Case Report

Multiple Bilateral Retinal Pigment Epithelial Detachments in a Patient with Systemic Lupus Erythematosus: A Case Report

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Abstract

Purpose: To report a case of multiple bilateral retinal pigment epithelial detachments (PEDs) in a woman with systemic lupus erythematosus (SLE).

Methods: Case Report.

Results: A 28-year-old female with mild blurred bilateral vision in both eyes (OU) without pain or any other symptom was admitted to the hospital due to worsening renal function and uncontrolled high blood pressure (HBP). Best-corrected visual acuity (BCVA) was 20/30 and 20/40, right and left eyes, respectively. She had SLE, glucose-6-phosphate dehydrogenase deficiency, and immune thrombocytopenic purpura. BP was over 150/90 mmHg for more than 1.5 years, and she used corticosteroids at varying doses for more than 4 years. During hospitalization, she was taking prednisone 60 mg daily as Class IV lupus nephritis was diagnosed. On fundoscopy, she had a lacy retinal pattern, remarkably on the macula in OU. Spectral-domain optical coherence tomography revealed multiple bilateral serous PEDs and pachychoroid. Angiofluoresceinography displayed multiple pooling hyperfluorescence areas. Six months afterward, while she was on prednisolone 10 mg daily, and antihypertensive medications, BCVA was improved to 20/25 OU. Nevertheless, she had no retinal or choroidal changes. Her findings could be related to SLE choroidopathy, central serous chorioretinopathy-like disease, and/or hypertensive choroidopathy.

Conclusions: Ocular involvement affects nearly one-third of SLE patients. The findings are variable and can include nearly any part of the eyeball. Multiple bilateral PEDs have been described in the literature; however, in this case, it is probably multifactorial and not only related to SLE.

Keywords: Central serous chorioretinopathy, Hypertensive choroidopathy, Retinal pigment epithelial detachments, Systemic lupus erythematosus

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Introduction

Systemic lupus erythematosus (SLE) is an intricate illness affecting multiple organs and connective tissue. Its prevalence ranges from 20 to 150 cases per 100,000 person, and its incidence rates range from 1 to 10/100,000 persons year globally. This disease predominantly affects women of reproductive age. Ocular involvement affects nearly one-third of SLE patients. It may anticipate other systemic symptoms and can be associated with disease activity.[1-3]

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The ocular findings are variable and can present as choroidopathy, optic neuropathy, retinal vasculitis, vaso-occlusive disorder, iridocyclitis, keratoconjunctivitis, and eyelid abnormalities. The most prevalent being keratoconjunctivitis sicca, present in approximately 33% and associated with secondary Sjogren's disorder, whereas choroidal and retinal conditions are more associated with vision loss.[1] Moreover, posterior segment involvement choroidopathy or retinopathy is related to poor

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systemic disease control as it indicates high disease activity with a correlation to renal impairment and central nervous system lupus.[3] Gladly, with the improvements in SLE management over the past decades, its incidence is significantly decreasing.[3,4] Dias-Santos *et al*. reported an incidence of 0.6% of SLE choroidopathy in their cohort.[3]

Nguyen *et al*. analyzed all SLE choroidopathy published cases from 1968 to 1998 and they observed best-corrected visual acuity (BCVA) of 20/40 or better in 64% of cases, with reported bilateral or unilateral blurred vision.[5] Fundus fluorescein angiography (FFA) demonstrates sensory retina multifocal serous elevations, related or unrelated to retinal pigment epithelial detachments (PEDs), or retinal pigment epitheliopathy.[4]

We report a clinical case of multiple bilateral PEDs in a woman with SLE.

