enhanced depth imaging OCT software (EDI) may be used to access choroidal morphology and thickness which may also be useful in the diagnosis and follow up of these patients. It was already demonstrated that patients with lupus nephritis present subclinical changes in indocyanine green angiography that are not present in SLE patients without renal involvement [1]. These changes may correlate with an increase in choroidal thickness which is highest during acute choroidopathy. An increase in choroidal thickness with disease activity

measured by EDI-OCT has been described in other pathologies like Vogt-Koyanagi-Harada [12] or Behçet's disease [13]. However, choroidal thickness changes have not been extensively studied in SLE nephritis. In this patient, we observed a bilateral increase in subfoveal choroidal thickness, which resolved with the improvement of renal function. Our patient required prolonged hospitalization for strict blood pressure, renal function and vision control. Conventional therapy combines glucocorticoids and other immunosuppressive agents such as cyclophosphamide, mycophenolate mofetil, and azathioprine. In the last decades, the use of targeted biologic therapy such as the off-label use of rituximab as first-line treatment has demonstrated similar efficacy to long-term conventional treatment, but with significantly lower PDN use [14]. Overall, our strategy proved to be highly effective. Strict monitoring of antiphospholipid antibodies and lupus anticoagulant is ongoing as she may require prophylactic anticoagulation. Our report highlights rare but significant posterior segment disease in lupus nephritis. We demonstrate how a multidisciplinary team was required for early recognition and effective treatment of ocular involvement in SLE.

References

1. Baglio V, Gharbiya M, Balacco-Gabrieli C, Mascaro T, Gangemi C, et al. (2011) Choroidopathy in patients with systemic lupus erythematosus with or without nephropathy. J Nephrol 24: 522-529.
2. Nguyen QD, Uy HS, Akpek EK, Harper SL, Zacks DN, et al. (2000) Choroidopathy of systemic lupus erythematosus. Lupus 9: 288-298.
3. Moraes-Fontes MF, Lúcio I, Santos C, Campos MM, Riso N, et al. (2012) Neuropsychiatric features of a cohort of patients with systemic lupus erythematosus. ISRN Rheumatol 2012: 989218.
4. Edouard S, Douat J, Sailler L, Arlet P, Astudillo L (2011) Bilateral choroidopathy in systemic lupus erythematosus. Lupus 11: 1209-1210.
5. Canamary A, Sousa J, Andrade G, Nascimento H (2017) Choroidopathy in systemic lupus erythematosus. Rev Bras Oftalmol 76: 2
6. Palejwala NV, Walia HS, Yeh S (2012) Ocular manifestations of systemic lupus erythematosus: a review of the literature. Autoimmune Dis 2012: 290898.
7. Dammacco R (2017) Systemic lupus erythematosus and ocular involvement: an overview. Clin Exp Med 18: 135-149.
8. Inês L, Silva C, Galindo M, López-Longo FJ, Terroso G, et al. (2015) Rheumatic Diseases Registry of the Portuguese Society of Rheumatology; Registry of Systemic Lupus Erythematosus Patients of the Spanish Society of Rheumatology. Classification of Systemic Lupus Erythematosus: Systemic Lupus International Collaborating Clinics Versus American College of Rheumatology Criteria. A Comparative Study of 2,055 Patients from a Real-Life, International Systemic Lupus Erythematosus Cohort. Arthritis Care Res 67: 1180-1185.
9. Gharbiya M, Pecci G, Baglio V, Gargiulo A, Allievi F, et al. (2006) Indocyanine green angiographic findings for patients with systemic lupus erythematosus nephropathy. Retina. 26: 159-164.
10. Kouprianoff S, Chiquet C, Bouillet L, Romanet JP (2010) OCT follow-up of systemic lupus erythematosus choroidopathy. Ocul Immun Inflamm 18: 113-115.
11. Hasanreisoglu M, Gulpinar Ikiz GD, Kucuk H, Varan O, Ozdek S (2017) Acute lupus choroidopathy: multimodal imaging and differential diagnosis from central serous chorioretiopathy. Int Ophthalmol 38: 369-374
12. Jap A, Chee SP (2017) The role of enhanced depth imaging optical coherence tomography in chronic Vogt-Koyanagi-Harada disease. Br J Ophthalmol 101: 186-189.
13. Kim M, Kim H, Kwon HJ, Kim SS, Koh HJ, et al. (2013) Choroidal thickness in Behcet's uveitis: an enhanced depth imaging-optical coherence tomography and its association with angiographic changes. Invest Ophthalmol Vis Sci 54: 6033-6039.
14. Gracia-Tello B, Ezeonyeji A, Isenberg D (2017) The use of rituximab in newly diagnosed patients with systemic lupus erythematosus: long-term steroid saving capacity and clinical effectiveness. Lupus Sci Med 4: e000182.