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A Rare Case Of Bilateral hypertrophic olivary degeneration

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Abstract : Hypertrophic olivary degeneration is a result of a primary lesion damaging the dentorubro- olivary pathway. Causes include primary lesions in contralateral dentate nucleus or ipsilateral central tegmental tract, focal brain stem insults leading to dentatorubral-olivary pathway disruption. Ischemia, demyelination, Hemorrhage, cerebrovascular malformation, or diffuse axonal injury following severe head trauma are most common causes for this degeneration. It is a transynaptic form of degeneration and unique, causing hypertrophy rather than atrophy of the inferior olivary nucleus. Progressive ataxia and palatal tremor(PAPT) is a rare distinct clinical entity characterized by symptomatic palatal tremor associated with progressive ataxia.Only few cases have been described in literature, and MRI findings show, in most cases, cerebellar atrophy and unilateral inferior olivary hypertrophy . We report a case where MRI study at 1.5T demonstrated marked bilateral olivary complex hyperintensity and hypertrophy and marked cerebellar atrophy.

Keyword : Hypertrophic olivary degeneration, dento-rubro-olivary pathway,

INTRODUCTION:

Hypertrophic olivary degeneration (HOD) is a rare form of neuronal degeneration that occurs secondary to any injury that disrupts the afferent fibers to the inferior olive within the dentatorubro- olivary tract (Triangle of Guillain-Mollaret) . Deafferentation causes transsynpatic degeneration and leads to hypertrophy of the inferior olive in a specific timeframe as demonstrated on MR imaging 1,2 . Classically, disruption of the triangle of Guillain and Mollaret results in clinical palatal myoclonus or other dentatorubral tremor. Olivary hypertrophy is not seen immediately after the brainstem insult but typically appears in a delayed fashion, usually within 4–6 months. The pathologic process persists and is frequently visible after 10 months. Clinical symptoms such as abnormal movement rarely improve. Although olivary hypertrophy typically resolves in 10–16 months, olivary hyperintensity on T2-weighted images may persist for years after resolution of the hypertrophy 2,4.

CASE HISTORY:

33 year old male presented with complaints of headache and vertigo since one year duration. He was conscious and had normal orientation**.** He had bilateral mild palatal myoclonus; the palatal arc was normal but he could not get his tongue out. No history suggestive of any fever, upper or lower respiratory tract infection. No history of previous surgery. Patient had a history of severe head injury 2 years back. No history of seizures. The patient is not a known case of diabetes or hypertension. Neurological examination showed bilateral vertical optokinetic nystagmus. Restriction in bilateral horizantal conjugate gaze. Vertical gaze was normal. The patient underwent MRI examination which revealed bilateral inferior olivary nuclei hypertrophy and hyperintenity on T2 weighted images, and mild prominence of superior cerebellar peduncle and diffuse cerebellar atrophy as shown in FIGURE 1.

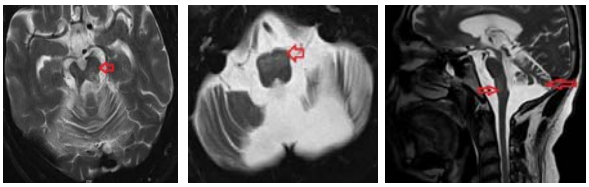

FIGURE 1: MRI T2 axial and sagital image shows bilateral inferior olivary nuclei hypertrophy and hyperintenity on T2 weighted images, and mild prominence of superior cerebellar peduncle and diffuse cerebellar atrophy .

