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IDIOPATHIC INTRACRANIAL HYPERTENSION AS AN INITIAL MANIFESTATION OF SLE: A RARE CASE REPORT

Dr. Ankith Krishnakumar	Junior Resident, Department Of General Medicine, Mahatma Gandhi Medical College And Research Institute, Pondicherry
Dr. Thumbanatham A	Professor, Department Of General Medicine, Mahatma Gandhi Medical College And Research Institute, Pondicherry
Dr. Dhruvi Reddy	Junior Resident, Department Of General Medicine, Mahatma Gandhi Medical College And Research Institute, Pondicherry

ABSTRACT Systemic Lupus Erythematosus (SLE) is an autoimmune illness affecting mainly females and presents classically with malar rash, arthritis, and renal damage. Neuropsychiatric manifestations of this disease have been well established and manifests most commonly as cognitive impairment. Widespread inflammation has been proposed to alter CSF flow and can lead to raised intracranial tension. Idiopathic intracranial hypertension presents usually as headache and blurring of vision. It has also known to be a manifestation of lupus. This is a case of a 36-year-old women who presents with headache and blurring of vision. Fundus evaluation showed Papilledema suggestive of raised intracranial tension. MRI brain showed no mass effects of hydrocephalus. Autoimmune workup confirmed a diagnosis of SLE. This case highlights the importance of the need for a high suspicion of SLE as it can present with a wide variety of symptoms. IIH if left untreated can lead to major complications such as vision loss.

KEYWORDS : systemic lupus erythematosus, idiopathic intracranial hypertension, headache

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a disorder characterized by clinical symptoms and signs such as headache, papilledema and vision loss which is attributed to elevated intracranial pressure, and presence of no other cause of intracranial hypertension evident on neuroimaging. Idiopathic intracranial hypertension is an unusual manifestation of neuropsychiatric SLE.

Case Report

A 36-year-old female patient with no known comorbidities presented to the hospital of mahatma Gandhi medical college in Puducherry with complaints of headache for 3 weeks, intermittent type, in the frontal and retro-orbital region, aggravated during early morning.

Headache was also associated with vomiting, which was non projectile. Patient also complained of transient blurring of vision. Patient had no significant previous medical history. Menstrual history was normal. There was no history of any skin lesions or rashes, photosensitivity, hair loss, any prolonged drug intake. On arrival to the hospital vitals were stable and systemic examination was within normal limits. Fundus evaluation was done which showed features of b/l papilledema.

MRI brain was done which showed features consistent with papilledema and raised intracranial tension. Patient was provisionally diagnosed as a case of idiopathic intracranial hypertension and was started on Tab. Acetazolamide.

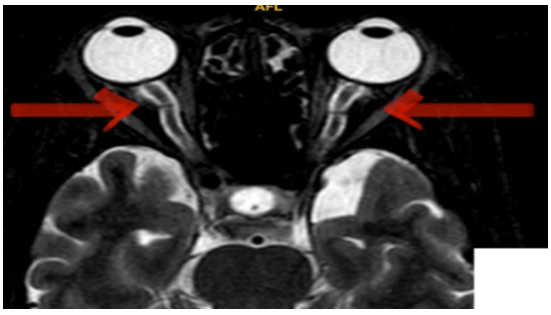

Figure 1: Brain Magnetic Resonance Imaging: T2 Flair Axial Section Showing Bilateral Mildly Bulky Tortuous Optic Nerves With Posterior Scleral Flattening Which Are Consistent With Papilledema.

Laboratory investigations showed pancytopenia with total counts of 2000, Platelets of 70,000 and hemoglobin of 8.4. Direct Combs test was positive. Urine routine examination showed albuminuria.

ANA was positive. C3, C4 levels were reduced and Anti – dsDNA was positive. Patient was diagnosed to have SLE as per SLICC criteria.

Patient was started on Pulse steroid therapy with Inj.Methylprednisolone and as per NIH protocol was initiated on monthly injections of cyclophosphamide. Renal biopsy could not be done due to logistic reasons. Patient was reviewed regularly. Patient's headache improved and repeat fundus evaluation showed resolving papilledema.

DISCUSSION

The above case shows an atypical manifestation of SLE with the patient presenting as a case of Idiopathic intracranial hypertension.

Idiopathic intracranial hypertension is manifested by elevated intracranial pressures without any mass effect or CSF abnormalities[1]. SLE is autoimmune in nature, and affects multiple systems with production of numerous autoantibodies such as ANA, Anti – dsDNA etc[2]. It is a disease mainly seen in females with a female to male ration of 9: 1. The etiology involves genetic susceptibility and HLA DR-3 AND HLA-DR 2 susceptibility. There is activation of innate immune system, T cells and B cells. Neurological manifestations of Lupus were seen in 25 to 75 % of patients and most commonly presents as cognitive impairment. Bettman *et al* reported a case of IIH and SLE in 1968[3]. Reasons for development of benign intracranial hypertension in patients with SLE mainly include autoimmune injury to the arachnoid villi leading to reduced CSF absorption, or a pro-thrombotic state causing obstruction of vessels in CNS[4].

Treatment of idiopathic intracranial hypertension in SLE mainly includes corticosteroids. Intracranial pressure can be lowered with drugs such as acetazolamide, diuretics such as

loop diuretics and intravenous mannitol. If left untreated IIH can lead to vision loss. Other conditions which are associated with IIH include Hypothyroidism, Hypervitaminosis A and anti-phospholipid antibody syndrome.

CONCLUSION

Idiopathic intracranial hypertension presenting with severe headache maybe the initial symptom of SLE[5]. Lupus can present in multiple ways and may not always present with classical symptoms such as malar rash. This case indicates the need for high degree of suspicion for lupus particularly in women of reproductive age group. A thorough search into the cause for IIH and treatment of that cause is required to prevent complications.

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Ethical Approval: Not required

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