■ Case vignette

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Epileptic Disorders

Auditory and emotion-sensitive jerks in a patient with subacute sclerosing panencephalitis

Ruchika Tandon, Varsha Ambwani

Department of Neurology, Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, 226014 India

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• **Correspondence:**
Ruchika Tandon
Department of Neurology,
Sanjay Gandhi Postgraduate Institute of Medical Sciences,
Lucknow 226014, India
<rtlib161080@gmail.com>

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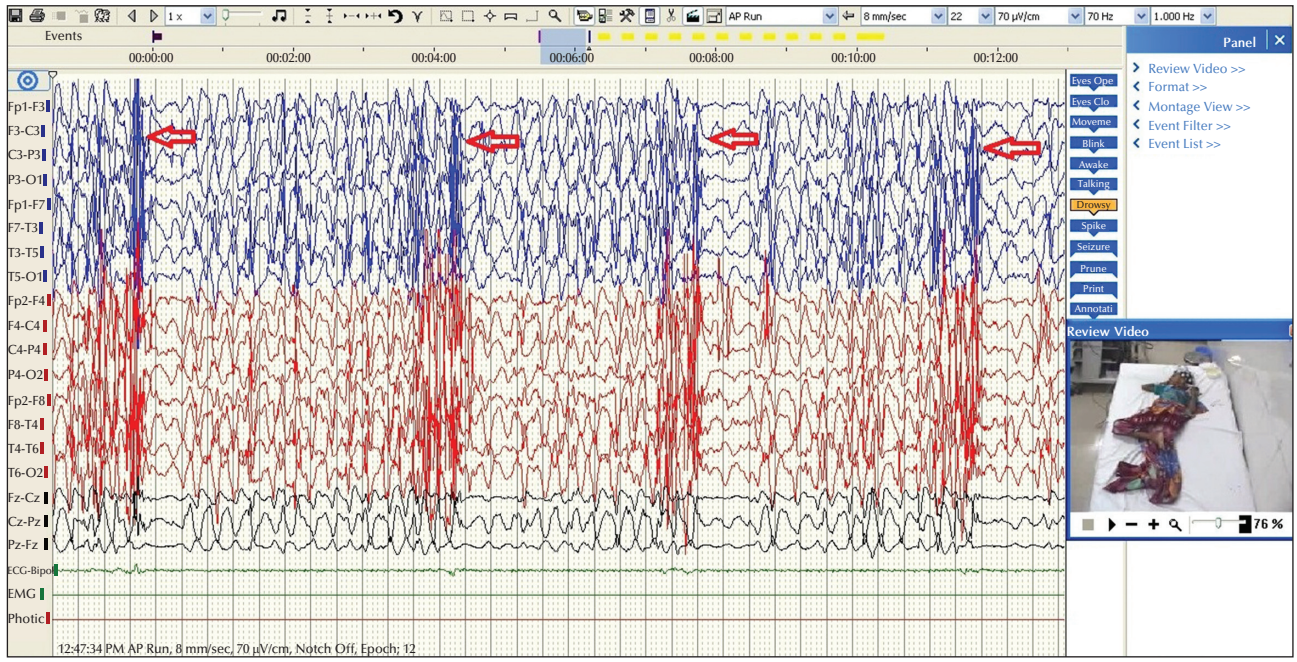
Stimulus-sensitive myoclonus is a known feature of progressive myoclonic epilepsies, although the types of stimuli may differ in these disorders [1]. Myoclonus in subacute sclerosing panencephalitis (SSPE) is, however, characteristically, non-stimulus-sensitive [2]. We describe a young boy in whom sound and emotional stimuli provoked myoclonic jerks, who was positive for measles antibodies.

A nine-year-old boy presented with altered behaviour including inappropriate laughing, irritability and crying, reduced scholastic performance over 9-10 months, sudden jerks and falls while walking for six months, an inability to walk without support for 2-3 months and was bed-bound for the last two months. He did not have generalized tonic-clonic seizures (GTCS), family history was unremarkable with no evidence of birth hypoxia, and he had normal developmental milestones up to eight months ago. According to his father, he had received scheduled vaccines but did not have a vaccination card. He was on levetiracetam and clobazam. On examination, he was conscious and followed commands. He was thin with increased muscle tone and demonstrated +2 limb reflexes, and was able to move his limbs spontaneously and often in response to commands. He had sudden brief jerky movements of the limbs that were precipitated by auditory and emotional (staring at him as if to scold him) stimuli (*video 1, 2*). Therefore, we considered the differential diagnoses of progressive myoclonic epilepsy, inborn errors of metabolism and SSPE. Fundus was normal. Brain MRI was normal and the age range ruled out dentatorubral-pallidoluysian atrophy. Serum TSH was 1.09 miu/L and HIV ELISA was non-reactive. EEG showed quasi-periodic slow-wave complexes every 6-10 seconds with background slowing and no photoparoxysmal response (*figure 1*). Visual evoked potentials were not recorded and somatosensory evoked potentials (SSEP) did not show giant SEPs (N19 on both sides= 17.7, right P40= 29.5 [1.2], left P40= 36.4 [0.47], right CCT= 11.35 and left CCT= 18.74).