Case Report

A female subject, 28 years old, reported for an evaluation, referred by the rheumatology team. Seven days before the ophthalmic assessment, she was admitted for worsening renal function, uncontrolled high blood pressure (HBP), and multifactorial anemia. She had a previous diagnosis of SLE, glucose-6-phosphate dehydrogenase deficiency, and immune thrombocytopenic purpura with inconsistent follow-up. BP was over 150/90 mmHg for more than 1.5 years, with peaks striking 220 mmHg of systolic blood pressure and she was on corticosteroids with varying doses for more than 4 years. During hospitalization, she was taking prednisone 60 mg daily as Class IV lupus nephritis was diagnosed. The study was approved by the Research Ethics Committee of the State University of Campinas (approval number CAAE: 53895321.9.0000.5404). All procedures were performed in accordance with the principles of the World Medical Association Declaration of Helsinki.

Her SLE diagnosis was in March 2017, at which time she had a positive antinuclear antibody titer of 1/1280 thick speckled, positive ribonucleoprotein antibody, positive SS-A/Ro antibodies, malar rash, and thrombocytopenia.

She complained of mild blurred bilateral vision in both eyes (OU) for a few months, without pain, headache, or any other symptoms, and no previous assessments. Furthermore, she had no history of ophthalmologic illness and denied previous surgeries, trauma, use of illegal substances, or ophthalmic lens use. She did not report any relevant family or social history.

Her BCVA was 20/30 and 20/40, right (OD) and left (OS) eyes, respectively. Anterior segment examination, pupillary reflexes, and extraocular movements were normal. Intraocular pressure with the Goldmann tonometer was 12 bilaterally.

The fundoscopy examination showed a lacy retinal pattern in OU, remarkably in the macula, with no cotton-wool spots or hemorrhages. In addition, it highlighted a degree of arterial narrowing, constricted and tortuous arterioles, enhanced arterioles light reflex (copper wiring), and a few arteriovenous nicking crossing, as registered with colored retinography [Figure 1a and b]. Spectral-domain optical coherence tomography (SD-OCT) revealed subfoveal choroidal thickness of 275 µm OD and 314 µm OS with multiple bilateral serous PEDs and choroidal pachyvessels [Figure 1c and d].

Fundus autofluorescence demonstrated, bilaterally, multiple well-defined posterior pole lesions with moderate autofluorescence, corresponding to PEDs areas seen on SD-OCT [Figure 1e and f]. FFA displayed fluorescein gradually filling the multiple fluid-filled spaces, distinguishing multiple pooling hyperfluorescence areas in OU [Figure 2a and b].

She remained under rheumatology care and returned 2 months later using sodic mycophenolate, prednisone 30 mg daily tapering regimen, and three antihypertensive drugs under regular use, nevertheless presenting with poorly controlled hypertension. BCVA was 20/25 OD and 20/30 OS. Despite BCVA improvement, examination showed no imaging changes.

Her last evaluation was 6 months afterward, she was still on a sodic mycophenolate induction dose and the prednisone regression dose was lower, 10 mg daily. She remained on three antihypertensive drugs, reporting better blood pressure control despite measurements never reaching below 130/90 mmHg. Subject refraction displayed OD −0.50 −1.25 160° and OS −1.25 30° with BCVA of 20/25. Although she demonstrated visual acuity improvements, her clinical and complementary examination revealed no retinal or choroidal changes [Figure 3] compared with her previous examinations.

Given these signs, the main hypothesis elaborated was a choroidopathy that in this patient, interestingly, could be related and explained by three possible etiologies: SLE choroidopathy, central serous chorioretinopathy (CSC)-like disease, and/or hypertensive choroidopathy.

The three etiologies above could be connected as she had SLE with active nephritis and had been using corticosteroids for many years, which could have predisposed her to a CSC-like condition. Moreover, she had uncontrolled HBP with inconsistent medication use that could contribute to hypertensive choroidopathy. Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

Discussion

SLE is a connective tissue illness affecting multiple organs. Its pathogenesis is multifactorial, leading to dysregulation of both compartments, cellular and humoral of the immune system, creating an autoimmune response against self-chromatin antigens. The clinical manifestations are highly dependent on the autoantibodies and their pathogenic effects by complement-mediated inflammation, immune complexes-mediated damage, and cell apoptosis. One of the

