Blurred Vision as a Presenting Feature of Systemic Lupus Erythematosus: Blurred Vision, Blurred Diagnosis

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ABSTRACT

Benign/idiopathic intracranial hypertension is a diagnosis of exclusion in patients presenting with headache, particularly in young female patients. It is one of the rarest neuropsychiatric manifestations of systemic lupus erythematosus and only a few cases are described in the literature where it was a presenting feature of systemic lupus erythematosus. Herein, we report a case of a 34-year married female who presented with complaints of headache and blurring of vision for 3 months on a background history of hypothyroidism and pains in the small joints of the hands. She was diagnosed as benign intracranial hypertension with systemic lupus erythematosus as the underlying cause for it. She was successfully managed with immunomodulatory therapy.

Key Words: *Benign/idiopathic intracranial hypertension, Systemic lupus erythematosus, Blurred vision.*

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INTRODUCTION

Systemic lupus erythematosus (SLE) is a chronic autoimmune disease that can affect any organ system of the body, ranging from the skin to the brain.[1] There are 19 distinct neurological and psychiatric manifestations of SLE involvement of the brain as described by the American College of Rheumatology (ACR). Headache is the most common manifestation of neuropsychiatric SLE (NPSLE).[2] Herein, a case of idiopathic intracranial hypertension (IIH) is described, which is not included in the list of NPSLE manifestations as described by ACR but does constitute a unique and important differential of any patient of SLE presenting with headaches and blurring of vision. The first case of the association of IIH and SLE was reported in 1968 by Betmann.[3] Since then, several cases have been reported in the literature of this rare manifestation of the disease.

CASE REPORT

A 34-year female, a known case of hypothyroidism, presented with a history of pain in small joints of hands and myalgias of 5 months duration.

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There was involvement of proximal interphalangeal (PIP) and metacarpophalangeal (MCP) joints bilaterally with no swelling or redness. She also gave a 3 month history of headache which was generalised, more in the mornings and in lying position for which she had been using over-the-counter analgesics. She also noticed a blurring of vision with the headache but denied any history of complete loss of vision, diplopia, or any focal neurological deficit. She also mentioned experiencing generalised weakness and fatigability. A couple of months ago, she suffered from localised alopecia in her frontal region (Figure 1); however, no cause for this manifestation was sought at that time. There was no history of oral ulcers, rash, photosensitivity, Raynaud's phenomenon, pregnancy losses, or deep vein thrombosis. The medical history was significant for hypothyroidism. On examination, she had a weight of 70 kg and a body mass index (BMI) of 27.34 kg/m^2. The vital signs showed a blood pressure of 130/80 mmHg, pulse of 84/min, and respiratory rate of 17/min. She was afebrile. Ophthalmological examination revealed normal-sized reactive pupils with bilateral grade IV papilledema with normal intraocular pressures on applanation tonometry (15 and 12 mmHg in right and left eyes, respectively). The visual fields on perimetry showed early nasal scotomas bilaterally. Extraocular movements and the rest of the cranial nerve examinations were normal. The neurological limb examination revealed a power of 4+/5 in both upper and lower limbs. Deep tendon reflexes and creatine phosphokinase (CPK) were normal. The MCP and PIP joints were tender and mildly swollen. Apart from the patchy alopecia, there were no other pertinent examination findings related to connective tissue disease. The relevant laboratory and radiological investigations are shown in Table I.

Table I: Laboratory and radiological investigations.

Laboratory investigations	Values
Haemoglobin	11.9 g/dl
Total leucocyte count	11900/mm^3
Platelets	290.000/mm^3
Prothrombin time	13/13 seconds
Activated partial thromboplastin time	30/26 seconds
International normalised ratio	1.0
Bilirubin	0.4 mg/dl
Alanine aminotransferase	56 IU/L
Urea	40 mg/dl
Creatinine	0.6 mg/dl
Sodium	138 mEq/l
Potassium	4.6 mEq/l
Urine routine examination	No blood, proteins, casts or white cells
Hepatitis B surface antigen	Both negative
Anti-HCV antibodies	
Electrocardiography	Normal
Chest x-ray	Normal
Echocardiography	Normal
MRI, MRV, and MRA brain with contrast	Normal
Fundoscopy	Bilateral Grade IV papilloedema
CSF routine examination	Opening pressure raised at 30 cm of water in the lateral recumbent position. Proteins 171 mg/dl WBCs: 150 ×10^6/L (100% lymphocytes) Glucose: Normal Gram and AFB stain: negative CSF culture: negative
ANA	Positive
Anti-dsDNA	48.5 IU/ml (Negative: <10 IU/ml)
ENA profile	
U1 RNP	50.79 (>5: positive)
SS-A/Ro	13.94 (>5: positive)
Anti-Sm antibodies	14.77 (>5: positive)
Lupus anticoagulant	Ratio= 1.5 (>1.1: positive)
β2-glycoprotein 1	Both negative
Anticardiolipin antibodies	
C 3 levels	61 mgl/dl (Normal: 83-193)
C 4 levels	12.6 mg/dl (Normal: 15-57)
Thyroid profile	
FT4	0.97 ng/dl (Normal: 0.70-1.48 ng/dl)
TSH	5.6 u IU/ml (Normal: 0.35-4.94 u IU/ml)

After excluding all the common causes for headache and blurring of vision in a young female, the final diagnosis of IIH was made in collaboration with the neurologist and the ophthalmologists. Moreover, IIH was noted to be the initial presenting symptom of SLE in this patient, which is very rare and has been reported in a couple of patients only.

