Reversible blindness in a patient with systemic lupus erythematosus: case report

Ochieng PO, Bandagi SS

Queens Hospital Center, Mount Sinai School of Medicine, Jamaica, New York

Abstract

Neuropsychiatric manifestations can occur in majority of patients with Systemic Lupus Erythematosus (SLE). We describe a patient who presented with acute onset binocular visual loss and was found to have inflammation of optic chiasma on imaging. The patient was treated with immunosuppression. She responded favorably and had complete visual recovery. We concluded that prompt management of atypical presentation of SLE can have enormous positive effect on outcome and quality of life.

Introduction

Systemic Lupus Erythematosus (SLE) is a multisystem disorder with multiple neuropsychiatric manifestations. Neuropsychiatric manifestations of SLE are common[1].

We present a rare case of SLE with isolated optic chiasmitis presenting with near total blindness that was reversed with treatment.

Case report

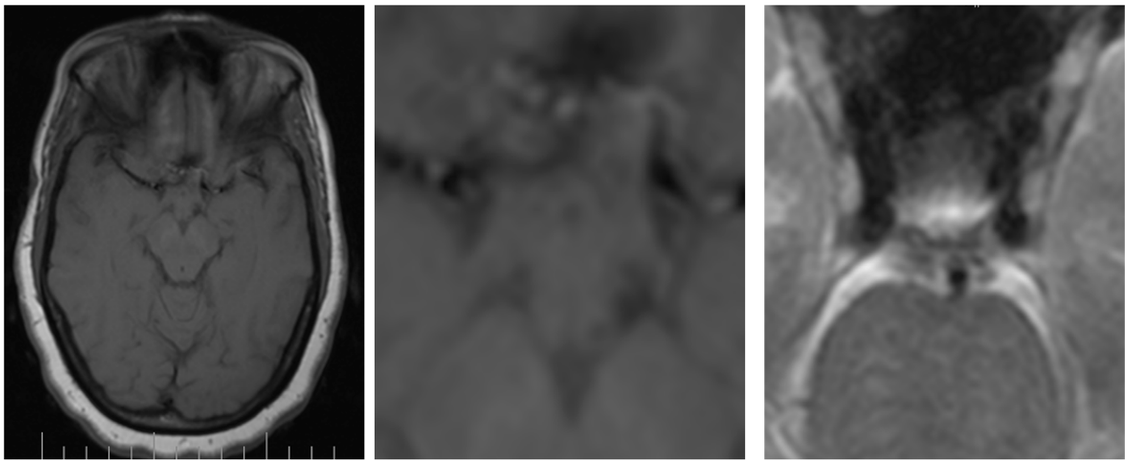
A forty eight year old black female with past medical history of remote pulmonary embolism and a ten year history of SLE presented with rapidly progressing loss of vision. She described acute onset blurring of vision affecting her peripheral vision with initial central sparing that ended in near complete bilateral loss of vision within two weeks. She reported no eye injury or pain, fever, headache, vomiting, weakness or numbness. She reported no rash, arthralgia, joint swelling or any other symptom. She had no similar symptoms in the past. Her only medication was hydroxychloroquine at a dose of 200 mg every 12 hours which she had taken for 8 years for SLE. She was a non-smoker and did not take alcohol or use drugs. She had one spontaneous abortion at gestational age of 8 weeks as her first conception followed by four full-term pregnancies with the last delivery at the age of 36.

On examination she was a well nourished female and was in no distress at rest. Her vital signs were within normal limits. Eye exam revealed visual acuity of 14/200 on the right and perception of light on the left with visual field and color perception testing was limited due to reduced visual acuity. The rest of the eye examination including fundoscopy was normal. Her systemic examination including the rest of neurological examination was within normal limits.

Hemogram revealed thrombocytopenia of 89,000/microLitre, hemoglobin of 12.2 g/dl and normal leucocyte count. CRP was elevated at 0.14 mg/dl (ref 0-0.8), Antinuclear Antibody (ANA) and anti-double stranded-DNA were positive at titers of 1:160 and 1:160 respectively with homogenous pattern of ANA. Anti-Ro was positive and Anti-La was negative. Her electrolytes, urea, creatinine and hepatic panel were unremarkable. Urinalysis was unremarkable. Lyme titer was negative. Anti-phospholipids, beta-2 glycoprotein and Anti-Neuromyelitis Optica (NMO) antibody were negative. Brain MRI revealed thickening, edema, and enhancement of the optic chiasm consistent with optic chiasmitis (Figure 1). Lumbar puncture results were normal with normal protein, no cells and negative for oligoclonal bands.

The hydroxychloroquine was discontinued at admission with initial concern for hydroxychloroquine toxicity. She was subsequently treated with intravenous pulse methyl prednisolone for 3 days followed by oral prednisone of 60 mg daily. She had gradual improvement of vision with full restoration of vision to visual acuity of 20/20 on both eyes in 6 weeks. Follow-up MRI was normal. The prednisone was tapered off gradually after 8 weeks and her vision remained normal on subsequent follow-up for over 1 year.

Figure 1: Pretreatment MRI of the brain (left- normal size T1 image, middle- T1 image zooming on the optic chiasma and right- T2 image zooming on the optic chiasma)



Discussion

This patient fulfilled at least four out of the eleven components of the diagnostic criteria for SLE with positive ANA, positive anti-double stranded DNA antibody, neurological manifestations and thrombocytopenia. There are at least 19 neuropsychiatric manifestations of SLE and they occur in up to 90% of SLE patients[1]. Most visual complaints from SLE result from retinopathy and anterior uveitis[2]. Optic chiasma involvement has however been rarely documented[3]. As in our case it is even more rare to have isolated optic nerve or optic chiasma involvement without other clinical manifestations. The leading differential diagnosis that was considered included glioma, granulomatous diseases like sarcoidosis and tuberculosis and lymphoma. Hydroxychloroquine eye toxicity as a possible cause of blindness prompted discontinuation of the medication although this was unlikely considering the absence of the pathognomonic corneal keratopathy characterized by whorl-like corneal epithelial deposits and maculopathy with "bull's eye" lesion or atrophy of the retinal pigment epithelium[4]. Another plausible differential diagnosis was neuromyelitis optica but this was considered less likely after the positive SLE markers and negative anti-NMO antibodies. A differential diagnosis for reversible blindness whose criteria this patient did not fulfill is posterior reversible leukoencephalopathy. This is a clinicoradiologic entity characterized by altered mental status, seizures and visual deficits associated with reversible changes on Magnetic Resonance Imaging (MRI) of the brain[5].

The pathophysiology of SLE optic chiasmitis remains an area of postulation and may involve auto-antibodies and immune dysregulation, vasculitis, non-inflammatory vasculopathy and thrombosis[6]. The inflammatory and immune pathogenesis is supported by response to immunosuppressants.

High dose steroid is the appropriate therapy for this sight threatening condition and the response is usually good if treated early. There have been few published case reports where alternative treatments like IV cyclophosphamide and methotrexate have been tried[7]. Hydroxychloroquine does not seem to have protective effect against this condition as observed both in our case and in the case reported by Frohman *et al*[3].

Conclusion

SLE remains a great medical masquerader and clinical vigilance is necessary in atypical presentations. Prompt therapy, therefore, can make an enormous quality of life difference in extreme presentations of SLE.

References

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