AN UNUSUAL CASE OF SUBACUTE SCLEROSING PANENCEPHALITIS PRESENTING AS MYOCLONUS AND CERVICAL DYSTONIA

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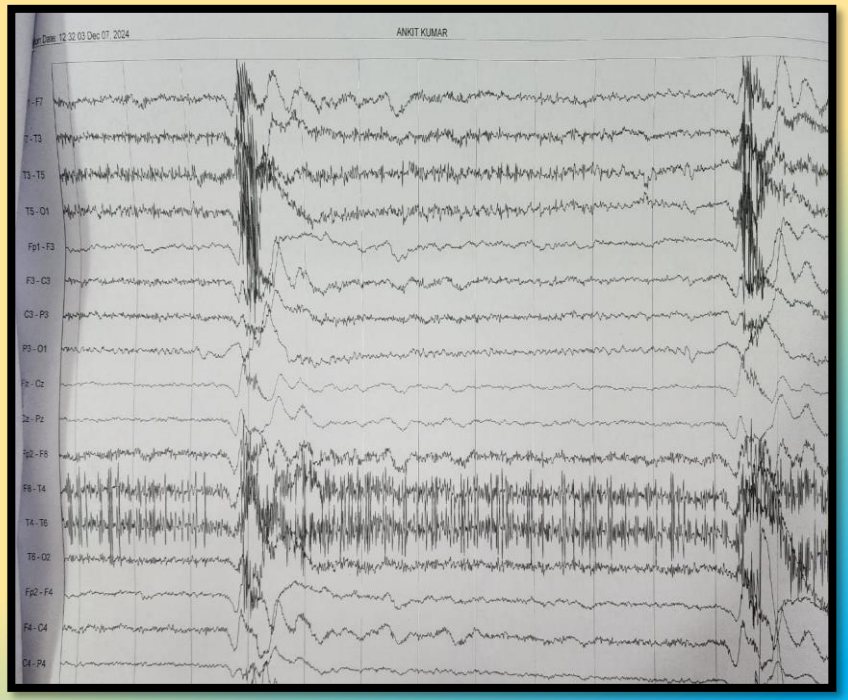
INTRODUCTION

- Subacute sclerosing panencephalitis (SSPE) is characterized by progressive deterioration of cognitive and motor function and death within 1-3 years.
- It often develops in a person who had measles at an age of less than 2 years.
- It is diagnosed with Dyken's criteria including clinical features, high titre anti measles IgG in serum and CSF and electroencephalogram pattern.
- A very few adult onset SSPE cases have been reported in literature.

CASE REPORT

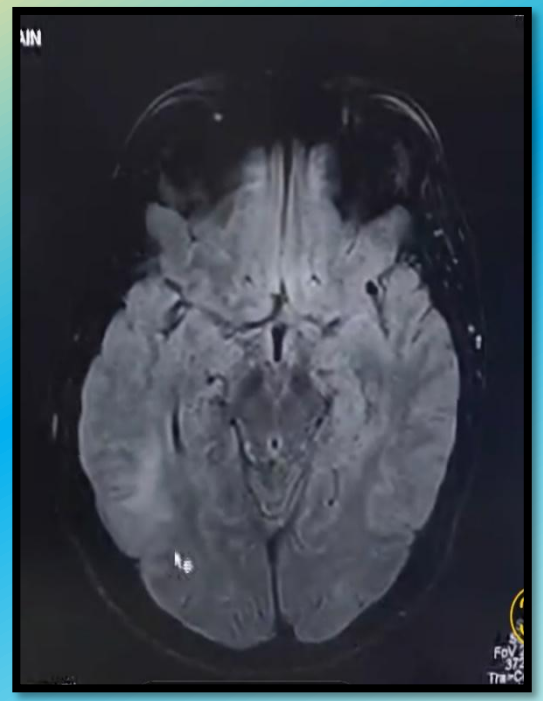
- A 17 year old male with normal birth and developmental history and immunization schedule presented with history of poor scholastic performance and abnormal involuntary movements since 1 year.
- On clinical examination, patient had cognitive impairment, generalized spontaneous and stimulus sensitive myoclonus. He also had cervical dystonia with no sensory, autonomic and bowel/bladder involvement.
- His workup for Wilsons disease and progressive myoclonic epilepsy was non-contributory.

