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Palatal myoclonus in a child: Herald of acute encephalitis

To the Editor: In adults, palatal myoclonus occurs months after the onset of a lesion involving the triangle of Mollaret (inferior olive, red nucleus, and contralateral cerebellar nuclei); it may be caused by an infarct, demyelination, or a neoplasm. There have been few cases in children and none in which palatal myoclonus was the presenting feature of an evolving lesion, as in the following case.

C.M., a 5½-year-old boy, had been treated for acute lymphocytic leukemia with prednisone, intrathecal methotrexate, and cranial irradiation. He had been in bone marrow and CNS remission for 2 years, with no clinical or CT evidence of latent adverse effects. Except for a mild respiratory infection, he had been well; following a dose of intrathecal methotrexate, he was admitted for the onset of "twitching" eyebrows, dysphagia, and dysarthria.

Examination revealed a cachectic boy with no fever, rash, or striae. He was alert and followed three-step commands, but did not speak and uttered only broken syllables. Cranial nerve functions were normal except for continuous, rhythmic, "blinking" contractions of the orbicularis oculi muscles. The movements were bilateral, sometimes more pronounced on the right. Similar movements of the soft palate were synchronous with the orbicularis movements, at a rate of about 1.5 per second. The eyes were not affected; pupils and fundi were normal. Facial strength was equal and full, and a gag reflex was present. Proximal limb wasting and weakness were attributed to steroid myopathy.

During the next week, right facial contractions appeared and were asynchronous with the orbicularis and palatal movements. Following this, right hand and toe jerks appeared, also out of phase with the facial movements. The palatal myoclonus became intermittent. After the fifth hospital day, the child became lethargic.

Brainstem evoked potentials and brain CT were normal. EEG revealed a left slow-wave focus that followed the facial contractions but preceded limb jerks (figure). Later, the EEG showed encephalopathic changes compatible with the child's lethargy. On the second day, the CSF fluid was acellular with normal protein and glucose, but the peripheral blood white cell count was less than 1,000. Though smears and cultures for bacteria, tuberculosis, and fungi were negative, viral cultures yielded a picornavirus. On the ninth hospital day, CSF was still normal except for myelin basic protein content of 8.4 mg/ml. On the sixteenth day, the CSF showed 15 white cells and protein of 57 mg/ml. By that time, encephalitis was manifest by obtundation, slow EEG, and diffusely increased contrast in cortical areas on CT. By the end of the second week, there were asynchronous myoclonic jerks of the eyes, right side of the face, left shoulder, and right hand and foot. Phenytin and phenobarbital had no effect; IV diazepam, administered under EEG monitoring, abolished the movements without affecting the EEG. The myoclonus, palatal and generalized, was completely controlled by a combination of clonazepam (0.08 mg/kg/d in three divided doses) and diazepam (1 mg po tid).

A brain biopsy on the seventeenth day revealed microglial nodules suggesting an acute viral infection, but cultures for herpes and picornavirus were negative. The white matter appeared normal with no leukemic infiltrates. Over the following 6 months, the patient remained free of myoclonus, with a normal mental status but with persistent dysarthria and dysphagia as well as a right hemiparesis.

Matsuo and Ajas suggested denervation supersensitivity as the underlying mechanism for palatal myoclonus. They noted the "obligatory" lag time between onset of a lesion (hemorrhage, infarction, and so on) and the appearance of the palatal myoclonus. The onset of the movement disorder was always at least 2 months after the inciting event, with a mean interval of 10 to 11 months. Thus, in all reported cases, palatal myoclonus has been a new symptom related to an old, established lesion.

Our patient had been in remission from acute lymphocytic leuke-

Figure. EEG depicting contraction of the facial muscles preceding the onset of a slow cortical wave, maximal at the left temporal leads.