A Rare Presentation of Brain Stem Involvement (Encephalomyelitis) In Systemic Lupus Erythematosus

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Abstract- **Brainstem involvement in systemic lupus erythematosus is an uncommon but serious manifestation which present with various neurological symptoms that includes cranial nerve deficits such as ophthalmoplegia, facial numbness, diplopia, ataxia, vertigo, dysarthria, hemiparesis and altered consciousness. We report a rare case of 20-year-old female who is a known case of SLE with brainstem involvement.**

I. INTRODUCTION

SYSTEMIC LUPUS ERYTHEMATOSUS, tissue damage begins with deposition of auto antibodies and/or immune complexes followed by destruction mediated by complement activation and release of cytokines/chemokines.
Prevalence of SLE in India range from 14 to 16 per 100000 people

II. CASE REPORT

A 20-year-old female patient, resident of penugonda, k/c/o SLE with AIHA since four years on tab.wysolone 5mg informant being father came with chief complaints of :

- Fever since 5 days
- Vomitings(Projectile) since 2 days
- Headache since 2 days

Then she developed vertigo and diplopia on arrival to the hospital and on further evaluation she had nystagmus, restricted movements of left eye on lateral side(suggestive of sixth nerve involvement), ptosis and miosis of left eye(suggestive of Horner's syndrome), positive Romberg's sign, truncal ataxia, gait ataxia, stance ataxia, titubation which was having on and off pattern and high grade fevers even with antibiotics suggestive of autoimmune pattern and mild elevation in dsDNA titres(1.89,ref <o.8) and complements on lower side suggesting active CNS lupus, patient was given methyl prednisolone pulse doses and rituximab . There was_significant clinical improvement (fever decreased, improvement in other signs).

III. INVESTIGATIONS

- Anti ds DNA- 1.89 (ref range: <0.8)
- Complements - c3- 63.4(ref range:90-180 mg/dl)
- c4- 7.5(ref range:10-40mg/dl)
- CSF culture - sterile
- CSF zn staining - negative
- CRP – negative

- MRI BRAIN- defined t2 and flair hyperintensity with no restriction on DWI and no significant post contrast enhancement in left half of medulla extending into cervico medullary junction -? encephalitis

- MRI C SPINE: t2 hyperintense signal in visualized brain stem and cervical spinal cord with cord expansion - likely acute disseminated encephalomyelitis.

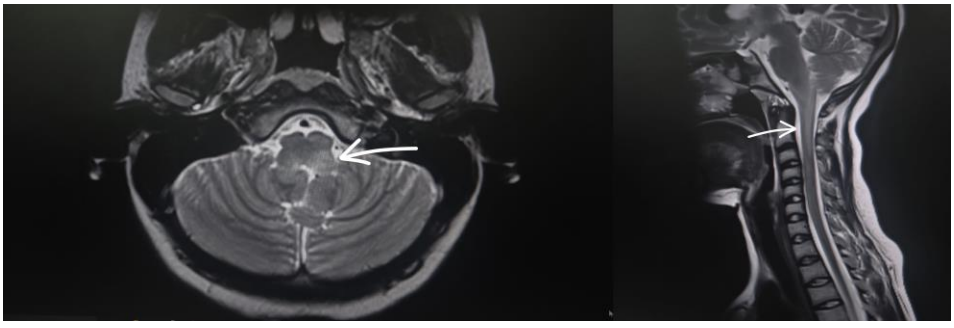

Fig. MRI of Brain and C Spine.

IV. RESULTS

Based on clinical presentation and investigative findings, patient was diagnosed with brainstem involvement in SLE flareup.

V. MANAGEMENT AND OUTCOME

The patient was initiated on pulse dose steroids and then patient was given Rituximab then advised further doses of Rituximab and patient symptoms were subsided.

VI. DISCUSSION

The reported incidence of CNS in SLE varies across studies, estimates ranging from approximately 40 to 70% and patients with brain stem involvement may exhibit symptoms including cranial nerve deficits (e.g.: diplopia due to sixth nerve palsy), ataxia, dysarthria, nystagmus that results from inflammatory, ischemic and antibody mediated processes affecting brain stem.
Diagnosis: MRI BRAIN, CSF analysis, ANA profile, Complement.
Treatment: Pulse dose corticosteroids (Methyl prednisolone), Immunosuppressive agents.

VII. CONCLUSION

Brainstem involvement in systemic lupus erythematosus is a rare but potentially life-threatening manifestation that requires prompt recognition and aggressive immunosuppressive therapy.
Early diagnosis, supported by neuroimaging and serologic markers, combined with timely treatment, can significantly improve neurological outcomes.

CONSENT

The consent was taken and signed by the respective patient.

CONFLICTS OF INTEREST

The authors have no conflicts of interest.

REFERENCES

[1] Kumar, Susheel, et al. "A case of systemic lupus erythematosus with extensive brain stem involvement." Clinical rheumatology 28 (2009): 69-71.
[2] Shibata, Machiko, et al. "Diffuse central nervous system lupus involving white matter, basal ganglia, thalami and brainstem." Brain and development 21.5 (1999): 337-340.
[3] Ameer, Muhammad Atif, et al. "An overview of systemic lupus erythematosus (SLE) pathogenesis, classification, and management." Cureus 14.10 (2022).
[4] Firestein and Kelley’s textbook of Rheumatology, 12th edition
[5] Harrison’s Principles of Internal Medicine, 21st edition