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CNS Vasculitis in A Patient with Systemic Lupus Erythematosus: A Case Report

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ABSTRACT

Systemic Lupus Erythematosus (SLE) is a chronic idiopathic autoimmune disease with multi-system involvement. Clinical manifestations range from mild constitutional symptoms to life-threatening systemic manifestations. Neurological involvement in SLE occurs in 10 to 80 percent of patients and has a broad spectrum of symptoms, including chronic headache, impaired cognitive function, seizures, delirium, psychosis, vasculitis, and thromboembolic events. The pathophysiology by which SLE affects the nervous system includes vasculopathy, auto-antibodies, secondary factors (infections, metabolic dysfunction, and drug-induced), and other miscellaneous factors like inflammatory mediators (cytokines, chemokines, neuropeptides, nitric oxide etc.).

Keywords: SLE, vasculopathy, auto-antibodies, secondary factors.

CASE REPORT

A 32-year-old female who is a known case of SLE of 2 years and not on regular treatment for the past 6 months presented with the complaints of low-grade intermittent fever for the past 3 months not associated with chills or rigors and headache for the past 2 months not associated with blurring of vision, photophobia or vertigo. History of weight loss of over 15 kilograms present over the past 6 months associated with loss of appetite. Patient also gives history of tinnitus in the right ear associated with hearing loss. History of poly-arthralgia present predominantly involving the bilateral inter-phalangeal joints and knee joints. There is no history of skin rashes, seizures, decreased urine output, chest pain palpitations, or dyspnea. The patient has no other co-morbidities. On examination, the patient was conscious, oriented, febrile with a temperature of 99.8ºF, and other vitals were stable. Systemic examination revealed no significant abnormalities. Complete blood count showed neutrophilia and decreased RBC count. ESR was raised to 82 mm, and CRP was 1.2 mg/dl. LFT, RFT, Serum electrolytes, and urine routine were found to be expected. ANA (IFA) was positive.

Complement C_3 and C_4 levels were low (60 mg/dl and 9.1 mg/dl, respectively). Serum LDH levels were raised to 305 U/L. Anti-dsDNA level was 60 IU/ml. Anticardiolipin antibodies and lupus anticoagulant was 0.8 (Negative). D-Dimer was elevated to 1.18 mcg/ml. MRI brain showed diffuse and multiple punctate T2 FLAIR hyperintensities in the bilateral brain parenchyma, suggestive of vasculitis. Hence a diagnosis of CNS vasculitis was made and the patient was started on T. Mycophenolate Mofetil 500mg twice daily, T. Hydroxychloroquine 200 mg HS and T. Methylprednisolone 8mg OD. The patient's condition improved, and her symptoms subsided completely within two weeks of commencing treatment.

Table 1. Different parameters of blood biochemistry

Investigations	**Values**
ESR	82 mm
CRP	1.2 mg/dl
LDH	305 u/l
D-DIMER	1.18
ANA (IFA)	Positive
LUPUS ANTICOAGULANT (DRVVT METHOD)	0.8 (negative)
ANTICARDIOLIPIN ANTIBODIES: IGM:	Negative

IGG	3 u/ml 3.3 u/ml
Complement C3	60 mg/dl
Complement C4	9.1 mg/dl
ANTI-DSDNA	60 iu/ml

TABLE-1 showing summary of investigations

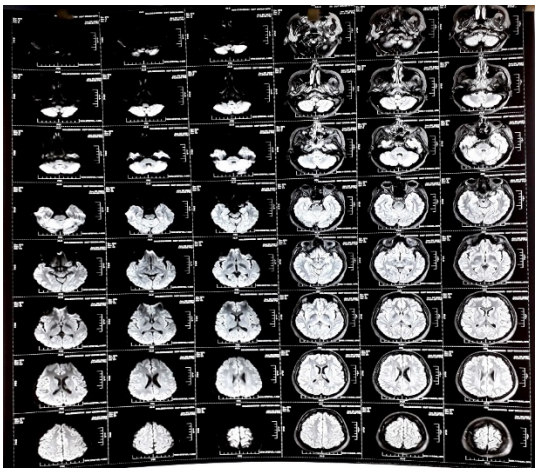

FIG -1 showing MRI brain having diffuse and multiple punctate T2 FLAIR hyperintensities in bilateral brain parenchyma.

Discussion

True CNS vasculitis is relatively rare and occurs in only 7% of patients with SLE. It usually presents with recurrent bouts of fever, severe headache, and acute confusional states, which can progress rapidly towards psychosis, seizures and coma (Everett *et al.,* 2008). Underlying active Lupus is usually demonstrable with lab investigations revealing hypocomplementemia and elevated anti-dsDNA levels (Rowshani *et al.,* 2005). MRI brain is usually abnormal showing focal defects. EEG, CSF analysis and SPECT scan can reveal the extent of neurological dysfunction if performed. It is important to differentiate between CNS vasculopathy and true vasculitis of the brain to frame treatment modalities (Kakati *et al.,* 2017). Initial aggressive treatment with IV Cyclophosphamide and IV methylprednisolone if the disease progresses rapidly. Azathioprine and Mycophenolate Mofetil have been used as second-line agents in case cyclophosphamide toxicity and have been equally effective. Anti-CD20 antibody drug-like Rituximab have also been considered effective in remission of the disease.

Conclusion

Active treatment of SLE is required to prevent the progression of systemic involvement. In addition, prompt clinical assessment and imaging studies are required to diagnose neurological complications of SLE, and aggressive treatment with immunosuppressants is needed to bring about remission.

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Study significance:

Active treatment of SLE is required to prevent the progression of systemic involvement.

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