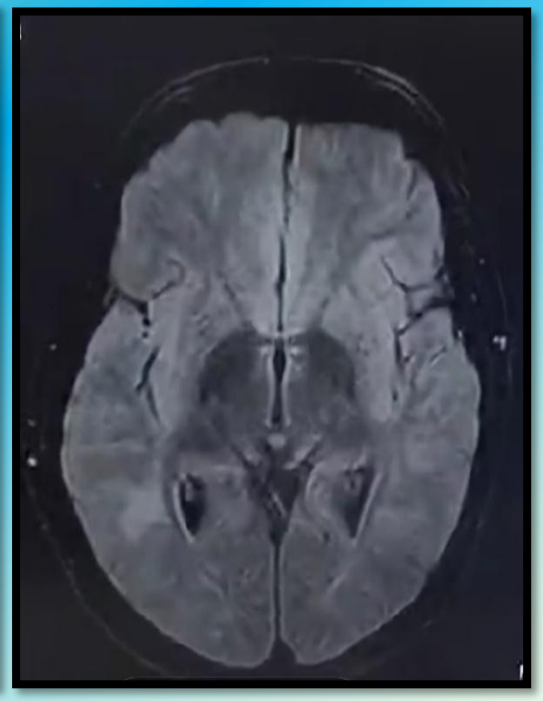
INVESTIGATIONS	
CBC, LFT, KFT, Urine routine	normal
Serum ceruloplasmin	382 mg/L
Serum IgG measles CSF IgG measles	473.64 U/mL 617.57 U/mL
CSF/serum quotient for measles antibody	Positive (5.16)