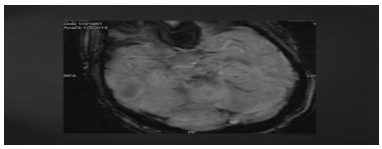

FIGURE 2:SWI shows no evidence of hemorrhage

DISCUSSION:

The dento-rubro-olivary pathway was described by Guillain and Mollaret and is referred to as ‘‘the triangle of Guillain and Mollaret’’(FIGURE 3). The triangle is defined by three anatomical structures: the dentate nucleus in the cerebellum; the contralateral red nucleus (at midbrain level); and the contralateral inferior olivary nucleus (ION) in the medulla. The afferent pathway to the olives originates in the dentate nucleus and travels through the superior cerebellar peduncle to enter the contralateral red nucleus —the dentate rubral tract. It then traverses downwards through the central tegmental tract to connect the red nucleus with the contralateral ION. Efferent

fibres from the ION then cross superiorly through the inferior cerebellar peduncle back to the dentate nucleus — the olivocerebellar tract 3,4.

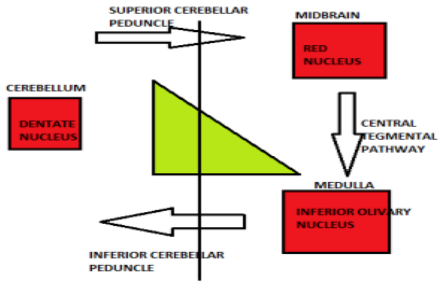

FIGURE 3:The triangle of Guillain and Mollaret

CAUSES:

Include primary lesions in contralateral dentate nucleus or ipsilateral central tegmental tract, focal brain stem insults leading to dentatorubral-olivary pathway disruption. in Olivary enlargement, unusual vacuolar degeneration of cytoplasm occurs which cause hypertrophy and increase in number of astrocytes occur in 6- 15 months. Gliosis follows at 15-20 months. Decreased number of purkinje cells in contralateral cerebellar cortex. Contralateral dentate reduced in size, possibly due to iron depletion secondary to axonal iron transport block. Most common signs/symptoms include symptomatic palatal tremor and myoclonus. Rhythmic involuntary movement of soft palate, uvula, pharynx, and larynx. Severe myoclonus may also affect cervical muscles and diaphragm with or with out dentatorubral tremor 5. All patients who develop palatal myoclonus after brain insult will have hypertrophic olivary degeneration. But bilateral hypertrophic olivary degeneration is rare. If it is associated with bilateral cerebellar atrophy with symptoms of ataxia, restricted horizontal gaze and palatal myoclonus we can consider progressive ataxia and palatal tremor (PAPT) which is a subgroup of the symptomatic palatal tremor (SPT)5,6. Palatal tremor has been subdivided into essential (EPT) and symptomatic palatal tremor (SPT). PAPT may be divided into sporadic and familial forms. Published details of cases of PAPT are sparse . Sporadic PAPT is a subtype of SPT in which progressive cerebellar degeneration is the most symptomatic feature, other symptoms include vertical nystagmus. The cause of sporadic PAPT remains uncertain. Familial PAPT is associated with marked brainstem and cervical cord atrophy with corticospinal tract findings, but the typical olivary MRI abnormalities have not been reported. Sporadic form of progressive ataxia and palatal tremor is a disorder of both the cerebellum and brainstem 7. In our patient there is no specific posterior fossa mass/pontaine hemorrhage(FIGURE 2), except for the symptoms of vertigo, tinnitus. On examination ,he had palatal clonus, mild ataxia and restricted horizontal gaze and thus we evaluated and found to have the possibility of progressive ataxia and palatal tremor syndrome of sporadic type with the MR imaging finding of bilateral hypertrophic olivery degeneration and cerebellar atophy with mild prominence of superior cerebellar peduncle.

CONCLUSION: Hypertrophic olivary degeneration is a rare and potentially confusing imaging finding with unusual histological features causing macroscopic enlargement rather than atrophy of the inferior olive. Palatal myoclonus or other movement disorders may not always occur; however, the possibility of developing Hypertrophic olivary degeneration and its clinical sequelae is a relevant concern for clinicians and patients when surgery is contemplated. Knowledge of the condition and its MR characteristics on the part of the radiologist can prevent erroneous diagnoses of more sinister pathology.

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