The patient was started on pulse methylprednisolone, 500 mg, IV once a day for a total of five doses alongside vitamin D and calcium supplementation and proton pump inhibitors. She was also prescribed acetazolamide at a dose of 250 mg three times a day to lower the intracranial pressure along with hydroxychloroquine for her SLE.

After pulse methylprednisolone, immunomodulatory therapy using cyclophosphamide was commenced to halt the disease process. Following the first dose of cyclophosphamide, she was discharged home on prednisolone at 1 mg/kg body weight and was called for subsequent cyclophosphamide pulses according to the Euro Lupus regimen. A repeat lumbar puncture to relieve the pressure was also done which showed that the pressure had reduced to 20 cm of water after one and a half months. She had improved symptomatically over the course of treatment and her papilloedema settled completely after the third dose of cyclophosphamide. Unfortunately, optical coherence tomography (OCT) could not be done prior to starting treatment but it was done after the third dose of cyclophosphamide and showed some permanent damage to visual field (Figure 2). She has currently completed her induction therapy with cyclophosphamide and is on mycophenolate mofetil as a maintenance therapy.

Figure 1: Patchy scaring alopecia.

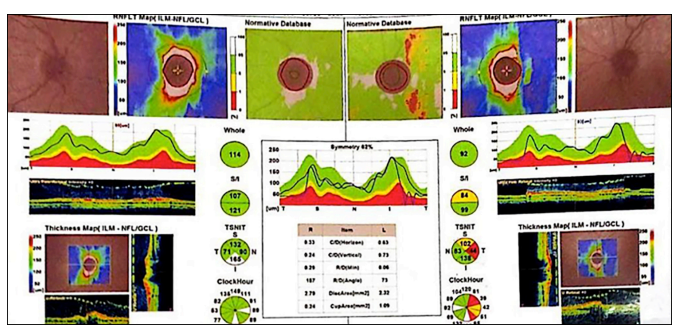

Figure 2: Optical coherence tomography (OCT) results.

DISCUSSION

In this case report, we have described IIH as the presenting feature of SLE. This patient fulfilled the following clinical criteria; alopecia, arthritis, neuropsychiatric manifestation i.e headache secondary to IIH and immunological criteria of positive ANA, elevated anti-dsDNA and positive anti-Smith antibody, to be diagnosed as a case of SLE based on the Systemic Lupus International Collaborating Clinics (SLICC) criteria.[4] This patient also fulfilled the criteria for diagnosis of IIH, as rest of the causes of raised intracranial pressure, like vascular, structural, and infective causes were ruled out.

Two very large retrospective cohort studies on the causal association of IIH and SLE demonstrated that out of 1084 patients of SLE, eight had IIH.[5] Alternatively, 1% of the cohort of 410 patients diagnosed with IIH had SLE.[6]

The pathophysiology behind the development of IIH in SLE patients is twofold. Firstly, there is disruption of the normal absorption of the cerebrospinal fluid as the arachnoid villi are damaged by the immune-mediated mechanisms of SLE. Secondly, there is abnormal venous outflow which is attributed to hypercoagulability due to concomitant nephritis or the presence of antiphospholipid antibodies leading to microthrombotic phenomenon.[7] In this patient, SLE was active as evidenced by

the high titers of anti-ds DNA and low complements contributing to the immune-mediated damage. Weakly positive lupus anticoagulant could also be a contributing factor in this patient.

The best way to treat IIH is to remove the underlying cause e.g. withdrawal of causative agents, like tetracyclines, high doses of vitamin A, retinoids, steroids, and oral contraceptive pills, or by controlling obesity or other conditions like SLE. Measures like therapeutic lumbar punctures and medications to reduce intracranial pressure like acetazolamide, furosemide, and mannitol in certain cases are also undertaken. Steroids are generally not indicated unless waiting for a surgical correction. However, in patients with underlying autoimmune phenomena like SLE as was seen in this case, steroids in high doses along with immunosuppressive therapy are needed as documented by Shah *et al*. from Nepal, Rajasekharan *et al.* from India and, Kalanie from Iran and many others as well.[8-10]

In conclusion, headache is the most common neuropsychiatric manifestation of SLE; however, sometimes it is the harbinger of other potentially dangerous pathologies so it should always be investigated. IIH, though a rare association, should always be watched out for as prompt diagnosis and treatment can save a patient's vision and the long-term morbidity associated with it.

PATIENT'S CONSENT:
Written informed consent was obtained from the patient.

COMPETING INTEREST:
The authors declared no competing interest.

AUTHORS' CONTRIBUTION:
TK: Conceptualisation, referencing, and proofreading.
SA: Literature review, proofreading.
SN: Literature review, critical reviewing of the manuscript.
SSA: Manuscript writing, literature review.

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