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Hypertrophic olivary degeneration due to the presence of pontine cavernomas

Saumya H Mittal, KC Rakshith, ZK Misri, Shivanand Pai, Nisha Shenoy, Gautam Gundabolu
Department of Neurology, Kasturba Medical College Hospital, Mangalore, Karnataka, India

Correspondence Address:
Saumya H Mittal
Department of Neurology, Kasturba Medical College Hospital, Mangalore, Karnataka
India

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Full Text

Sir,

A 48 year old gentleman developed sudden onset of right-sided hemiparesis associated with slurred speech, facial deviation and difficulty in swallowing. He was found to have cavernomas involving the dorsal pons, inferior midbrain on the left side and the parietal subcortical white matter [Figure 1].{Figure 1}

After the episode, the patient had a persistent feeling of abnormal repeated vertical upsurge and downsurge of his visual field. He presented to the hospital with ocular bobbing [Video 1]. On examination, he was found to have Horner's syndrome and palatal myoclonus [Video 2]. A repeat magnetic resonance imaging (MRI) was suggestive of cavernomas involving the dorsal pons, left caudal midbrain and left parietal subcortical white matter. The finding of T2/fluid-attenuated inversion recovery hyperintense areas with mild expansion involving the left half of the medulla was suggestive of hypertrophic olivary degeneration [Figure 2].{Figure 2}

Hypertrophic olivary degeneration was first recognized by Oppenheim in 1887. Hypertrophic olivary degeneration is a deafferentation syndrome. There is a neuronal loss and reactive gliosis. This happens after a loss of synaptic input from the afferent fibers. This is known to occur at several areas of the central nervous system, e.g., damage to the optic nerve or optic tract causes changes in the lateral geniculate body. Hypertrophic olivary degeneration is the end result of transynaptic degeneration of the inferior olivary nucleus after an injury to the dentato-rubro-olivary tract. This pathway of degeneration was described as the anatomical basis of palatal myoclonus by Guillain and Mollaret in 1931.[1]

Hypertrophic olivary degeneration is a characteristic entity wherein there is enlargement rather than atrophy despite the degeneration. The etiopathogenetic mechanism responsible for this phenomenon is

unknown. However, gliosis and increased water content leading to neuronal ballooning, increased protoplasmic astrocytes, and axonal remodeling may result in the hypertrophy and the associated signal hyperintensity on MRI. Over a period of time, cell death results in neuronal as well as astrocytic loss leading to olivary atrophy.[1]

The symptom of palatal myoclonus (rhythmic, involuntary movement of the oropharynx, including the soft palate and uvula) is almost pathognomonic of hypertrophic olivary degeneration and should heighten its suspicion. There may be a loss of inhibitory control by damage to the dentatorubral pathway with consequent hyperactivity of the olivary neurons leading to palatal myoclonus and other involuntary movements. The symptoms persist even after the resolution of hypertrophy due to the lack of feedback mechanism and disturbance of natural rhythmicity.[1] Cases have been reported where the patient presented with dentatorubral tremors but no palatal myoclonus.[2] Another case was reported with Holmes tremors (2–5 Hz tremors present at rest and increased on intention in an upper limb).[3] Besides the palatal myoclonus and dentatorubral tremors, ocular myoclonus may also be seen.[2] Severe myoclonus of the cervical muscles and the diaphragm, preceding the palatal myoclonus, have also been reported.[4] Other findings include choreodystonia, ataxia, dysarthria, and diplopia.[5]

Radiologically, the T2-hyperintense lesion is seen anywhere in the triangle of Guillain and Mollaret including the dentate nucleus, superior cerebellar peduncle, red nucleus, or pontine tegmentum. The changes may occur as early as 1-month post insult, and persist for 3–4 years. Olivary hypertrophy is the other finding of this entity that comes late at 6 months post injury and resolves at 3–4 years.[1]

Another possible modality for detection of this entity is the diffusion tensor imaging. There is radial diffusivity suggestive of demyelination and axial diffusivity suggestive of neuronal hypertrophy. Brain stem neoplasms and infections may also show these changes but may be distinguished from hypertrophic olivary degeneration as the former entities also show a contrast enhancement. Amyotrophic lateral sclerosis, Wallerian degeneration, and adrenoleukodystrophy would show hyperintensity of the corticospinal tract.[2]

The triangle of Guillain and Mollaret is formed between the red nucleus (located in the midbrain), inferior olivary nucleus (located in the anterolateral medulla) and contralateral dentate nucleus [Figure 3].[2] Red nucleus receives afferents from the contralateral dentate nucleus through the cerebellorubral and dentatorubral tracts (that traverses the superior cerebellar peduncle). Inferior olivary nucleus receives afferent fibers from the ipsilateral red nucleus by central tegmental tract (that traverses the central brainstem tegmentum). Finally, olivocerebellar tract and its efferent fibers traverse the inferior cerebellar peduncle to connect the inferior olivary nucleus to the contralateral dentate nucleus.[2]{Figure 3}

The inferior olivary nucleus has a role in coordination while the dentatorubral tract plays a role in fine voluntary motor movements. Olivodentate tract maintains the cerebellum and lesion of this tract is known to cause cerebellar atrophy (but not palatal myoclonus or hypertrophic olivary degeneration).[1] Therefore, if the primary lesion is in the central tegmental tract, olivary hypertrophy is ipsilateral. If the primary lesion is in the dentate nucleus or cerebellar peduncle, olivary degeneration is contralateral. If the lesion is in both central tegmental tract as well as cerebellar peduncle, bilateral involvement is seen.[1]

The sequence of pathological changes includes no olivary changes, degeneration of olivary amiculum, mild olivary hypertrophy, olivary enlargement, olivary pseudohypertrophy, and olivary atrophy. The changes occur over a period of time ranging from immediately after the primary insult to several years later.[4],[6]

Three radiological changes are seen on MRI--olivary changes in acute stage, increased signal in T2 images within approximately 6 months up to 16 months, and persistence of signal intensity changes on T2 images with resolution of hypertrophy beyond 16 months.[4]

Other causes found in our review were surgical resection of low-grade astrocytoma attached to floor of fourth ventricle, surgical resection of posterior fossa epidermoid cyst, posterior inferior cerebellar infarct as a complication of surgery for trigeminal neuralgia, old pontine infarct, old non-hypertensive cerebellar and

brainstem hemorrhage and post-traumatic hemorrhagic contusion to the superior cerebellar peduncle and midbrain.[1],[2],[3],[4],[5]

The differential diagnosis of hypertrophic olivary degeneration include infarction, demyelination, primary tumor, metastasis and infection.[1]

The treatment options for palatal myoclonus in the literature review included medications such as carbamazepine, clonazepam, valproic acid and/or botox injection to the tensor veli palatine muscle.[1]

The usual presentation of hypertrophic olivary degeneration is palatal myoclonus. Occasionally, opsoclonus is seen in these cases. We present a case of hypertrophic olivary degeneration with ocular bobbing. Furthermore, in our review, we found that pontine cavernomas are not a known cause of hypertrophic olivary degeneration.

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Conflicts of interest

There are no conflicts of interest.

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