Idiopathic intracranial hypertension as a cause of severe intractable headache in a patient with Systemic Lupus Erythematosus: A case report from Eastern Nepal

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Abstract
The common risk factors for Idiopathic intracranial hypertension are obesity, female gender, hypervitaminosis A, and steroid withdrawal. Even though Idiopathic intracranial hypertension is considered as a neuropsychiatric manifestation of Systemic lupus erythematosus, it is often missed by the physician as a cause of a headache in a patient with Systemic lupus erythematosus. We report a case of 21-year-old female who presented in our outpatient department with a history of a severe intractable progressive headache for a duration of four weeks and blurring of vision for five days who was later diagnosed as a case of idiopathic intracranial hypertension with Systemic lupus erythematosus. She recovered dramatically with the institution of steroid and acetazolamide therapy.

Introduction

The prevalence of Idiopathic intracranial hypertension (IIH) or pseudotumor cerebri in the Western world was 0.9/100000 person in females of 15-44 years age group.[1] The common risk factors for IIH are obesity, female gender, hypervitaminosis A, and steroid withdrawal.[2] Bettmen J reported the first case of idiopathic intracranial hypertension in patients with Systemic Lupus Erythematosus (SLE) in 1968 A.D. Since then there were sporadic case reports of association of IIH and SLE from different parts of the world.[3] Headache is a common feature of SLE and considered as one of the neuropsychiatric manifestations of SLE.[4] The common causes of a headache in patients with SLE are migraine, tension headache, aseptic meningitis, central nervous system vasculitis, sagittal vein thrombosis and steroid withdrawal.[5] Even though IIH is one of the neuropsychiatric manifestations of SLE, it is often missed as a cause of a headache in a patient with SLE. Here, We report a case of 21-year-old female who presented with severe intractable headache and diagnosed as a case of Systemic lupus erythematosus with idiopathic intracranial hypertension.

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Case Report

TA 21-year-old female patient presented to the outpatient department of Internal Medicine of B.P. Koirala Institute of Health Sciences, Dharan, Nepal with presenting complaints of headache for a duration of four weeks occurring mostly in the bi-frontal and occipital region, throbbing in nature, severe in intensity with 9/10 in pain scale and restricting her daily activities. The headache was aggravated on lying down position and occurred mostly in the morning time. The intensity of a headache decreased with the sitting position. It was associated with vomiting. She also had history blurring of vision and double vision for five days prior to presentation to our hospital. The double vision occurred on looking laterally and with both eyes open and improved on the closure of one eye. She also had the history of photosensitivity and hair loss as well. There was no history of the seizure, trauma to head and fever. She had visited two nearby hospitals prior to presenting to our centre. She was prescribed Tablet Ibuprofen 400 mg per oral thrice a day and tablet tramadol 50 mg once a day but that didn't relieve her headache. Past history was not significant. Family history did not reveal a chronic headache and autoimmune disease in the first-degree relative.

On examination, she was conscious, oriented to time, place and person. She had blood pressure of 100/60 mmHg, respiratory rate- 18 cycle/minute, pulse rate- 92 beat/minute, temperature- 98oF. She was pale but icterus, oedema, cyanosis, and clubbing was absent. Her weight was 45 Kg, height was 1.51 meter and Body mass index was 19.7 kg/m2. On systemic examination finding, there was bilateral 6th cranial nerve palsy. The other central nervous system findings, respiratory system, gastrointestinal system and cardiovascular system revealed no abnormality. A consultation was done with an ophthalmologist who reported the bilateral papilledema and bilateral lateral rectus palsy in the patient. The photograph of the fundus of the right eye of the patient at the first visit was shown in Figure 1.

Table 1: Baseline laboratory reports of the patient

Variables	Values	Normal values
Haemoglobin	5.7 gm/dl	11-16gm/dl
Total leucocyte count	4600 cell/mm cu	4000-11000 cell/ mm cu
Platelet	210000 Cell/ mm3	1,50000-4,00000Cell/mm3
Reticulocyte count	7.8	
Serum Iron	70 ug/dl	60-150 ug/dL
Serum UIBC	91ug/dL	250-400 ug/dL
Serum Ferritin	2034 ug/L	15-150 ug/L
Serum LDH	1370 U/L	125-135 U/L
Direct comb test	Negative	
Serum Urea	16 mg/dL	15-40 mg/dL
Serum Creatinine	0.4 mg/dL	0.6-1.3 mg/dL
Plasma ANA	38.4 U/ml	<10U/ml
Plasma Anti-ds DNA	50.45 IU/ml	<30 IU/ml
C3 Level	108 mg/dl	90-180 mg/dl
C4 Level	21mg/dl	10-40 mg/dl
24- hour urine protein	0.57g/day	<0.15 g/day
TSH	3 mIU/L	0.4-6.2 mIU/L

Abbreviation: UIBC Unbound iron binding capacity, LDH- lactate dehydrogenase,
ANA- Antinuclear antibody, Anti-ds DNA- Anti double-stranded deoxyribonucleic acid, C3- complement 3, C4- complement 4, TSH- thyroid stimulating antibody

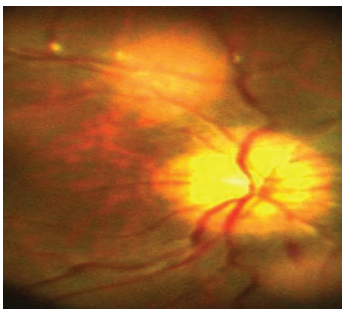

Figure 1: Fundus showing blurring of disc margin,elevation of optic disc, engorgement of veins

The baseline investigation reports of the patient are illustrated in Table 1. The magnetic resonance imaging (MRI) of the brain revealed no abnormality as shown in Figure 2. A Lumbar puncture was done on the 4th day of hospital admission. The cerebrospinal fluid analysis showed no cell count, protein level - 30 mg/dl and glucose level 80mg/dl.

