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Case Report

SSPE with Ocular Manifestation "Eye as Window to the Brain"

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Introduction

Subacute Sclerosing Panencephalitis (SSPE), is a progressive neurodegenerative disease caused by the persistence of wild measles infection commonly seen in children and young adults [1-4] The estimated risk is 4–11/100,000 cases worldwide [5], while developing countries like India reported an incidence rate is 21 cases per million [6]. The reported risk factors such as younger age of measles (16 times greater risk), poverty, rural area, overcrowding, higher birth order, and higher number of siblings [7]. The clinical presentation of SSPE is variable and characterized by progressive cognitive decline, periodic slow myoclonus, and extrapyramidal motor dysfunction with ocular manifestations. The classic age at presentation is 8–11 years. Herein, we report a case of SSPE with bilateral papilledema, emphasizing the role of ocular examination as a tool in early diagnosis. CASE : A 6 year-old boy, born to a non-consanguineous couple, antenatal period being uneventful , delivered by a normal vaginal delivery with the postnatal period being uneventful. The child attained developmental milestones age appropriate. Immunization history reveals that he is a defaulter, received only vaccination at birth.

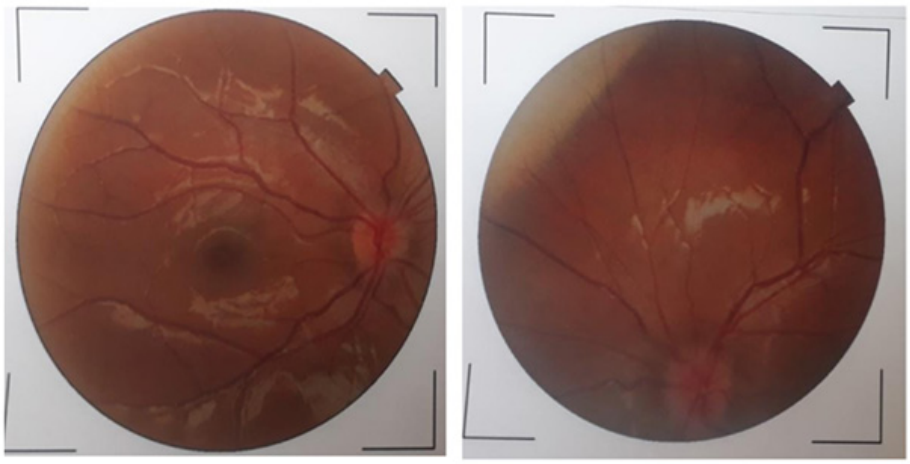

Figure 1: Fundus photography suggestive of OD (Right Eye) and OS (Left Eye) Papilledema.

Six months prior to admission to the hospital, the child demonstrated personality changes consisting of outbursts of anger, decreased interest in playing outdoor, and deterioration in school performance. He was presented to us with a 2 month history of continuous involuntary jerky movements of both upper limbs and trunk. Multiple episodes of generalized seizures heralded the onset of illness, with drop attacks, following which he had a progressive decline in cognitive function. At presentation to our hospital, he was in a minimally conscious state but able to recognize parents unable to hold the neck, sit or stand without support. On examination, apart from altered sensorium, he had expressive aphasia(mute) and central hypotonia and he was getting repetitive axial myoclonic jerks. On Ophthalmological evaluation, his fundus showed bilateral papilloedema correlating well with fundus photography (Figure 1).

On investigation, MRI is suggestive of focal T2-flair hyperintensities in bilateral corona radiata and centrum semiovale. EEG revealed periodic generalized complexes consisting of bilaterally symmetrical, high voltage (>200μV) bursts of sharp waves and delta waves which repeat at an interval of 3 to 15 seconds intervals with a slow background (Figure 2).

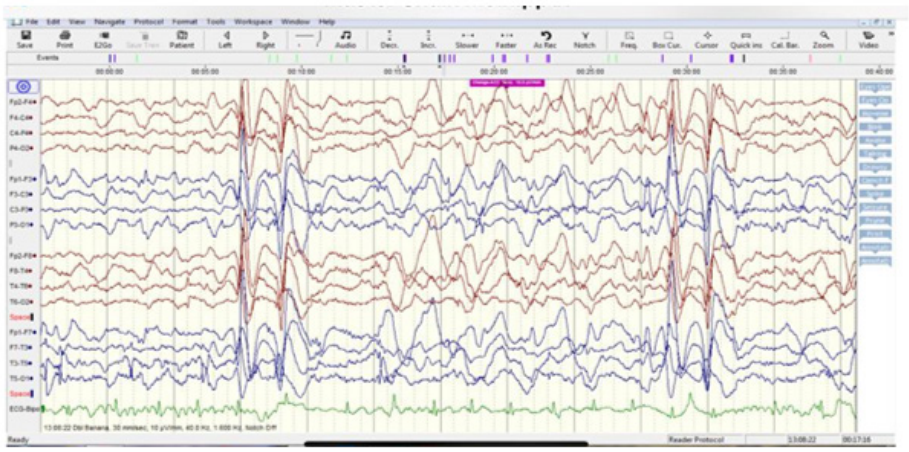

Figure 2: Generalized periodic epileptiform discharge with slow background.

The diagnosis of SSPE was confirmed with elevated titers of cerebrospinal fluid (CSF) anti-measles IgG antibody levels (3.74mg/dl) and serum anti-measles IgG antibody levels (4814). CSF/serum quotient (CSQrel) was positive (4.27). Cerebrospinal fluid was clear with 4 cells (all lymphocytes), CSF protein 16 mg/dL, and glucose 72 mg/dL. Complete blood count, serum electrolytes, liver, and kidney function test, ESR, Chest X-ray were normal. The child was initiated on Isoprinosine, amantadine, and antiepileptic drugs (valproic acid and clonazepam).

Discussion and Conclusion

Diagnosis of SSPE can be very challenging, particularly in atypical cases when myoclonus is absent or subtle. Another study by Prashant et al. revealed that the rate of misdiagnosis was as high as 78.8%, with the interval between accurate diagnosis and presentation being 6.2 ± 11.3 months (range 0.2–96 months) [11]. Visual involvement can be there in as many as 50% of patients [9]. The most common ocular manifestations include papilloedema and nystagmus followed by optic neuritis, ptosis, optic atrophy and macular pigmentary clumps [11]. Even though, chorioretinal involvement being common, optic nerve involvement can be seen in significant number of cases [12]. Ocular involvement can sometimes precede neurological symptoms by months or even years [9,10]. Hence, ocular examination in young children with SSPE is an important clinical handle for the neurologist.

Acknowledgement

None.

Conflict of Interest

No Conflict of interest.

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