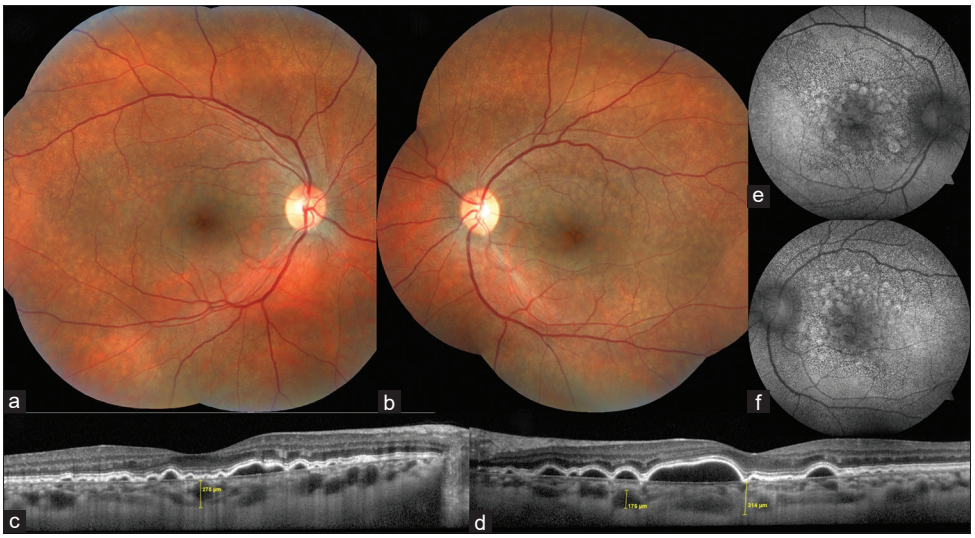

Figure 1: Colored retinography showing lacy retinal pattern on the macula, arterial narrowing, constricted and tortuous arterioles, enhanced arterioles light reflex (copper wiring), and a few arteriovenous nicking crossing in both eyes (OU) (right eye [OD] [a], left eye [OS] [b]). Pigment epithelial detachments and pachychoroid with 275 μm OD and 314 μm OS subfoveal thickness on spectral-domain optical coherence tomography (OD [c], OS [d]). Fundus autofluorescence showing multiple well-defined posterior pole lesions with moderate autofluorescence OU (OD [e], OS [f])

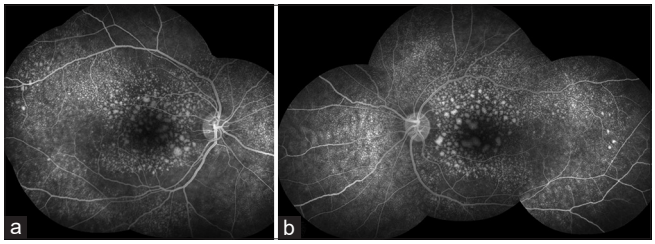

Figure 2: Fundus fluorescein angiography displaying multiple pooling hyperfluorescence areas in both eyes in the right eye (a) and left eye (b)

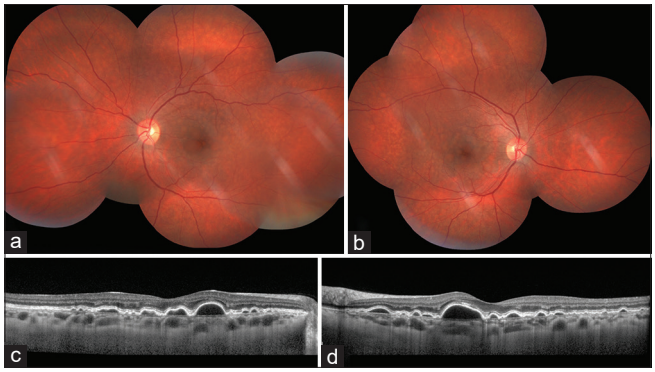

Figure 3: Colored retinography 8 months after presentation in the right eye (OD) (a) and the left eye (OS) (b) spectral-domain optical coherence tomography 8 months after presentation in the OD (c) and the OS (d)

most severe manifestations is kidney involvement, which affects 50%–80% of SLE patients and frequently appears in the first 3 years of the disease. Ocular involvement affects approximately one-third of SLE patients with variable severity and nearly all parts of the eyeball can be affected. It may be associated with systemic disease activity or be a complication of systemic or topical therapy.[1-3,6]

