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MR Demonstration of Cryptic Vascular Malformation Producing a Palatal Myoclonus

—A Case Report—

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A 47-year-old man had suffered oscillopsia associated with palatal myoclonus for 10 years. High-field magnetic resonance imaging (MRI) revealed a cryptic vascular malformation within the "Guillain-Mollaret triangle" which was thought to be the responsible lesion.

Key Words: *Palatal myoclonus, High-field magnetic resonance imaging, Cryptic vascular malformation Guillain-Mollaret triangle*

INTRODUCTION

Palatal myoclonus denotes a rhythmic involuntary jerky movement of the soft palate. This rhythmic contraction may spread to the adjacent muscles such as external ocular muscles, tongue, larynx, face, neck or diaphragm, but most often is restricted to the oropharyngeal muscles (Cooper, 1958). This condition is usually attributed to a localized lesion of the brainstem or cerebellar nuclei. Although vascular malformations affecting the brainstem are not unusual (Mc Cormic et al., 1968), cases causing either palatal or palato-ocular myoclonus (POM), to the best of our knowledge, have not been so far reported.

In this communication, we describe a patient with POM in whom a cryptic vascular malformation of the brainstem was demonstrated on a high-field MR imaging.

CASE REPORT

A 47-year-old man had been well until the age of 37, when he first noticed surroundings bouncing up and down and felt pins and needles sensation in his left cheek. The abnormal sensation had then progressed over the next 48 hour span to the right accompanied by some weakness of the right extremities. Examination done elsewhere revealed a mild ataxic gait which resolved spontaneously thereafter. However the oscillopsia and mild right sided sensory disturbance has persisted until the present time with patient being on no pertinent medication.

On examination, he was alert and mild hypesthesia on the right side of the face with decreased corneal reflex was noted. Ocular examination was remarkable in that there were rhythmic torsional movements in both eyes, the excursion of the right eye were of a greater amplitude and more rotatory than those of the left. In addition, the soft palate and posterior pharynx showed bilateral rhythmic involuntary movements. These movements were apparently synchronous with the ocular oscillations at a rate of 100 per minute and subsided during sleep. The extraocular eye movements were full and the oscillopsia was independent of fatigue. His speech was slurred, and on phonation there was a definite quiver of his voice. The remainder of neurological examination were not remarkable.

The results of routine hematological, biochemical and serological tests were normal. And the CSF ex-

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amination was also unrevealing including tumor marker studies. Brainstem auditory evoked responses (BAER) revealed a prolongation of central conduction time which suggested functional derangement in the brainstem. A brain CT performed elsewhere disclosed a hyperdense and poorly enhanced lesion of 1.5×1.8 cm in size at the posterior aspect of the cerebral peduncle. Four-vessel angiography failed to reveal any pathologic vessels or arteriovenous shunting. MR imaging was done on a 2.0-T superconducting system using spin-echo pulse sequences, which revealed a circumscribed region of low intensity containing central focus of high-intensity signal in the brainstem. After the clinical diagnosis of palato-ocular myoclonus caused by cryptic vascular malformation, several medications such as trihexyphenidyl, diazepam, and clonazepam were tried without any benefit. Currently he is on carbamazepine which still gives no clinical improvement.

DISCUSSION

Palatal myoclonus is a classical example of focal myoclonic disorder restricted to specific muscle group and usually due to a well-defined focal brainstem lesion (Lapresle, 1986). This unusual, queer movement disorder is most commonly due to cerebrovascular disease (infarct more often than hemorrhage), but can also be seen after a variety of neurologic lesions including multiple sclerosis, tumors, trauma and metabolic encephalopathy (Marsden et al., 1982). The pathological lesion responsible for palatal myoclonus has been claimed to be in the so-called Guillain-Mollaret triangle, an area including the red nucleus, the inferior olivary nucleus, the dentate nucleus or their connections, especially the central tegmental tract and the dentato-olivary pathway (Nathanson, 1956; Lapresle and Ben Hamida, 1970; Koeppen et al., 1980). Cases of palatal myoclonus that have been submitted to postmortem examination have revealed