Clinicians excluded juvenile myoclonic epilepsy due to intellectual decline and on the basis of the EEG as well as mitochondrial disorders on the basis of normal baseline and post-exercise lactate levels. Fast intellectual decline and very fast progression of myoclonic jerks helped in ruling out Unverricht-Lundborg disease, despite stimulus-sensitive myoclonus. A negative family history and absence of photosensitivity ruled out Lafora body disease, and the absence of cherry red spot and movement- and touch-sensitive myoclonus excluded sialidosis. The absence of photosensitivity excluded neuronal ceroid lipofuscinosis. Negative screening for inborn errors of metabolism ruled out Gaucher's disease and Tay-Sachs disease. Hence, our workup did not reveal findings suggestive of other progressive myoclonic syndromes.



■ **Figure 1.** EEG image of the patient showing quasi-periodic slow-wave complexes every 6-10 seconds with background slowing.

Routine CSF was normal. CSF measles IgG was 11588.4 U/mL, serum measles IgG was 10632.5 U/ml, CSF total IgG was 8.87 mg/dL (normal: 0-3.4 mg/dL), serum total IgG was 1640 mg/dL (normal: 700-1600 mg/dL) and CSF/Serum Quotient was 4.25 (positive > 1.5). Hence, we diagnosed SSPE and administered isoprinosine and intrathecal alpha-interferon, sodium valproate, clobazam and zonisamide, which reduced seizure frequency. It was not possible to perform genetic testing as his parents could not afford this.

SSPE is rare (as a result of increased immunization), progressive and usually fatal (<5% survival), affecting children aged 8-11 years with a higher prevalence in developing countries. It may cause altered behaviour, myoclonus, cognitive decline, gait unsteadiness and coma [3]. Persisting latent mutant measles virus causes SSPE [4]. Here, epileptiform discharges probably stimulate the motor cortex resulting in myoclonus jerks. Sensitivity to unexpected somatosensory stimulus delivered to the mantle area of the cortex causes startle reflex. Hyperekplexia is pathological, acquired or inherited, resistant to habituation, and involves an exaggeration of normal startle responses (brainstem reflex) [5]. In SSPE, periodic pattern and motor movement is usually unaffected by external stimuli [6]. There are some reports of myoclonic jerks elicited by stimulus in patients with SSPE or perhaps hyperekplexia [6, 7]. No explanation is available for this phenomenon, but one possible explanation could be heightened sensitivity, such as hyperacusis in SSPE, resulting in auditory stimulus-responsive myoclonus [8]. Radermecker *et al.* have stated that, early during the disease course, sound, light, or touch stimuli may elicit EEG complexes. Later, however, complexes are no longer affected by stimuli [9]. Hence, other possible hypotheses for stimulus-sensitive jerks could be based on SSPE complexes evoked by sensory stimuli to the cortex when the rhythmic pattern is unstable, and after establishing the periodic pattern, EEG complexes and motor accompaniments would remain unaffected by external stimuli [10]. Our patient had spontaneous jerks and sound and emotional stimuli, such as scolding, induced jerks, however, we did not observe giant SEPs. Although there is significant overlap between myoclonic jerks seen in hyperekplexia and those seen in our patient, and we usually see these jerks only in the early stages of the disease and they are similar to myoclonic jerks observed without stimuli, we therefore feel that these jerks represent a type of epileptic myoclonus which is stimulus sensitive initially but and later becomes non-stimulus sensitive. To our knowledge, auditory and emotional stimulus-sensitive myoclonus is not well known in SSPE and this finding should not dissuade physicians from diagnosing SSPE. ■

Disclosures.
None of the authors have any conflicts of interest to disclose.

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Legends for video sequences

Video sequence 1

Video of the patient with SSPE showing the duration between two myoclonic jerks at rest and a jerk following a stimulus.

Video sequence 2

Video of the patient showing simultaneous EEG recording.

Key words for video research on www.epilepticdisorders.com

Phenomenology: myoclonic seizure
Localization: generalized
Syndrome: progressive myoclonic epilepsy
Aetiology: infection (brain)

TEST YOURSELF

(1) What are the common causes of stimulus-sensitive myoclonus?

(2) Can SSPE present with stimulus-sensitive jerks?

(3) When can SSPE present with stimulus-sensitive myoclonus?

Note: Reading the manuscript provides an answer to all questions. Correct answers may be accessed on the website, www.epilepticdisorders.com.