

## Case Report

### Subacute Sclerosing Panencephalitis

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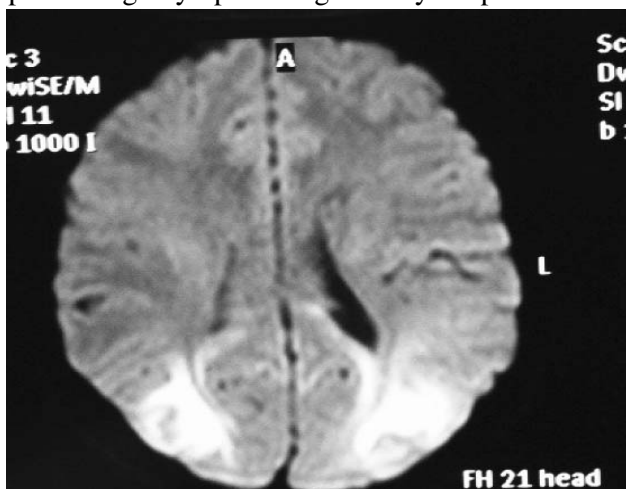
#### Abstract

Subacute sclerosing panencephalitis (SSPE) is a rare, invariably fatal degenerative disease of central nervous system developing after measles infection. Neurological symptoms are initial presenting symptoms but rare reports of pure psychiatric symptoms preceding neurological symptoms have been reported. We here report a case of 18 years old male patient suffering from Subacute sclerosing panencephalitis (SSPE) who initially had behavioural changes and then subsequently developed neurological symptoms.

**Keywords:** Subacute sclerosing panencephalitis, measles, psychiatric symptoms

#### Introduction

Subacute sclerosing panencephalitis (SSPE) is a rare and serious disorder of the central nervous system. It is a slow virus infection caused by defective measles virus. The term subacute sclerosing panencephalitis has been used since Greenfield suggested it in 1960 to designate a condition due to a persistent infection by a virus involving both grey matter and whitematter<sup>1</sup>. Initial presenting symptoms generally reported are



*Fig.1 MRI scan showing confluent hyperintensities in bilateral parieto occipital regions*

myoclonic jerks, falling attacks, changing gait, abnormal movements, speech impairment, inability

to walk or stand, seizures, dementia, visual disturbance, pyramidal and extra pyramidal signs.<sup>2</sup>We report a case of 18 yrs old male who presented with behavioural changes and was subsequently diagnosed as case of SSPE.

#### Case report

An 18 years old boy was referred to the department of medicine on October 16, 2012 because of recent behavioural changes with progressive cognitive impairment, disorientation for last 11 months and involuntary movements, uncontrolled defecation and micturition over the past 9 months. Medical history was unremarkable except for an episode of measles 12 months back. No history of encephalitis was reported. Before hospital admission patient's mother noticed memory problems, confusion, apathy and an inability to stay alone without supervision. She observed nocturnal myoclonic jerks after two months of illness.

Results of the initial mental status examination revealed poor attention and concentration, disorientation, perseveration, acalculia and ideational apraxia. Findings from basic neurological examination showed bilateral myoclonic jerks with hyperreflexia and bilateral Babinski sign. Ophthalmic examination was unremarkable.

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## Subacute Sclerosing Panencephalitis

Extensive medical and laboratory work up was done to rule out other diseases, the result of these tests showed no abnormalities. The electroencephalogram showed periodic sharp and slow wave discharges. The MRI imaging scan showed confluent hyperintensities in bilateral parieto occipital regions with few discrete white matter hyperintensities in bilateral fronto parietal regions(Fig.1). which are the findings of sequele of viral encephalitis (possibility of subacute sclerosing panencephalitis) . Serum measles IgG titer was done which turned out to be positive and confirmed the diagnosis.

### Discussion

Our patient fulfilled the criteria for SSPE, which include fulfillment of at least three of the following five criteria<sup>3</sup>

A typical clinical picture: Personality and behavioral changes, worsening school performance, followed by myoclonic seizures, paresis, dyspraxias,

memory impairment, language difficulties, blindness, and eventually obtundation, stupor, and coma  
Characteristic EEG changes

Elevated CSF globulin levels greater than 20% of total CSF protein

Raised titers of measles antibodies in blood and CSF

Typical histopathological finding in brain biopsy or autopsy

Presentations with psychiatric symptoms have been described in the literature. Cases of schizophreniform psychosis, paranoid psychosis, schizophrenia catatonia and depression are well described.<sup>4,8</sup> Since the disease is slowly progressive occurring years after measles infection, it is possible that full neurological manifestations may develop only after some period of time.

In conclusion high suspicion is needed to detect SSPE because of its atypical presentation and rare form. Detailed work up to rule out organic conditions is required.

### References

1. Greenfield JG. Encephalitis and encephalomyelitis in England and Wales during last decade. *Brain* 1950;**73**:141-66. <http://dx.doi.org/10.1093/brain/73.2.141> PMID:14791785
2. Prashanth LK, Taly AB, Sinha S, Ravi V. Subacute sclerosing panencephalitis (SSPE): An insight into the diagnostic errors from a tertiary care university hospital. *J Child Neurol* 2007; **22**: 683-8. <http://dx.doi.org/10.1177/0883073807303999> PMID:17641252
3. Dyken PR. Subacute sclerosing panencephalitis. *Neurol Clin* 1985; **3**: 179-85. PMID:2581121
4. Forrest G, Stores G. Subacute sclerosing panencephalitis presenting with psychosis and possible sexual abuse. *Eur Child Adolesc Psychiatry* 1996; **5**: 110-3. <http://dx.doi.org/10.1007/BF01989503>
5. Duncalf CM, Kent JN, Harbord M, Hicks EP. Subacute sclerosing panencephalitis presenting as schizophreniform psychosis. *Br J Psychiatry* 1989; **155**: 557-9. <http://dx.doi.org/10.1192/bjp.155.4.557> PMID:2611581
6. Jahnel M. Paranoid-hallucinatory psychosis as primary manifestation of subacute sclerosing panencephalitis (SSPE) in a 19-year-old man. *Psychiatr Prax* 2003; **30**: S70-2. PMID:14509042
7. Khadilkar SV, Patil SG, Kulkarni KS. A study of SSPE: Early clinical features. *J pediatric neurology* 2004; **2**: 73-7.
8. Khwaja GA, Gupta M, Sharma DK. Subacute sclerosing panencephalitis. *J Assoc Physicians India* 1991; **39**: 928-33. PMID:1816221