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Pearls & Oy-sters: Oculopalatal tremor with one-and-a-half syndrome after pontine hemorrhage



CLINICAL PEARLS 1. The palatal myoclonus in symptomatic cases is usually silent as the involved muscle is the levator veli palatini. This is in contrast to the audible clicking sound (generated by the synchronous collapse of the eustachian tube) in patients with essential palatal myoclonus in which the involved muscle is the tensor veli palatini.

2. The hyperintense signal on T2-weighted MRI in transneuronal hypertrophic degeneration of the inferior olivary nucleus reflects increased water content and gliosis. Enlargement of the inferior olivary nucleus results from astrocytic hypertrophy and vacuolar cytoplasmic degeneration. Eventually, permanent atrophy of olivary neurons ensues within 3 to 4 years.

CASE REPORT A 63-year-old man with a history of treated hypertension presented with sudden onset of right hemiparesis and horizontal diplopia that was worse on rightward gaze. His blood pressure was 220/120 mm Hg. On neurologic examination, the patient had a right hemiplegia. Examination of the extraocular movements demonstrated conjugate left lateral gaze palsy, impaired adduction of the left eye, and abduction nystagmus of the right eye. These signs were consistent with a left “one and a half syndrome.” Further assessment revealed a bilateral facial nerve palsy of the lower motor neuron type. The cranial CT revealed intraparenchymal brainstem hemorrhage involving the left side of the pontine tegmentum with extension into the fourth ventricle (figure 1). His motor weakness and the facial diplegia improved, but the extraocular eye movements remained limited with persistent horizontal diplopia, particularly on rightward gaze.

Six months later, he presented again with a precipitous onset of oscillopsia with exaggerated sense of motion and blurring of the visual world during head movements, particularly while walking or driving. On examination, a new finding of a vertical pendular nystagmus was observed with larger amplitude in the right eye (video 1). Another finding of a palatal

Figure 1

Initial CT of the head showing the intraparenchymal brainstem hemorrhage that mainly involved the left side of the pontine tegmentum with extension into the fourth ventricle

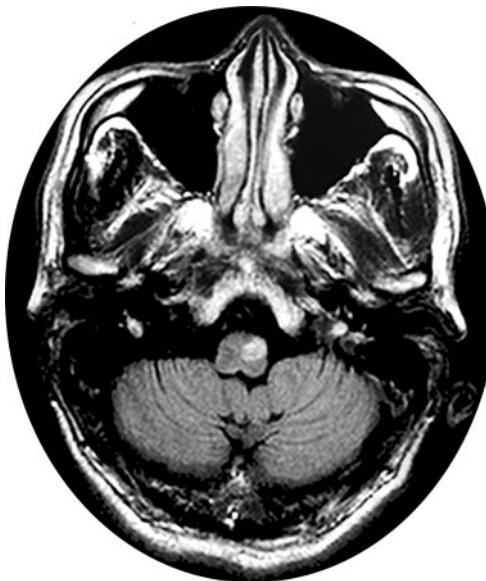


tremor was noted which was more prominent on the right side (video 2). On further inquiry, he did not describe any specific bulbar complaint that could be linked to his palatal tremor. He also was not aware of any unusual noise or clicking sounds in his ears. The palatal tremor persisted during sleep while the pendular nystagmus disappeared. Cranial MRI showed an enlargement of the left inferior olivary nucleus with a hyperintense signal on T2-weighted images (T2WI) (figure 2). The clinical presentation supported by the imaging was consistent with the diagnosis of oculopalatal tremor. The patient was initially treated by a trial of gabapentin for symptomatic relief, but this aggravated the dizziness. He was subsequently prescribed clonazepam with significant improvement in his symptoms despite the persistence of the ocular and palatal tremors.

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Figure 2 Axial FLAIR MRI at the level of the inferior olives revealing the high signal intensity and the hypertrophy of the left inferior olfactory nucleus



DISCUSSION Palatal tremor (PT) is a segmental myoclonus involving the muscles of the soft palate. The movement consists of rhythmic and repetitive, rather than oscillatory, contractions of the agonist muscles only. Occasionally, this hyperkinetic movement may spread to involve adjacent structures derived from the brachial arch such as the larynx and the pharyngeal wall.¹ Oculopalatal tremor (OPT) refers to the synchronous or asynchronous combination of PT and pendular nystagmus. The pendular nystagmus can be symmetric or asymmetric in amplitude with vertical, torsional, or mixed trajectories. Symptomatic PT is usually the delayed sequela of lesions interrupting the Guillain-Mollaret triangle (GMT) and typically manifests several months after the acute event. The myoclonus is generated by the levator veli palatini muscle innervated by the nucleus ambiguus through the facial or glossopharyngeal nerves.

