Palatal myoclonus induced by extirpation of a cerebellar astrocytoma

Case report

Kazuyuki Nishigaya, M.D., Masami Kaneko, M.D., Yoshishige Nagaseki, M.D., and Hideaki Nukui, M.D.

Iwata Neurosurgical Hospital, Iwata, Shizuoka, Japan; and Department of Neurosurgery, Yamanashi Medical University, Nakakoma-gun, Yamanashi, Japan

A 45-year-old woman developed a rare case of palatal myoclonus with no other neurological signs after undergoing extirpation of a small cerebellar low-grade astrocytoma that was located in the right dentate nucleus. The palatal myoclonus has persisted for 4 years after the operation. Magnetic resonance T$_1$-weighted imaging revealed a high-intensity lesion in the left inferior olive. Palatal myoclonus associated with the removal of cerebellar tumors is unusual but may easily be overlooked.

Key Words • myoclonus • palatal muscle • astrocytoma • cerebellum • dentate nucleus

Palatal myoclonus is a rare movement disorder characterized by involuntary, rhythmic contractions of the palatal muscles. This hyperkinetic movement develops as a result of lesions located within a pathway that extends from the contralateral dentate nucleus via the brachium conjunctivum and the ipsilateral central tegmental tract to the inferior olive, originally known as the Guillain–Mollaret triangle. The most common identifiable cause of palatal myoclonus is cerebrovascular disease; this is followed by brainstem tumors, multiple sclerosis, encephalitis, brainstem trauma, and obstructive hydrocephalus. Although the symptomatology of the dentatofugal projection system remains unclear, patients who undergo surgery for tumors involving the cerebellar hemisphere usually present with intention tremor or ataxia in the ipsilateral extremities rather than palatal myoclonus. Only two cases of palatal myoclonus caused by removal of a cerebellar hemangioblastoma have been reported.

We describe a rare case of palatopharyngeal myoclonus that developed 3 months after removal of a small low-grade astrocytoma located in the right dentate nucleus. The case was exceptional in that palatal myoclonus was the only symptom, and no other profound neurological dysfunction appeared.

Case Report

History. This 45-year-old woman sustained a minor head trauma and consulted staff at our hospital in October 1992 because she suffered from a continuous headache.

Examination. Neurological examination detected no deficit, but noncontrast-enhanced computerized tomography (CT) scanning revealed a round high-density mass lesion in the right cerebellar hemisphere (Fig. 1). Magnetic resonance (MR) T$_1$-weighted imaging demonstrated an inhomogeneous low-intensity mass lesion measuring 2 cm in diameter. The central region of the mass was enhanced by administration of gadolinium-diethylenetriamine pentaacetic acid (Gd-DTPA). The T$_2$-weighted image displayed a high-intensity area surrounding a small round low-intensity mass. The tumor was suspected to involve the region corresponding to the right dentate nucleus on MR imaging (Fig. 2).

Operation. In November 1992, a suboccipital craniotomy was performed with the patient in the lateral decubitus position. A corticotomy was made in the inferomedial surface of the right cerebellar hemisphere with the aid of the operating microscope, and dissection was advanced in the superolateral direction. A greenish yellow, hard elastic area of tumor and a whitish yellow, soft elastic area of tumor with many tiny vessels were recognized in the inferior and the superior portions of the tumor, respectively. The mass lesion was poorly defined with respect to normal cerebellar tissue and gliosis was suspected during the operation; therefore, extensive tumor removal was not performed. However, a second operation was undertaken.
1 month later to extirpate the remaining tumor until normal tissue was identified because the histological diagnosis of the specimen obtained at the first operation was astrocytoma.

Postoperative Course. The patient developed no neurological deficits. However, approximately 3 months after the first operation, she complained of dysarthria. Examination revealed an involuntary rhythmic palatopharyngeal movement on the right side that appeared at a rate of approximately 140 times per minute. Although no definite involuntary movement was found in her vocal cords, she reported that her voice became falsetto in the midst of singing a song and tired after talking for more than 30 minutes. She never heard a continuing rhythmical ear-click. Clonazepam was administered; however, the palatopharyngeal myoclonus did not regress. The patient’s myoclonus has persisted, but she has made no further complaint.

