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Clinical Spectrum and Electroencephalographic Correlation in Subacute Sclerosing Panencephalitis: A Case Series

Dr. Sansmon Thomas[1], Dr. N Veena[2], Dr. G Sree Ranga Lakshmi[3]

[1, 2, 3]Department of Neurology, Osmania Medical College, Hyderabad, India

Abstract: ***Background: Subacute Sclerosing Panencephalitis (SSPE) is a fatal, progressive neurodegenerative disorder caused by persistent, aberrant measles virus infection in the central nervous system. Characterized by cognitive deterioration, behavioral abnormalities, progressive motor deficit, and stereotyped slow myoclonic jerks, its clinical hallmarks are intricately linked to classic electroencephalographic (EEG) periodic slow-wave complexes (Radermecker complexes). Case Presentation: We report a series of four distinct clinical presentations of SSPE Case 1 describes a 20-year-old male with initial focal myoclonic seizures, asymmetric hemiparesis, cognitive decline, subsequent progression to generalized myoclonus, and typical generalized Radermecker complexes post-intrathecal interferon-alpha therapy. Case 2 illustrates an unimmunized 10-year-old boy presenting with classic scholastic failure, prominent axial and proximal appendicular slow myoclonus, gait impairment, and stereotypic high-voltage periodic slow complexes. Case 3 details an 18-year-old female with a confounding history of hydrocephalus requiring surgical decompression, presenting with scholastic decline, periodic slow-wave discharges, and slow myoclonus. Case 4 outlines an atypical, protracted decade-long course in a 20-year-old male misdiagnosed with primary psychiatric illness and movement disorders before presenting with severe cognitive failure, gait cessation, and classic periodic discharges. Discussion & Conclusion: These cases demonstrate the broad phenotypic spectrum of SSPE—spanning classic juvenile presentation, focal onset, neurosurgical confounders, and protracted psychiatric-extrapyramidal masquerades. Electroencephalography remains the pivotal real-time neurophysiological biomarker. Early clinico-electrophysiological suspicion is mandatory to avoid catastrophic diagnostic delays.***

Keywords: Subacute Sclerosing Panencephalitis, SSPE, Radermecker complexes, Periodic slow-wave complexes, Slow myoclonus, Measles virus, Clinico-electrophysiological correlation, Jabbour staging

1. Introduction & Review of Literature

Subacute Sclerosing Panencephalitis (SSPE), originally described by James Dawson in 1933 as 'inclusion body encephalitis' and subsequently delineated electrophysiologically by Joseph Radermecker in 1949, is a relentless, progressive neurodegenerative disorder of children and young adults. It is caused by persistent infection of the central nervous system (CNS) by hypermutated measles virus (MeV) strains. SSPE persists in endemic pockets, particularly in developing regions where childhood measles outbreaks occur and vaccination coverage remains suboptimal.

Clinically, SSPE is categorized into four progressive stages according to Jabbour's staging system:

- Stage I (Incipient / Mental Stage): Insidious behavioral alterations, personality changes, emotional lability, subtle scholastic decline, and cognitive slowing.
- Stage II (Convulsive / Myoclonic Stage): Hallmark periodic slow myoclonic jerks ,extrapyramidal movement disorders, chorioretinitis, and progressive intellectual deterioration.
- Stage III (Comatose / Rigidity Stage): Decerebrate or decorticate rigidity, extrapyramidal dystonia, autonomic instability, stupor, and unresponsiveness.
- Stage IV (Terminal / Vegetative Stage): Quadriparesis, mutism, loss of cortical function, flexor spasms, autonomic crisis, respiratory failure, and death.

Diagnostic confirmation rests upon Dyken's revised criteria which encompass elevated cerebrospinal fluid (CSF) and serum anti-measles IgG antibodies, typical periodic complexes on EEG, elevated CSF immunoglobulin ratio, and compatible clinical findings.

Electroencephalography plays a paramount role in both early diagnosis and longitudinal assessment of SSPE. The pathognomonic EEG hallmark consists of 'Radermecker complexes'—bilaterally symmetrical, synchronous, high-voltage (200–1000 µV), stereotyped polymorphic delta waves or sharp-and-slow wave discharges recurring regularly every 4 to 15 seconds against a progressively disorganized and suppressed background. Crucially, these periodic complexes correlate precisely with visible slow myoclonic jerks and clinical lapses in postural tone.

2. Case 1

A 20-year-old right-handed male presented to the neurology service in a bedridden, profoundly cognitively impaired state with frequent generalized jerks. His illness had commenced at 17 years of age with insidious myoclonic jerks preferentially involving the left upper and lower extremities. Initial evaluation had revealed background slowing and diagnostic lumbar puncture confirmed markedly elevated CSF anti-measles IgG antibody titers, establishing the diagnosis of SSPE (Jabbour Stage II).