In regard to the possible etiologies for the patient's choroidopathy, it is important to highlight that lupus choroidopathy (LC) can develop with lupus retinopathy or isolated. The latter retinopathy is accredited to vasculopathy rather than vasculitis, meaning ordinarily immune complex-mediated microangiopathy, with complement activation per immune complex deposition and vascular damage. Hypertensive and diabetic retinopathy show comparable microangiopathy.[6] Therefore, her fundus vascular alterations could be related to either SLE or hypertension, making it difficult to distinguish both.

The precise mechanism of LC remains unclear; however, a few decades ago, histopathologic studies showed the existence of mononuclear inflammatory cell infiltrates, as well as deposits of immunoglobulin and complement in the choroidal vessels.[7-9] Most likely, LC results from a combination of inflammation, thrombosis, and hypertension with the occurrence of occlusion in the choroidal vessels from microthrombus formation.[5,10] It is a very rare condition, with only 28 cases (47 involved eyes) reported in the English literature from 1968 to 1998.[5]

Recent studies postulate that LC could express drusen-like deposits (DLD) in advanced disease.[11,12] The DLD is situated between the retinal pigment epithelium (RPE) and the Bruch membrane and may be associated with complement pathway dysregulations in the eye.[10-12] They appear similar to drusen deposits and could present with hyporeflective content, however, differently from our patient, they do not have serous content.

One limitation of our case is the absence of indocyanine green angiography. Choroidal modifications not visible on the other examinations could be the reason for the vision improvement noticed in the subsequent visit.

Concerning hypertensive choroidopathy, it may contribute to PEDs and sensory retinal detachments by ischemia and the breakdown of the outer blood-retinal barrier at the level of the RPE, due to constriction of choroidal blood flow.[7] In addition, hypertension supplementary hydrostatic force results in fluid leakage into the detachment.[7] Our patient did not have sensory retinal detachments, which makes this a less probable diagnose. However, Karatepe Hashas *et al*. showed another case with isolated PEDs and also considered hypertensive choroidopathy as a differential diagnosis.[13]

The other possible association, in this case, CSC, is the part of the pachychoroid spectrum. Its pathogenesis is multifactorial and not fully understood yet. It is characterized by RPE disturbances, inflammatory, and circulatory changes leading to the existence of a thick choroid associated with increased vascular permeability and neurosensory retina serous detachment, as fluid crosses the RPE. Men are commonly affected, in a male-to-female ratio of 3:1, the mean age group is 35–55 years, and this condition is generally unilateral. Exogenous corticosteroid use has been proved to have a strong association with the disease onset.[14] Although it is less common in women, her choroidal pachyvessels, PEDs, and corticosteroids use collaborate with this diagnose.

As stated above, all the conditions – SLE activity, glucocorticoid use, and uncontrolled hypertension – were fully addressed considering the patient's illness limitations. For instance, her SLE activity makes it impossible to stop the glucocorticosteroids. Moreover, the antihypertensive medications progressively introduced did not fully control her blood pressure.

Hence, it is hard to separate only one etiologic factor for the patient's choroidopathy. Therefore, she requires multidisciplinary care with regular visits to improve the conditions that can be contributing to her ocular and systemic alterations.

When facing a choroidopathy in a patient with many comorbidities, like our patient, it is primordial to state all the possible associated factors as it may be contributing to more than one disease. In general, the degree of visual acuity deterioration is variable, and early, multidisciplinary treatment can contribute to better disease control and reduce the occurrence of associated complications.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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