Teaching Video NeuroImage: Slow Axial Myoclonus in Subacute Sclerosing Panencephalitis


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Slow periodic myoclonus (up to 1 second in duration) is a distinctive phenomenology described in fulminant subacute sclerosing panencephalitis (SSPE).1 We describe slow axial myoclonus in 2 children having SSPE (Videos 1 and 2). The slow myoclonus causes sudden alteration in tone of axial musculature, causing head drop or backward and sideways tilting of the trunk (axial tilt sign). The involvement of sensorimotor integration mechanism and basal ganglia ictal activity have been proposed as possible mechanisms for the periodic dystonic myoclonus.2 Basal ganglia involvement may also be responsible for associated dystonia. Fulminant Wilson disease and nonrhythmic, repetitive axial myoclonic jerks of propriospinal origin are important differential diagnoses.3,4

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