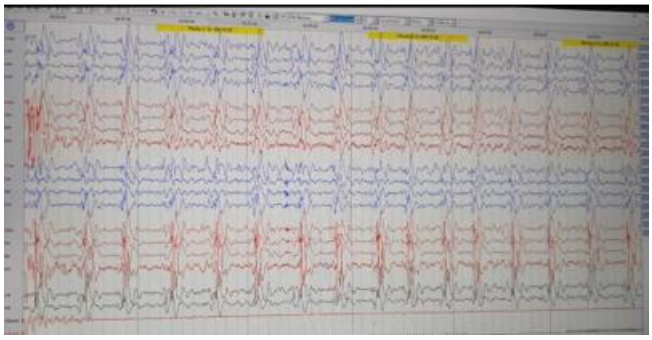
Following diagnosis, the patient was initiated on intraventricular/intrathecal Interferon-alpha (IFN-α) therapy, receiving three sequential doses alongside oral antiepileptic therapy (valproate and clonazepam). Despite therapeutic intervention, he had progressive neurocognitive deterioration over the next 36 months. He developed a left hemiparesis,

bradyphrenia, loss of speech output, severe scholastic and cognitive decline, eventually becoming bedridden.

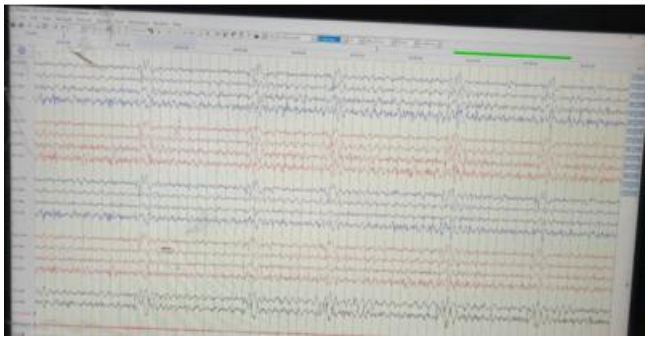
Neurological Examination
Cranial nerve examination demonstrated preserved pupillary light reflexes, roving horizontal eye movements. Motor examination revealed spastic left hemiparesis with hypertonia bilateral ankle clonus with bilateral extensor plantars. Generalized, stereotyped, slow myoclonic jerks occurred rhythmically every 5–8 seconds, involving the axial trunk and all four extremities, predominantly affecting the right side.

Electrophysiological & Neuroimaging Correlation
Current electroencephalography (EEG) demonstrated marked diffuse suppression and severe slowing of the background rhythms with theta-delta dominance. Superimposed generalized, bilaterally synchronous, stereotypic periodic slow-wave complexes (Radermecker complexes) recurring at intervals of 6 to 9 seconds, each lasting 1.2 to 2.0 seconds with amplitudes exceeding 450 µV, were seen,showing synchronization with the clinical slow myoclonic jerks. Brain MRI revealed progressive bilateral subcortical white matter hyperintensities on T2/FLAIR images with prominent cerebral atrophy and secondary ex-vacuo ventricular dilatation, corresponding to advanced Jabbour Stage III disease.

Case 1 EEG



Case 2 EEG



3. Case 2

A 10-year-old boy was brought by his parents with a 1 year history of rapidly declining scholastic performance, cognitive slowing, behavioral regression, and abnormal involuntary jerking movements. The child was unimmunized during early infancy and had suffered a severe, febrile exanthematous illness clinically diagnosed as measles at approximately 14 months of age. Over the preceding four months, his teachers reported inability to comprehend instructions, loss of handwriting skills, and frequent drop attacks.

Involuntary movements had emerged insidiously as slow, rhythmic, shock-like jerks primarily affecting his neck (resulting in periodic head-nodding/head-drop spells), as well as bilateral proximal shoulder girdle and pelvic musculature. The jerks resulted in progressive gait unsteadiness, frequent stumbles, and eventual inability to walk unassisted.

Neurological Examination
On examination, the child was conscious but severely abulic, disoriented, and demonstrated marked memory impairment. Cranial nerves were intact without optic atrophy or chorioretinitis. Motor system assessment revealed generalized hypotonia distally with preserved muscle bulk. Periodic slow myoclonic jerks were observed every 4 to 6 seconds, producing synchronous neck flexion, shoulder abduction, and brief hip flexion, followed by a brief post-myoclonic atonic phase, accounting for his gait collapse.

