HYPERTROPHIC OLIVARY DEGENERATION SECONDARY TO PONTINE CAVERNOMA: A RARE RADIOLOGICAL DIAGNOSIS

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ABSTRACT

Hypertrophic olivary degeneration is caused by a lesion in the triangle of Guillain and Mollaret. It presents clinically as palatal myoclonus, ataxia, hemiparesis and tremor. MRI demonstrates T2 hyper-intensity and hypertrophy of the inferior olivary nucleus. We present a case of hypertrophic olivary degeneration associated with a pontine cavernoma.

Key Words: Hypertrophic olivary degeneration; MRI; Brain; pontine cavernoma.

Introduction

Hypertrophic olivary degeneration is a rare entity that develops after an injury to the dentato-rubro-olivary pathway. Hypertrophic olivary degeneration is due to presumed trans-synaptic degeneration resulting in vacuolation of neurons, an increase in the number of glial cells, demyelination, and shrunken neurons. It is considered to be a unique type of degeneration because it is associated with enlargement rather than theatrophy of the inferior olivary neurons.

Case Report

We report a case of a 45 year old male, hypertensive, non-diabetic, presenting with headache, vertigo and right upper limb rhythmic tremor. The symptoms had appeared over the past 6 months. He had no history of trauma or any neurological intervention. Neurological examination revealed palatal myoclonus on right side. However no motor or sensory deficit was present. No cerebellar signs were found.

MRI evaluation was done which revealed a well-defined lesion at the level ofpons that was iso-to-hypo-intense on T1-weighted images. On T2-weighted images, the lesion showed heterogenous, predominantly hypo-intense lesion in posterior pons involving the left central segmental tract, representing pontine cavernoma.

Figure 1: Axial T2-weighted Fast-spin echo (FSE) image at the level of ponto-mesencephalic junction showing a heterogenous, predominantly hypo-intense lesion in posterior pons involving the left central segmental tract, representing pontine cavernoma.
At level of the inferior olivary nucleus, axial T2-weighted and Fluid Attenuated Inversion recovery (FLAIR) images revealed hyper-intense signal and relative enlargement of the left anterolateral medulla, consistent with left inferior olivary nucleus hypertrophy (Fig. 3, Fig. 4). There was no contrast enhancement or diffusion restriction. No other focal lesion was noted.

Discussion

Hypertrophic olivary degeneration was first reported in 1887 by Oppenheim. In 1926, Foex et al. described a process referred to as “trans-synaptic degeneration” where neurons undergo neuronal loss and reactive gliosis after losing synaptic input from injury to their afferent fibers. The triangle of Guillain and Mollaret consists of a set of connecting tracts including the cerebellar dentate nucleus, the brachium conjunctivum, and the contralateral olive. The efferents from the dentate nucleus ascend through the superior cerebellar peduncle, cross in the decussation of the brachium conjunctivum inferior to the red nucleus, and then descend to the inferior olivary nucleus by way of the central tegmental tract. The triangle is completed by inferior olivary nuclear efferents crossing the midline, entering the inferior cerebellar peduncle, and terminating on the original dentate nucleus.

Hypertrophic olivary degeneration is a unique form of degeneration because it results in enlargement of the affected structure rather than atrophy. The mechanism underlying these unique pathologic changes is still not well understood. What has been clearly demonstrated is that hypertrophic olivary degeneration represents the end-result of a lesion that damages the neuronal connections between the
dentate nucleus of the cerebellum, the red nucleus, and the inferior olivary nucleus. High intensity in the anterolateral part of the medulla has a wide differential diagnosis including infarction, demyelination, tumor, infection and other inflammatory processes. However, limitation of the lesion to inferior olivary nucleus with sparing of surrounding medullary tissues and association with focal olivary enlargement is strongly suggestive of hypertrophic olivary degeneration. Hypertrophic olivary degeneration is almost always unilateral; however, rare bilateral cases have been reported. Lesions in the brainstem involving the central tegmental tract cause ipsilateral hypertrophic olivary degeneration, while lesions in the cerebellum (dentate nucleus and superior cerebellar peduncle) cause contralateral hypertrophic olivary degeneration. Lesions involving both the central tegmental tract and the superior olivary nucleus may cause bilateral hypertrophic olivary degeneration. Infarct, hemorrhage, traumatic brain injury, tumor and surgery for vascular lesions have all been reported as causes of hypertrophic olivary degeneration. MR imaging due to its exquisite contrast sensitivity and lack of bone artifact in the low posterior fossa, is the most sensitive and specific tool for the diagnosis of olivary hypertrophic changes. Birbamer et al described the evolution of olivary changes after an acute lesion onset by assessing MR imaging data obtained in patients. These authors proposed three stages: 1) the acute stage without olivary changes, 2) olivary enlargement and increased signal on T2- and proton density weighted images and 3) disappearance of the hypertrophy with some persistence of increased signal. The diagnosis of hypertrophic olivary degeneration on MR imaging is made by presence of T2-hyperintense non-enhancing olivary lesion in association with another lesion in the contralateral dentate nucleus, contralateral superior cerebellar peduncle, ipsilateral red nucleus or ipsilateral pontine tegmentum. The increased olivary signal on T2-weighted images can appear as early as 1 month post-insult, and persist for at least 3-4 years. Olivary hypertrophy is a later finding, with initial development around 6 months and resolving at around 3-4 years. This resolution appears as a result of olivary atrophy. It has been implicated that the initial hyperintense signal relates to the early phases of gliosis due to demyelination and increased water content. The hypertrophy of olivary nucleus, therefore, is the stage of pathological changes that leads to cell death of both neurons and astrocytes. This process eventually results in atrophy, and olivary shrinkage. Vacuolar degeneration of the cytoplasm occurs at 6-15 months and gliosis follows 15-20 months after the onset of the primary lesion.

Conclusion

Hypertrophic olivary degeneration is a rare trans-synaptic degeneration with a self-limiting course. Only symptomatic treatment is recommended. The T2-hyperintensity may appear as early as 1 month after insult and persist for many years. Olivary nucleus hypertrophy develops around 6 months and resolves by 3-4 years. Radiological imaging plays a crucial role in accurate diagnosis and is essential to avoid misinterpretation.

References