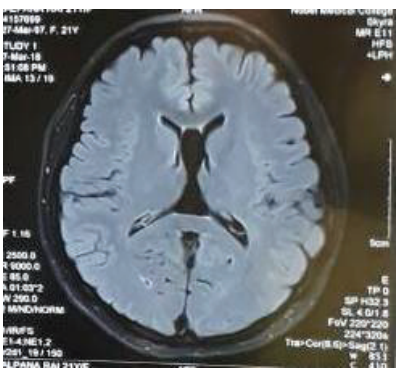

Figure 2: Magnetic Resonance Imaging of brain showing normal sized ventricles

The patient was treated with injection Methylprednisolone 1gm intravenous once daily for five days, Tablet acetazolamide 500 mg thrice a day and tablet folic acid 5mg once daily. The headache improved with treatment and headache severity decreased from 9/10 to 3/10 on the pain scale. We discharged the patient on oral prednisolone 50 mg once a day, tablet acetazolamide 500 mg thrice a day and tablet folic acid 5 mg once a day after five days of hospital admission. The patients had a follow-up visit two weeks after hospital discharge and there was a significant improvement in her headache and vision. The photograph of the fundus taken by Optical Coherence Tomography (OCT) after two weeks of treatment with steroid and acetazolamide is shown in figure 3.

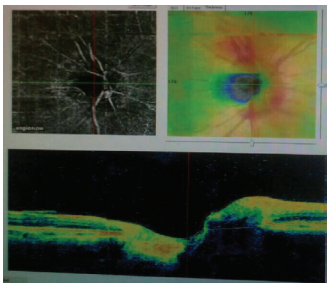

Figure 3: Optical coherence tomography showing minimal blurring of disc margin, normal optic disc, normal vessels.

Discussion

Here we reported the case history of a 21-year-old female who presented in our centre with a history of severe intractable headache and blurring of vision. She was diagnosed as a case of Systemic Lupus Erythematosus with idiopathic intracranial hypertension. Her headache subsided with steroid and osmotic diuretic therapy.

She was diagnosed as a case of SLE on the basis of fulfilling the clinical criteria of alopecia, haematological manifestation (autoimmune hemolytic anaemia), renal manifestation (24-hour proteinuria of more than 500mg/day), neuropsychiatric manifestation (headache) and immunological criteria of positive ANA and elevated Anti-ds DNA.[6] Idiopathic intracranial hypertension was established as she had symptoms and signs of increased intracranial pressure (headaches, transient obscurations of vision, papilledema), no localizing neurologic signs other than bilateral VI nerve paresis, CSF can show increased pressure with no cytologic or chemical abnormalities, and normal to small symmetric ventricles as demonstrated by the CT scan/ MRI of head.[7]

Our patient presented to the outpatient department with a history of severe intractable headache and blurring of vision. A similar case report was done by Sahey P et al. where a 13-year-old girl was presented with a severe headache and blurring of vision and later diagnosed as SLE with idiopathic intracranial hypertension.[8] The reported clinical manifestation of IIH in SLE were headache, vomiting, blurring of vision and diplopia as.[9] Kim J et al. reported that among 1084 patients of SLE, eight patients were diagnosed with idiopathic intracranial hypertension.[10] Dave S et al. in their retrospective study among a cohort of 410 IIH patients demonstrated that the percentage of patients with SLE was 1%.[11] These all case reports support the possible significant association between IIH and SLE, however, the definite association between the IIH and SLE was not established yet. However, there were literature proposing the pathogenesis of IIH in SLE are venulitis, aseptic meningitis, immune complex deposition or micro-occlusion of arachnoid villi.[12]

The recommended medical treatment of IIH in obese patients are weight reduction, acetazolamide, frusemide and topiramate. Steroids are not recommended unless waiting for the surgical intervention in the fulminant condition of IIH.[9] In contrast to the guidelines for obese IIH, We have treated our patient with steroid and acetazolamide as our patients had IIH with SLE. Rajasekharan C et al reported a case of 14-year-old girl diagnosed as IIH associated with SLE who had dramatic recovery of signs and symptoms of raised Intracranial tension with treatment of steroid.[13] Similar improvement in a headache and vision with treatment with steroid in a patient with IIH with SLE was reported by Masai H et al .[14] Our patients too improved symptomatically with the steroid and acetazolamide therapy. Patients with IIH and SLE should be treated with steroids. As far as authors' knowledge this is the first case report of Idiopathic intracranial hypertension presenting as the first manifestation of Systemic Lupus Erythematosus from Nepal.

The measurement of the CSF opening pressure was not possible due to the lack of facilities in our centre. This case highlights the need for a study to conduct among a large population of SLE to determine the prevalence of IIH among SLE patients.

Conclusion

Even though Idiopathic intracranial hypertension is mostly associated with the obesity, hypervitaminosis, steroid withdrawal, the possibility of SLE as its cause should be kept as differential diagnosis especially in non-obese female with IIH. The cause of a headache in patients with SLE ranges from migrainous to non-migrainous, however, the treating physician should keep IIH as one possibility if SLE patient presented with severe intractable headache.

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