Teaching Video NeuroImage: Slow Axial Myoclonus in Subacute Sclerosing Panencephalitis

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Slow periodic myoclonus (upto 1 second in duration) is a distinctive phenomenology described in fulminant subacute sclerosing panencephalitis (SSPE).\(^1\) We describe slow axial myoclonus in two children having SSPE (Videos 1 and 2). The slow myoclonus causes sudden alteration in tone of axial musculature; causing head drop or backwards 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 Wilsons disease and non-rhythmic, repetitive axial myoclonic jerks of propriospinal origin form important differential diagnoses.\(^3,4\)

**Video Case 1: 10-year-old boy with slow axial myoclonus and head drop**

10-year-old boy with history of measles in childhood (around 15 months age); presented with recent onset progressive cognitive decline, seizures and periodic slow myoclonus in axial muscles – causing the neck and trunk to fall back. In addition, there was segmental myoclonic in neck and upper limbs - synchronous in flexor muscles; classical of SSPE. Myoclonic jerks were also noted while walking (see the last few seconds of the video), but they did not cause a fall. MRI brain screening was normal. EEG showed high amplitude, generalized spike and wave periodic discharges. The diagnosis of ‘probable’ SSPE was established using Dyken’s criteria (periodic, stereotyped, high voltage discharges + positive CSF anti-measles antibody).

**Video Case 2: 9-year-old girl with slow axial myoclonus and axial tilt sign**

9-year-old girl, never vaccinated and with history of fever in infancy; presented with recent onset cognitive decline, ataxia and abnormal movements. The video shows repetitive, periodic, slow myoclonic movements in axial muscles causing tilting of trunk backwards and to left (axial tilt sign). She also has dystonia in hands and toes; along with slow segmental myoclonic jerks involving flexor muscles of lower limbs. These myoclonic jerks do not interfere with consciousness and do not cause her to fall while walking. A non-contrast CT scan of brain did not show any major abnormality. A diagnosis of ‘probable’ SSPE was made using Dyken’s criteria (periodic sharp-slow wave discharges + positive CSF anti-measles antibody).


References:

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