

Periodic Myoclonus and Bilateral Macular Chorioretinitis in a 12-Year-Old Boy: A Multimodal Diagnostic Approach to Subacute Sclerosing Panencephalitis

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Abstract:

Background: Subacute sclerosing panencephalitis (SSPE) is a progressive, fatal measles-related encephalitis that persists where measles immunisation is incomplete, and its early features are often non-specific.

Case Presentation: A 12-year-old boy, probably unvaccinated against measles, presented with a 10-month history of headache and progressive visual loss, followed by behavioural change, cognitive decline, periodic involuntary jerks for four months and generalised seizures for one month. He was drowsy and mute, with increased tone and paucity of movement in the left limbs and a left extensor plantar response. Fundoscopy showed bilateral pigmented macular chorioretinal scars. Electroencephalography (EEG) showed generalised, stereotyped, high-amplitude periodic slow-wave complexes recurring every 4–5 seconds over a suppressed background, and simultaneous EEG–electromyography polygraphy demonstrated time-locked muscle bursts, confirming periodic cortical myoclonus. MRI brain revealed asymmetric (right > left) cortical–subcortical T2/FLAIR hyperintensities with patchy diffusion restriction in the right parietal and left occipital lobes. Cerebrospinal fluid (CSF) was acellular with mildly raised protein (65 mg/dL) and a markedly elevated measles IgG titre (625 by ELISA), fulfilling the modified Dyken criteria for definite SSPE. He was started on isoprinosine and levamisole without clinical improvement, and serial video recordings documented worsening myoclonus over one month.

Conclusion: Visual loss from macular chorioretinitis may precede the neurological signs of SSPE by several months. Integrating fundoscopy, periodic EEG complexes, EEG–EMG polygraphy, MRI and CSF measles serology allows a confident diagnosis, while measles immunisation remains the only effective prevention.

Keywords:

Subacute sclerosing panencephalitis, measles, periodic myoclonus, Radermecker complexes, chorioretinitis, EEG–EMG polygraphy.