



Case Reports & Case Series

Intracranial hypertension as an initial clinical manifestation of Systemic Lupus Erythematosus

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ABSTRACT

Background: Systemic Lupus Erythematosus (SLE) is an auto-immune systemic disorder that may affect multiple organ systems including central nervous system. Although involvement of the nervous system and eye is not uncommon in SLE, the presence of intracranial hypertension (IH) with papilledema in SLE a rare occurrence and it requires high level of suspicion to be diagnosed, especially when the combination of IH and papilledema is the initial presentation of SLE.

Case presentation: A 21-year-old female presented with a month long history of progressively deteriorating bilateral frontotemporal headache of throbbing quality. In neurological examination the only finding was bilateral papilledema with mild peripheral visual field Constriction. A brain Magnetic Resonance Imaging (MRI), MR angiography and venography showed no abnormal findings. Cerebrospinal fluid (CSF) analysis revealed a high opening pressure (320 mm H₂O) with normal CSF indices. She also had normocytic, normochromic anemia, mild thrombocytopenia, increased quantitative CRP, ESR and LE cell was also reported. Upon further work-up, it was revealed that she had positive ANA and Anti-ds-DNA, decreased level of complement C3 and C4. She, with SLE, was treated with corticosteroid which resulted in significant improvement in patient's manifestation of IH.

Conclusions: The association of IH and SLE has already been established and one should be aware of this co-existence and in IH cases, SLE should also be considered as an underlying cause and subsequently appropriate diagnostic tests should be performed.

1. Introduction

Systemic Lupus Erythematosus (SLE) is a chronic, auto-immune, multi-system disease that is believed to be a complex interplay of genetics, infectious, and Immunologic factors. It is particularly prevalent in women often with relapsing remitting course. The disease is characterized by diverse manifestations encompassing almost all organ systems. Neuropsychiatric Lupus may range from subtle cognitive or behavioral disorders to coma and death. Intracranial hypertension (IH) is included among the rare neuropsychiatric manifestations of SLE [1]. We discuss here a case of a 21-year old female who presented with only features of IH and on evaluation we found her to have Systemic Lupus Erythematosus.

2. Case report

A 21-year-old girl was admitted to our neurology service at Mehr

Hospital, Tehran, Iran, with a month history of progressively deteriorating bilateral frontotemporal headache of throbbing quality. Headache was worse in morning and upon lying flat. She also complained of episodes of transient visual blurring along with nausea sensation. She had no remarkable past medical history, was afebrile and denied diplopia, dysuria, throat pain, rash, cough, dyspnea, joint pain or abdominal pain. She had no seizure or behavioral abnormalities but complained of loss of appetite and loss of weight for the past 4 to 6 weeks. Upon examination she had normal vital signs. Her height was 154.5 Cn, her body weight was 55 kg and her body mass index indicated that she was not obese. Her general medical examination was unremarkable. She had no skin rash, organomegaly or lymphadenopathy. In neurological examination the only finding was bilateral papilledema (Fig. 1) with mild peripheral visual field Constriction, which was confirmed by neuro-ophthalmologist. Her pupils were symmetrical and reactive to light. She had no other cranial neuropathies, no abnormal motor or sensory signs, no pathological reflexes and had normal

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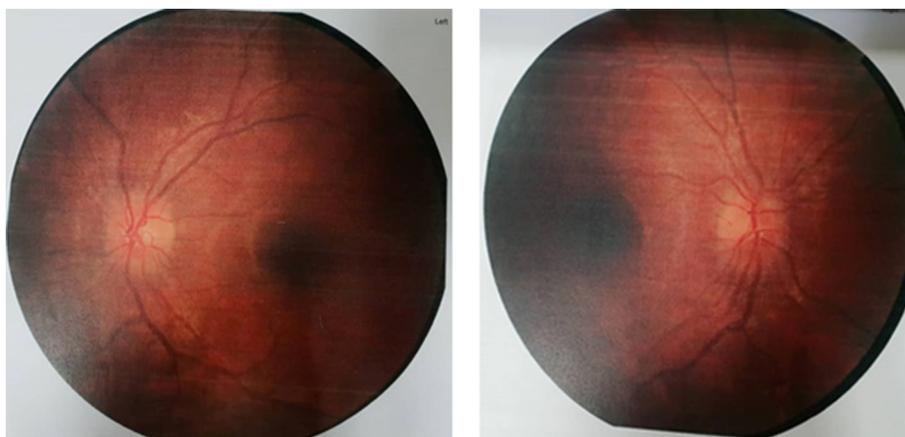