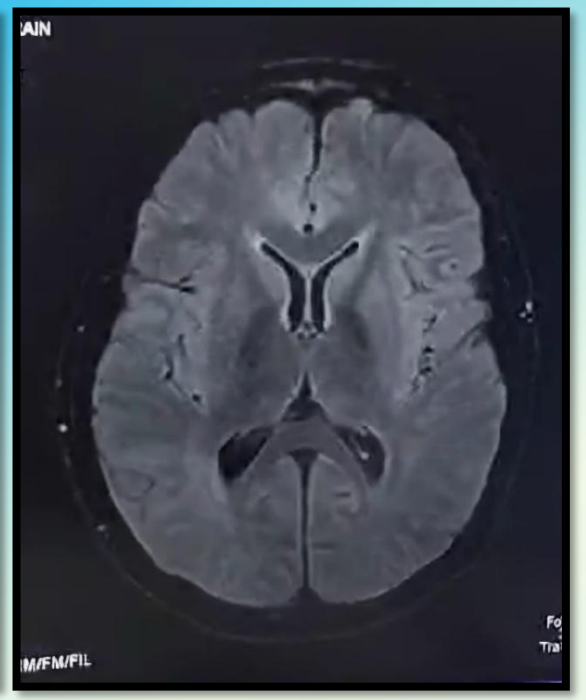


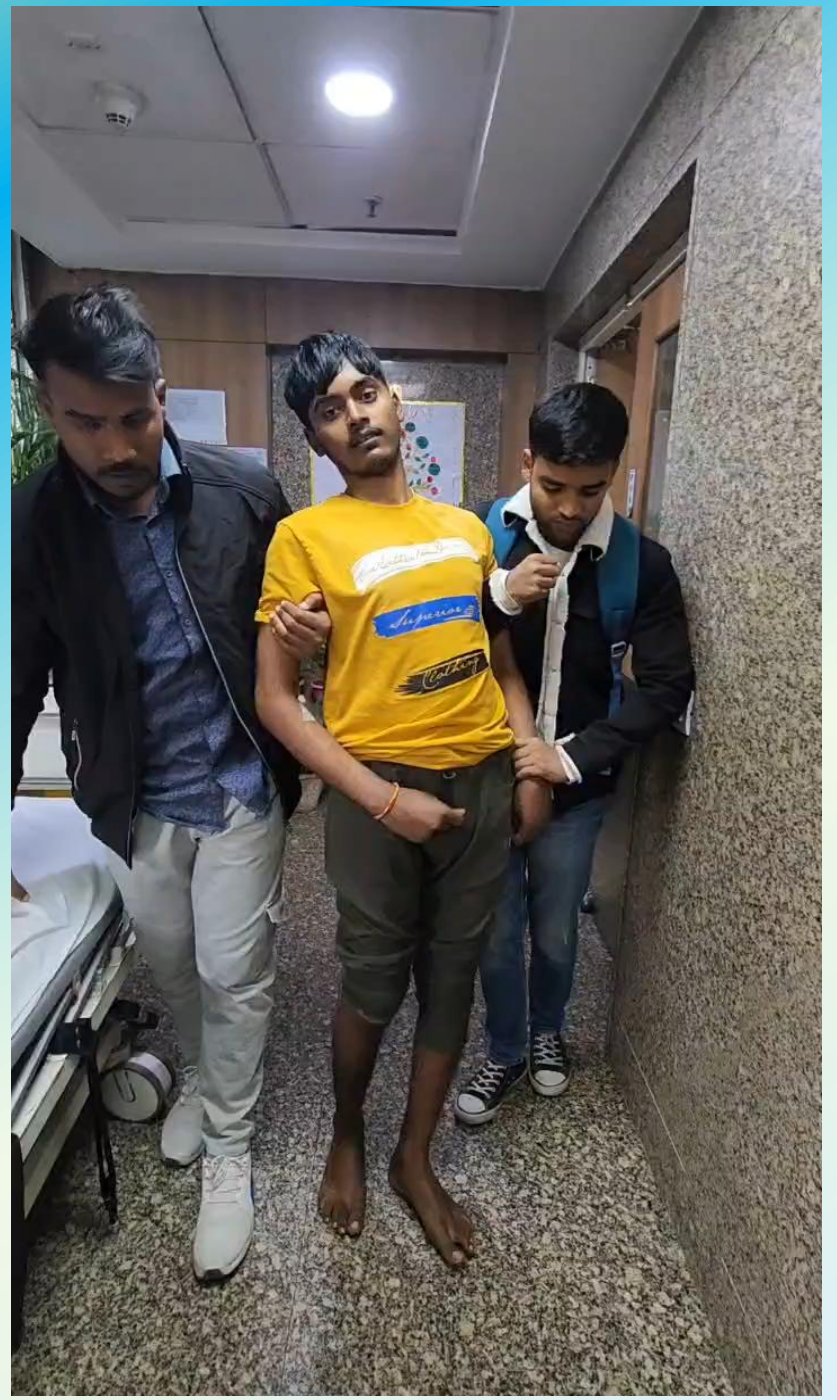
His EEG showed periodic long interval diffuse discharges (Radermecker complexes) occurring at 5-20 seconds interval, time locked with myoclonic discharges.

His MRI brain revealed hyperintense signal in bilateral insular cortex, occipito-temporal lobes and subcortical left high frontal lobe.









DISCUSSION

- He had history of repeated respiratory tract infections before 2 years of age.
- Due to the presence of high titre CSF anti-measles antibodies, Radermecker complexes and brain imaging findings, a reasonable diagnosis of SSPE was considered.
- We started treatment with valproate, clonazepam, levetiracetam and trihexyphenidyl.

CONCLUSION

- SSPE usually results from a measles virus infection acquired earlier in life, which usually develops 7 to 10 years later.
- It is a chronic progressive neurodegenerative condition resulting in akinetic mutism with persistent vegetative stage.

REFERENCES

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2. Garg D, Kakkar V, Kumar A, Kapoor D, Abbey P, Pemde H, Mukherjee SB, Sharma S. Spectrum of Movement Disorders Among Children With Subacute Sclerosing Panencephalitis: A Cross-Sectional Study. J Child Neurol. 2022 May;37(6):491-496.