Rapidly Progressive Subacute Sclerosing Panencephalitis

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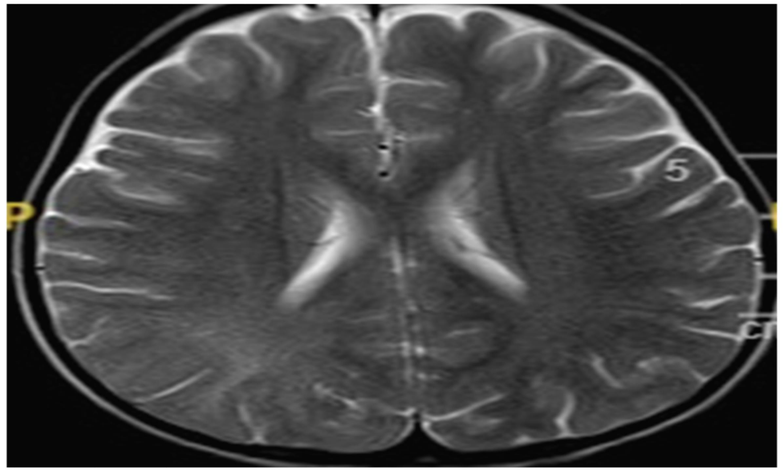
Introduction

Subacute sclerosing panencephalitis (SSPE) is a slowly progressive neurodegenerative disease associated with a complication of measles. The initial clinical features of SSPE in early childhood differ from those in adults.

Aim

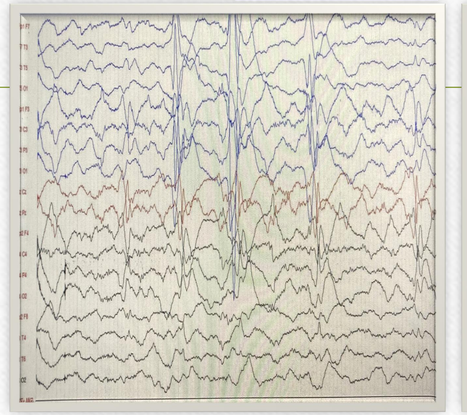
We discuss a 4-year-old male patient who started with drop attacks, showed rapid deterioration within 1month, and was diagnosed with SSPE.



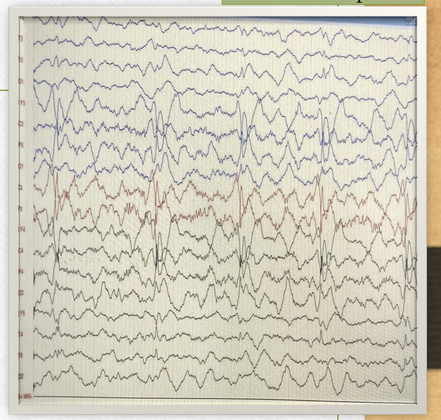
Case

A 4.5-year-old male was brought to hospital due to seizure. It was learned that the patient was previously healthy, generalized myoclonic jerks started after the fall attacks, followed a rapidly progressive course, and lost all developmental milestones within 1 month. He was born from a non-consanguineous family. The pregnancy, perinatal, neonatal, developmental histories were unremarkable. He developed clinically diagnosed measles at 1 years of age and did not have vaccinations. In neurological evaluation, he was in the vegetative stage and deep tendon reflexes were increased along with extensor plantar responses. He had generalized repetitive myoclonic jerks during the waking period and were stimulus sensitive. Magnetic resonance imaging (MRI) showed high signal intensity on T2 and FLAİR-MRI bilateral periventricular white matter. Electroencephalogram showed bilateral, synchronous high-amplitude slow waves at regular intervals, no suppression was observed after diazepam. The measles-specific IgG index obtained from the cerebrospinal fluid (CSF) was 35.56 u/ml and a diagnosis of SSPE was made.

Before diazepam



After diazepam



Conclusion

Clinically, SSPE has a stereotypical four-stage protracted course (1). But rarely
It has an acute and severe course leading to serious neurological disability and death occurs within a few weeks. The earlier the age of measles infection, the higher the risk of developing SSPE (2,3). Data on SSPE in preschool age is limited, and despite the slow progressive course of SSPE, our case is important in that it follows a very rapid clinical course.

References

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