No high-density mass lesion was revealed on postoperative CT scanning and no tumor recurrence has been identified on biannual MR imaging for 54 months after resection of the tumor. A low-intensity area was identified in the right cerebellar hemisphere on $T_1$-weighted images; this area was not enhanced by addition of Gd-DTPA (Fig. 3). On $T_2$-weighted imaging, a high-intensity lesion was located in the left inferior olivary nucleus (Fig. 4).

Histological Examination. The tumor cells had medium-sized round nuclei with a prominent chromatin network, and fine neuroglial fibrils occupied the matrix. Mitoses and cellular atypism were absent. The tumor was composed mostly of fibrillary astrocytoma and included microcystic degeneration. Gliosis and calcification were present in the surrounding area. This tumor was diagnosed as a low-grade (Grade 2) astrocytoma (Fig. 5).

Discussion

Nine cases of palatal myoclonus provoked by cerebellar tumors have been reported in the literature. Two patients developed palatal myoclonus 4 and 12 months after removal of the tumors, both of which were hemangioblastomas. However, the lesion that induced the palatal myoclonus could not be conclusively identified in these patients because they had suffered complicating brainstem infarction as a result of surgery. All of the patients with cerebellar tumors had various other neurological signs in addition to palatal myoclonus and their lesions were comparatively large. In contrast, our patient exhibited only palatopharyngeal myoclonus with no other profound cerebellar or brainstem dysfunction. Furthermore, the lesion was restricted to the dentate nucleus where the tumor was located and to a small inferior portion of the cerebellar cortex that had been injured by intraoperative dissection. Why our patient only presented with palatal myoclonus and not with intention tremor or ataxia in the ipsilateral extremities as a cerebellar sign is un-
known. However, this may be related to the different functions topologically localized in the dentate nucleus.

The inferior olive is a likely location for the “oscillator” that provoked the palatal myoclonus. Secondary to denervation, the inferior olive undergoes transsynaptic pseudohypertrophic degeneration over a period of weeks or months. Olivary pseudohypertrophy can be detected by MR imaging and has been observed at autopsy. Histological examination of the pseudohypertrophic inferior olive shows gliosis, enlargement and vacuolation of the neurons, and demyelinization. The serial changes on MR imaging are considered to correspond to chronological changes in the pathological characteristics of olivary pseudohypertrophy. Both T2- and proton-weighted MR imaging of olivary pseudohypertrophy show a gradual development to irregular intensity and slightly atrophic size from homogeneous high intensity and maximum size. In our case we suspect that the dentate nucleus, damaged by extirpation of the tumor, led to denervation of the dentatoolivary pathway and resulted in olivary pseudohypertrophy. The inhomogeneous high-intensity lesion in the ventral portion of the medulla of our patient was also found on the MR image obtained approximately 4 years after the palatal myoclonus appeared.

Cerebellum that has sustained a vascular, neoplastic, inflammatory, or degenerative insult loses the capacity to inhibit the firing of lower motor neurons and motor nuclei of the cranial nerves, resulting in rhythmical contractions.
of the muscle supplied by these neurons. These muscle groups include the ocular muscles, tongue, facial muscles, palatal muscle, pharynx, larynx, and diaphragm. Although many patients who harbor tumors involving the dentate nucleus undergo surgery, these tumors or the operations performed to excise them rarely result in palatal myoclonus. It is possible that palatal myoclonus occurs much more frequently than it is recognized. The typical complaint of patients with essential palatal myoclonus is a continuing rhythmic earclick as the only subjective manifestation. Because earclicks are rare in symptomatic palatal myoclonus, this is the cardinal symptom of essential palatal myoclonus. Patients with symptomatic palatal myoclonus are often unaware of the palatal movements but may complain of coincidental oscillosopia and have symptoms and signs of brainstem or cerebellar dysfunction. Accordingly, palatal myoclonus may easily be overlooked during routine physical and neurological examinations unless the physician is particularly aware of the possibility of this phenomenon or the patient complains of clicking sounds in the throat or ear. There is wide variation in the time interval between the occurrence of lesions within the dentatorubroolivary pathway and the clinical manifestation of palatal myoclonus. Thus, examination of the soft palate should continue to be performed in patients who have undergone surgery for a cerebellar tumor.

References