Symptomatic PT is often the result of an ischemic or hemorrhagic stroke disrupting the GMT, but also can result from other lesions to this area such as demyelination and space-occupying lesions. The GMT is the dentato-rubro-olivary pathway that connects the dentate nucleus with the contralateral red nucleus through the superior cerebellar peduncle (SCP), the red nucleus with the ipsilateral inferior olfactory nucleus (ION) through the central tegmental tract (CTT), and the ION back to the contralateral dentate nucleus via the inferior cerebellar peduncle (ICP). The topographic localization of the lesions

can therefore be localized to 1) dentate nucleus, 2) SCP, 3) decussation of SCP, 4) red nucleus, 5) CTT, or 6) ICP.² In our patient, the lesion disrupted the left CTT.

The etiopathology of symptomatic OPT is related to transneuronal hypertrophic degeneration of the ION in response to deafferentation. The gamma-aminobutyric acid (GABA)-dependent synaptic transmission within GMT modulates the rhythmic excitation and inhibition of inferior olivary neurons. OPT is thought to arise from hypersynchronous firing of olivary neurons due to interruption of the supranuclear GABAergic control.^{3,4} This abnormal rhythm is subsequently transmitted through ICP to the contralateral cerebellar flocculus and, thus, interferes with physiologic regulations of the oculomotor system. Disruption of the left CTT in this case resulted in hypersynchronous firing from the left ION toward the right cerebellar flocculus. Impaired supranuclear control over the left ION, therefore, may explain the higher amplitude of nystagmus in the contralateral eye. Alternatively, the pendular nystagmus in OPT has been postulated to result from an impaired adaptation of the vestibulo-ocular reflex (VOR) due to degeneration of the ION. Recently, the pendular nystagmus has been linked to the structural distortion of the adjacent vertical neural integrators or their efferent projections either by the primary lesion or by the hypertrophied ION.⁴

The horizontal one-and-a-half syndrome is characterized by ipsilateral conjugate horizontal gaze palsy and internuclear ophthalmoplegia. The combination results from a unilateral pontine lesion involving the paramedian pontine reticular formation or the abducens nucleus and the ipsilateral medial longitudinal fasciculus. The association between one-and-a-half syndrome and subsequent development of OPT has been initially described by Wolin et al.⁵ In that series, all of the five patients had concomitant facial nerve palsy during their initial presentations. The facial nucleus lies in the close vicinity of the CTT within the pontine tegmentum and the facial nerve fascicles can predictably be injured as they sweep around the abducens nucleus. This close anatomic proximity suggests that the concomitant facial palsy may be a predictor of subsequent OPT after the one-and-a-half syndrome. Nevertheless, the predictive value of an initial facial palsy may not be established from that retrospective review or from this case report.⁵

Pathologically, the olivary enlargement results from neuronal and astrocytic hypertrophy with vacuolar cytoplasmic degeneration. Disintegration of the ION occurs in a few years with dissolution and eventually permanent atrophy of olivary neurons.⁶ Tem-

poral correlation of the pathologic processes is demonstrated on imaging studies with high signal intensity of ION on T2WI reflecting the increased water content and gliosis within the ION. The high-intensity signal within the ION appears as early as 3 weeks after the initial lesion and may persist for 3 to 4 years before permanent olivary atrophy ensues.⁷

In approximately one quarter of cases, however, no structural lesion can be found and the condition is termed “essential palatal tremor.” Unlike symptomatic PT, essential PT involves the tensor veli palatini muscle which is innervated by the trigeminal nerve. The involvement of the tensor veli palatine muscle accounts for the audible clicking sounds often perceived by patients with essential PT and is due to the collapse of the eustachian tubes, the site of insertion of the tensor veli palatini muscle. In contrast to symptomatic PT, essential PT is not associated with abnormal ocular oscillations and the myoclonus tends to dissipate during sleep.¹

The treatment of PT has involved traditional agents used for myoclonus and tremor, particularly GABA agonists such as gabapentin and benzodiazepines. Other pharmacotherapies have been used with variable success including baclofen, anticholinergics, and antiepileptic drugs. Successful control of essential PT has also been achieved with botulinum toxin injections into the tensor veli palatini muscle. Our patient had a good response to clonazepam with

significant improvement in his symptoms. The symptomatic improvement may be theoretically related to the enhanced GABAergic transmission within the GMT. Arguably, the improvement in the oscillopsia might be attributed to different compensation mechanisms which characterize the natural course of this phenomenon.

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