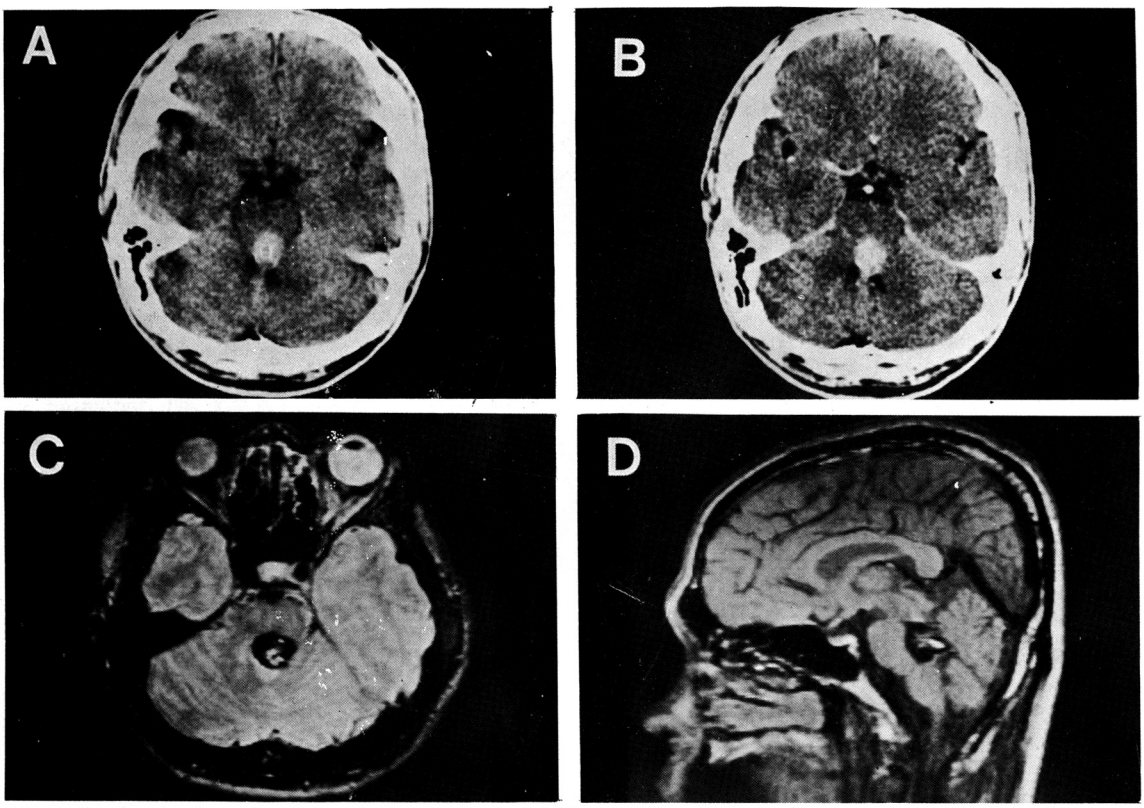

Fig. 1. Cryptic vascular malformation. A, Unenhanced axial CT scan demonstrates hyperdense pontine lesion with little mass effect. B, Axial CT scan with IV contrast. C, Axial T2-weighted image (TR=2,000 ms, TE=60 ms) shows a hypointense periphery and the *variably* isointense center in the brainstem. D, Sagittal T1-weighted image (TR=500 ms, TE=30 ms) localizes the lesion in the Guillain-Mollaret' triangle.

hypertrophic degeneration of the inferior olivary nucleus (Nathanson, 1956: Lapresle and Ben Hamida, 1970). While this hypertrophy of inferior olivary nucleus is the main pathologic finding, histologically it is characterized by enlarged, vacuolated neuron with hypertrophic astrocytes. Recent histochemical study revealed that hypertrophic neurons and their dendrites contain increased acetylcholinesterase reaction products (Koeppen et al., 1980).

The case here reported has a 10-year history of oscillopsia with a non-progressive palatal myoclonus detected in adult years. When the first non-enhanced CT scan revealed a hyperdense lesion (Fig. 1-A, B), the initial presumption was hemorrhagic neoplasm. But subsequent 4-vessel angiography revealed no tumor stain or abnormal vasculature. And the lack of progression of symptoms during the last 10 years prompted us to search for another diagnostic consideration. High-field MR images disclosed a characteristic appearance in the brainstem, i.e., circumscribed region of low signal intensity interpersed by various signal intensities within it (Fig. 1-C). The peripheral zone of low intensity is usually attributed to the paramagnetic effect of hemosiderin deposition. And the characteristic finding found has been described to be specific and virtually pathognomonic of cryptic vascular malformation (Gomori et al., 1985: Gomori et al., 1986).

In summary, we have observed palato-ocular myoclonus in a patient who was thought to have an intrinsic vascular malformation occult to angiography in the brainstem. The high-field MRI localized the lesion which we considered is responsible in the so-called Guillain-Mollaret triangle (Fig. 1-D). Thus our recommendation is to use high-field MRI routinely in an appropriate clinical setting and once the underlying neoplasm could be excluded, the cryptic vascular malformation should be included among the etiologic considerations causing palato-ocular myoclonus.

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