Unusual CT and MRI findings in palatal myoclonus

A 23-year-old man suffered from palatal myoclonus for 2 years. It had appeared one week after a minor head trauma. MRI and basal cisternography revealed a localized atrophy of the left paramedian part of the medulla, encroached upon the left vertebral artery. Clonazepam treatment was beneficial. This particular case is discussed in relation to etiopathogenesis and other causes of palatal myoclonus.

Palatal myoclonus usually is a delayed clinical manifestation of interruption of the dentato-olivary pathway at one of the preferential sites: the ipsilateral tegmental tract and the contralateral dentate nucleus (1). This leads to transsynaptic degeneration of the ipsilateral olivary nucleus in the lower medulla oblongata. It has most often been caused by a vascular lesion of the brainstem or cerebellum. Several other causes have rarely been associated such as multiple sclerosis, neoplasm, infectious or inflammatory diseases, electroshock therapy, Arnold-Chiari malformation, and so on (2-7). We now report another case with an unusual lesion visualized on both MRI and basal cisternography.

Case report

A 23-year-old man was referred to the department of neurology for evaluation of palatal myoclonus. He complained for two years of constant rhythmic palatal movements and continuous earclicking with tinnitus on both sides. He had suffered a minor head trauma without loss of consciousness one week prior to the beginning of the complaints. Previous treatment with carbamazepine in conventional daily doses for more than 4 months had no effect. The family history was negative. Clinical examination showed a bilateral and symmetrical myoclonus of the soft palate and of the anterior and posterior tonsillar pillars at a frequency of 100/min. There were no other associated movements. The neurological examination was otherwise normal.

CT-scan of the brain with special attention to the posterior fossa was unremarkable. MRI, however, showed a lesion on the left paramedian side of the medulla oblongata, at the level of the bottom edge of the clivus; next to the medulla was a small space-occupying lesion, separated from the medulla by a small, round, signal-free zone (Fig. 1). The lesion turned out to be the cranial part of the left vertebral artery, which lied against a localized atrophy of the medulla as shown by a basal cisternography (Fig. 2). Four-vessel angiography was normal.

Visual, somatosensory and auditory brainstem evoked potentials, EEG, standard X-rays of the skull and chest, oesophageal cineradiography, blood and spinal fluid analyses – including agargel electrophoresis and syphilis serology – were normal. Sympathetic and parasympatic function testing was normal.

The palatal myoclonus could be suppressed by clonazepam. The results of control investigations, including MRI, performed one year later were unchanged. Clonazepam was still capable of suppressing the palatal movements.

Discussion

Palatal myoclonus is characterized by involuntary and usually unconscious movements of the palate and the pharynx. This disorder is most commonly...
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Fig. 1. MRI shows a left-sided lesion at the level of the bottom edge of the clivus. The medulla is slightly displaced to the right by a small space-occupying process (arrow), separated from the medulla by a signal-free zone.

bilateral and symmetric, but it can be unilateral, the palate and the uvula then being drawn to one side. The movements are more or less continuous, usually between 100 and 150/min., with exceptional rates of 20/min. and 600/min. (1). Palatal myoclonus can be associated with synchronous movements of the larynx, eyes, face, the floor of the mouth, the tongue, the diaphragm and skeletal muscles (1, 8). Sometimes, as in our case, the patient can hear a synchronous clicking due to the rhythmic involvement of the Eustachian tube, or the clonic contraction of the tensor tympani and the stapedius muscles. Involvement of the oculomotor system is not rare and the condition is then termed "oculopalatal myoclonus" (7, 9).

Palatal myoclonus is due to lesions involving the dentato-rubro-olivary pathway. This pathway travels from the dentate nucleus through the superior cerebellar peduncle and crosses to the opposite side in the commissura of Wernerkink along the internal and dorsal surface of the red nucleus, before it reaches the inferior olivary nuclei via the central tegmental tract (1, 6).

The anatomic basis for the palatal myoclonus is a hypertrophic degeneration of the inferior olivary nuclei in the medulla oblongata. The lesion is considered to be transsynaptic in origin, as it is mostly associated with a supra-olivary causal lesion, the two preferential sites being the ipsilateral tegmental tract and the contralateral dentate nucleus. Microscopic findings include enlargement of the neurons or astrocytes or both (8, 10, 11, 12).

Cerebrovascular insults due to thrombosis, hemorrhage or embolism, affecting the brainstem or the cerebellum, are the most common causes, but also multiple sclerosis, tumors, trauma, infectious or inflammatory processes, electroshock therapy, vertebral artery aneurysm, subacute myelo-optic neuropathy, dialysis encephalopathy, Arnold-Chiari malformation and degenerative processes have been incriminated (2-7). In our case a localized atrophy of the left side of the medulla is visualized, wherein the medially shifted cranial part of the left vertebral artery is seen. We relate the clinical symptoms to this finding. Is the atrophy due to the pulsatile hammering effect of the vertebral artery, which in the long run has inflicted damage upon neighbouring structures, or to the head trauma? Neither possibility can be proved. All other classical causes were carefully ruled out. Four-vessel angiography was normal. Palatal myoclonus after head injury on the other hand, has been described (3), but not, as far as we know, after such a minor head trauma. The myoclonus in our patient also appeared rapidly after the trauma. Usually, the myoclonus is delayed with regard to the causal lesion, usually from 6 to 9 months (6), but ranging from 1 day (13) to 30 months (14).

Treatment of palatal myoclonus and of branchial myoclonus in general is often a matter of trial and
error. The neurotransmitters involved have not yet been defined, though some believe that serotonin or its precursors are implicated. Good results have been reported using 5-hydroxytryptophan combined with a decarboxylase-inhibitor (15, 16), but others saw no improvement at all. A lot of drugs have been reported to be beneficial in isolated cases: trihexyphenidyl (17), carbamazepine (18–20), phenitoin (21), valproic acid (22), clonazepam (23, 24), and tetrabenazine (24). Our present patient did not respond to carbamazepine treatment but improved dramatically after clonazepam administration.

References


Fig. 2. Basal cisternography shows the contrast surrounding the left vertebral artery (arrow) and the atrophic part of the medulla.