Electrophysiological Correlation
Although parental consent for lumbar puncture could not be obtained due to procedural reluctance, video-EEG provided definitive electrophysiological evidence. The interictal background showed diffuse delta slowing (2–3 Hz) with absence of normal posterior alpha rhythm. Highly rhythmic, stereotyped, generalized high-voltage periodic discharges (amplitude 350–650 µV, duration 1.5–2.5 seconds) occurred every 4 to 6 seconds across all montages (Radermecker complexes). Each periodic discharge was precisely coincident with the slow axial and proximal limb myoclonic jerks, establishing the diagnosis of classic Stage II SSPE.

4. Case 3

An 18-year-old female presented with progressive cognitive decline, behavioral changes, and involuntary rhythmic body jerking over a 9-month period. Her past medical history was notable for early childhood hydrocephalus diagnosed at 6 years of age, for which she had undergone a surgical decompressive procedure (ventriculoperitoneal shunt placement followed by subsequent revision). She had maintained normal neurodevelopmental milestones and completed secondary schooling successfully until the current illness.

The initial symptoms were attributed to shunt malfunction and scholastic fatigue; however, repeated neurosurgical and radiological evaluations confirmed normal functioning of the shunt.Over the subsequent months, she developed marked slowing of mentation, dyspraxia, loss of calculation abilities, and frequent shock-like muscle twitches.

Neurological Examination
Neurological evaluation revealed significant frontal-subcortical cognitive impairment, bradykinesia, and dysarthria. Fundus examinationwas normal. Motor examination revealed mild generalized spasticity with brisk deep tendon reflexes and bilateral extensor plantars. Slow, stereotypic myoclonic jerks involved the axial musculature, facial muscles, and bilateral upper extremities at 6–8 second intervals.

Electrophysiological Correlation
Lumbar puncture for CSF measles antibodies was deferred by the family. Digital EEG revealed a severely disrupted background rhythm with continuous polymorphic slow waves and recurring, generalized, bi-frontally predominant periodic sharp-and-slow wave complexes (Radermecker complexes) recurring every 6 to 8 seconds.

5. Case 4

A 20-year-old male presented with advanced cognitive decline, loss of independent ambulation, and involuntary movements with the course spanning nearly 8 to 10 years. In early adolescence he developed severe behavioral abnormalities, personality disintegration, emotional outbursts, and aggressive episodes. He was managed as a case of juvenile psychiatric illness.

Over the ensuing years, subtle movement disorders emerged, manifesting as postural tremors, choreoathetotic movements, later progressed insidiously into severe cognitive regression, speech arrest, and severe gait apraxia, leaving him non-ambulant and completely dependent for all activities of daily living.

Neurological Examination
Examination showed an uncommunicative, bedridden young male exhibiting profound dementia, lack of motor responses, and mutism, with severe extrapyramidal rigidity with cogwheeling, dystonic posturing. and hyperactive deep tendon reflexes. Slow, occasional, long-interval myoclonic jerks were observed, involving the head, trunk, and proximal extremities.

Electrophysiological Correlation
Digital EEG recording revealed profound diffuse cerebral dysfunction with complete absence of background alpha organization. Highly typical, high-amplitude (500–800 μV) periodic sharp-and-slow wave Radermecker complexes were observed occurring at lengthened intervals of 8 to 12 seconds, typical of prolonged, late-stage disease. While CSF measles antibody testing was not performed due to the advanced state, the clinicoradiological evolution and pathognomonic EEG periodic complexes confirmed SSPE.

6. Discussion

The present series underscores the rich phenotypic variability, diagnostic pitfalls, and electrophysiological correlations of SSPE in adolescent and young adult populations. While the classic presentation of SSPE involves insidious scholastic decline followed by generalized slow myoclonus, atypical manifestations are increasingly recognized and account for substantial diagnostic delays.

Electroencephalography remains the single most informative bedside investigation in SSPE. The quintessential neurophysiological signature is the Radermecker complex.

Our four cases delineate the broad spectrum of SSPE presentations This case series illustrate focal onset, neurosurgical confounders, unimmunized classic childhood presentation, and protracted psychiatric masquerades. The primary limitation is the lack of confirmatory CSF measles antibody testing in Cases 2, 3, and 4 due to parental refusal or advanced presentation. However, the presence of unequivocal, pathognomonic Radermecker periodic complexes time-locked to clinical slow myoclonus, together with characteristic clinicoradiological progression, firmly establishes the diagnosis.

7. Conclusion

Subacute Sclerosing Panencephalitis presents with a wide spectrum of clinical phenotypes beyond the classical textbook description. Clinicians must maintain a high index of suspicion in children and young adults presenting with unexplained behavioral changes, scholastic decline, focal neurological deficits, or movement disorders. Electroencephalography is an indispensable, non-invasive diagnostic modality where the identification of periodic slow-wave Radermecker complexes provides immediate, definitive diagnostic confirmation. Intensified measles vaccination efforts remain vital to eliminating this catastrophic neurodegenerative illness.

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