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]. Chorioidopathy 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: Special 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 (C3 0.40 g/L and C4 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
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 pinpoint areas of hyperfluorescent areas with pooling, corresponding to the areas of exudative retinal detachment [2].

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. 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

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 pinpoint areas of hyperfluorescent areas with pooling, corresponding to the areas of exudative retinal detachment [2].
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