Fig. 1. Fundus photography of both eyes (bilateral papilledema).

gait and coordination. The laboratory findings showed normocytic, normochromic anemia (Hb: 11.3) mild thrombocytopenia (PLT: 125,000) and increased quantitative CRP (10 mg/l).

Her ESR was 60 mm/h and an LE cell was present. Further work-up revealed Positive ANA, positive Anti-ds-DNA, Decreased level of complement C3 and C4, and negative VDRL, RE, and anti-phospholipid antibodies. Rest of laboratory tests including liver, renal and thyroid function tests, fasting blood sugar, electrolytes, PT, PTT, folate and B12 levels, homocysteine level, serum protein electrophoresis PPD skin test, urinalysis were normal. An abdominopelvic sonography and a chest x-ray did not show any pathology. A brain Magnetic Resonance Imaging (MRI), MR angiography and venography showed no findings of ischemia, thrombosis or vasculitis. Cerebrospinal fluid (CSF) analysis revealed an extremely high opening pressure (320 mm H₂O) with normal CSF indices and negative cytology for malignancy or infection. These findings indicated that patient had IIH with papilledema. A diagnosis of definite SLE was made on day 5 of admission according to the criteria of American College of Rheumatology (6–7). Intravenous Methylprednisolone pulse therapy (500 mg/day) was administered immediately for three days, followed by oral treatment with 60 mg of prednisolone daily. The papilledema and thrombocytopenia rapidly improved over the course of a week. Then she was referred to rheumatology department for follow up therapy with monthly intravenous cyclophosphamide pulse.

3. Discussion

The patient fulfilled American College of Rheumatology (ACR) diagnostic criteria for SLE [2]. She had hematological manifestations (anemia and thrombocytopenia), positive ANA, positive ds DNA, positive LE cell and neuropsychiatric manifestation of SLE (headache due to IIH). She also fulfilled modified Dandy criteria [3] for IIH which is defined as an elevated intracranial Pressure (CSF pressure > 250 mm H₂O), no clinical, laboratory or radiological evidence for structural lesions, no localizing signs, normal CSF composition and no unexplained symptoms or signs. Nervous system disease involvement in SLE varies widely and may occur in up to 50% of patients in the course of the disease. Headache has been classified by the ACR as a neuropsychiatric manifestation of SLE, however IIH is rarely a presenting manifestation

of the disease. Our case is probably the first case of SLE presenting itself as pure IIH. So far two cases with similar title are mentioned in the literature but besides IIH both had other symptoms: one had fever and reddish scaly lesions over her cheek [4], and second had joint pain and swelling to the wrists and knees [5]. The pathogenetic mechanism of IIH in SLE is not yet fully understood. It is not clear whether the overproduction of CSF, relative absorptive failure of CSF or increased cerebral blood flow is associated with the pathogenesis of IIH in SLE. Immune-mediated injury within the arachnoid villi and the resultant decrease in CSF absorption and/or thrombotic obliteration of cerebral arteriolar and venous system due to a hypercoagulable state are among the most probable mechanism.

Corticosteroid was effective for IIH in the present case. Most of the IIH in SLE cases can be treated using high-dose corticosteroids followed by immune-suppressants, which differ from IIH cases in general, which are treated using acetazolamide or mannitol and weight loss.

4. Conclusion

The association of IIH and SLE is probably not coincidental and one should be aware of this coexistence and in facing IIH seriously have SLE in mind and perform appropriate diagnostic tests for the patient.

Conflict of interest

All authors declare that they have no conflict of interest.

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