Case Report

Rare presentation of systemic lupus erythematosus with idiopathic intracranial hypertension

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Idiopathic intracranial hypertension (IIH) is an extremely rare condition and rare in systemic lupus erythematosus (SLE). Herein we report a case of young lady presenting with increased intra-cranial pressure (ICP), normal neurological examination and imaging. She was diagnosed with IIH and responded well to steroid and cyclophosphamide. Meticulous clinical and laboratory assessment must be carried out in suspicious case to avoid misdiagnosis. (Rawal Med J 201;43:360-362).

Keywords: Idiopathic intracranial hypertension (IIH), systemic lupus erythematosus (SLE), lupus cerebritis.

INTRODUCTION

Idiopathic intracranial hypertension (IIH) is a rare neuropsychiatric manifestation of SLE. It is a diagnosis of exclusion that is defined by modified Dandy criteria comprising of signs and symptoms of increased intracranial pressure (headache, blurring of vision etc.), absence of localized neurological signs, except unilateral/ bilateral abducent nerve palsy, increased cerebrospinal fluid pressure with no identifiable cause, normal CSF results, normal neuroimaging studies with no ventricular dilatation and awake and alert patient.[1] We herein present the rare occurrence of IIH in a SLE patient who has fulfilled this criteria.

CASE PRESENTATION

An 18 year old Malay lady who had no known medical illness, presented with throbbing frontal headache, altered behavior and cognitive impairment for two weeks. Headache was associated with vomiting and giddiness. She had become more forgetful, emotionally labile and socially withdrawn. She also complained of pain involving multiple interphalangeal joints of both hands with oral ulcers, photosensitivity and increasing hair loss over past one year. There was no seizure, blurring of vision or focal neurological deficit to suggest intracranial space occupying lesion. There was no fever or neck stiffness.

She was afebrile and hemodynamically stable. There was alopecia with bilateral, multiple interphalangeal joint tenderness with no deformity or swelling. Mini Mental State Examination (MMSE) showed moderate degree of cognitive impairment with scoring of 16/30, full neurological examination was normal. Ophthalmology examination was totally normal. Rest of examination was normal. Laboratory work up revealed pancytopenia, high ESR positive ANA with high titer of 1:2560, speckled pattern and hypocomplementemia. However, anti double stranded ANA was negative (Table).

CT angiography and MRI of brain were normal, so was CSF analysis (Table). A diagnosis of SLE was made according to American College of Rheumatology (ACR) SLE classification criteria, with neuropsychiatry symptoms and IIH.

Table. Laboratory results.

Test	Value	Unit	Normal range
White cell count	2.28	x 10^3 / uL	4.0 – 10.0
Hemoglobin	9.5	g/dL	12.0 -15.0
Platelet	22	x 10^3 / uL	150 - 400
ESR	55	mm/hour	< 30
CRP	2.29	mg/L	< 5
Complement 3	0.57	g/L	0.90 – 1.80
Complement 4	0.26	g/L	0.10 – 0.40
Anti-nuclear antibody	1 : 2560 , speckled pattern		
CSF findings:	Open pressure: 30.5cm H2O		
Total protein	21	mg/dL	10–40
Glucose	4.2	mg/dL	4.0–7.0
Cellular count	0		

She received pulse intravenous methylprednisolone 500 mg daily was given for 3 consecutive day followed with oral prednisolone at 1 mg/kg/day. Concurrently, monthly intravenous cyclophosphamide pulse at a dose of 500 mg was administered and planned for six cycles. She was also commenced on oral hydroxychloroquine 200 mg daily and calcium supplements.

The pancytopenia recovered and MMSE showed significant improvement from scoring of 16/30 prior methylprednisolone pulse therapy to 21/30 post pulse therapy reaching 30/30 after further three days. Clinically, the headache had resolved completely. We managed to taper prednisolone dose as patient currently in partial remission after three doses of methyl prednisone and cyclophosphamide.

DISCUSSION

Idiopathic intracranial hypertension is an extremely rare condition with annual incidence of 0.9/100,000 persons and 3.5/100,000 in females 15 to 44 years of age.[2] Our patient fulfilled all the modified Dandy criteria of IIH. Risk factors for IIH include overweight, women of child bearing age, medication such as corticosteroid, oral contraceptive pills, nalidixic acid, nitrofurantoin, tetracycline and nutritional such as vitamin A deficiency. It also associated with medical condition such as hypertension, sarcoidosis, autoimmune disease such as SLE, as seen in our case.[3]

Hershko et el in his 26 years experience of SLE with IIH reported that 10 of 651 hospitalized SLE patients (prevalence of 1.5%) were found to have IIH, exceeding the prevalence of 1 to 19 per 100,000 (prevalence of 0.001-0.02) reported in the general population.[4] IIH is usually associated with positive anti-ribosomal P protein and anti-neuronal antibody.[5] Our patient has risk factors of young female, serologically active SLE (low C3).

Pathophysiology of IIH in SLE is not clearly understood. It is postulated as impairment of venous outflow and CSF absorption due to the following mechanism: 1. Immune mediated injury within arachnoid villi and subsequent reduction in CSF absorption.[6] 2. Hypercoagulable state secondary to APLS, lupus nephritis with low levels of antibodies to thrombin III, presence of fibrin split products giving rise to micro-obliteration of cerebral arteriolar/ venous system, hence lead to impaired CSF absorption.[7-9] 3. Anemia, may cause increased cerebral blood flow leading to IIH, however, anemia (Hb 9.5g %) in our case report was not severe enough to cause this.

In general population, obesity is a risk factor, however, all 8 SLE patients with IIH in a study by Kim et al had acceptable BMI.[10] Our patient had BMI of 25kg/m^2. Mainstay of treatment for IIH in general population is by mannitol, acetazolamide and weight loss, where as in SLE is treated by corticosteroid followed by immunosuppressant cyclophosphamide,[4] suggesting that IIH in SLE is more of immune mediated.

In summary, IIH can be the first sole CNS manifestation of SLE. It is an important differential diagnosis to consider in young lady presenting with symptoms of increased ICP (headache, vomiting) but normal neuroimaging. Thorough history and examination must be done to avoid misdiagnosis, and in suspicious cases, meticulous investigations should be carried out.

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