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Bilateral Hypertrophic Olivary Degeneration-A Rare and delayed complication following pontine hemmoraghe

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ABSTRACT
Hypertrophic Olivary Degeneration is rare transsynaptic degeneration secondary to lesions involving the anatomical triangle of Guillain and Mollaret or the dento-rubro-olivary pathway. Here we present a case of 37 year male with history of pontine bleed 6 months back with residual left spastic hemiparesis, able to walk without support after 2 months of initial stroke. Now presented with 6 weeks history of severe ataxia of gait and in coordination of limbs with action and intention tremors of extremities. MRI Brain showed presence of bilateral Hypertrophic olivary degeneration. Thus in patients with brainstem strokes with improvement initially followed bydelayed worsening with fresh cerebellar signs and tremors, a possibility of hypertrophic olivary degeneration should be considered. This may help us avoid unnecessary investigations and evaluation.
Keywords: *Hypertrophic olivary degeneration Mollarets Pontine Rubral tremor.*

INTRODUCTION

Hypertrophic olivary degeneration is a unique and rare form of transsynaptic degeneration.

The degeneration is unique because here the degenerating inferior olivary nucleus goes into hypertrophy rather than atrophy.[1] It usually appears after an injury to the dentate-rubro-olivary tract like haemorrhage, Infarction, trauma, neoplasm, and demyelination [2–3]. The Hallmark clinical feature is Palatal Myoclonus followed by other findings like dendatorubral tremors and ocular myoclonus. It occurs after a period of latency ranging from 2-49 months (median duration being 10-11 months) [4]. Here we present our case of Bilateral HOD after a pontine bleed 6 months back and which did not show the hallmark sign of palatal myoclonus.

CASE REPORT

37 year old Hypertensive, Smoker with history of pontine bleed 6 months back with residual left spastic hemiparesis. The patient had gradually improved after the stroke and was able to walk without support after 2 months of initial stroke. Patient now presented with 6 weeks history of gradual onset and progressive ataxia and thereby difficulty of walking. He also complained of new onset tremors involving both upper and lower

limb along with head tremors. It was gradually progressive and because of progressive ataxia and tremors he could not walk with support and became wheel chair bound for past 2 weeks.

On examination he had vertical nystagmus in primary position with severe Resting, postural and action tremors of both upper limbs more towards the left side. Motor Tone showed mild hypertonia involving left upper and lower limb, power was MRC Gr 4+ in left upper and lower limb and normal in the right side .Deep Tendon Reflexes showed brisk reflexes on the left upper and lower limbs equivocal plantar in the left side .He had yes-yes type of head tremor. No palatal myoclonus was seen.

In view of progressive deficits a CT Brain was done to rule out any fresh structural causes which were found to be normal and showed total resolution of the pontine bleed. MRI Brain was done, T2 flair images showed bilateral olivary hyperintensities without any evidence of diffusion restriction or contrast enhancement. GRE sequences showed old hemosiderin deposits in pons. Thus the imaging was suggestive of bilateral hypertrophic olivary degeneration secondary to the old pontine bleed. The cause of delayed worsening could be explained by hypertrophic olivary degeneration which classically presents with gradual onset and progressive cerebellar signs and rubral tremors. The Hallmark finding of palatal myoclonus may not be present initially or may not occur at all in a small proportion of cases.

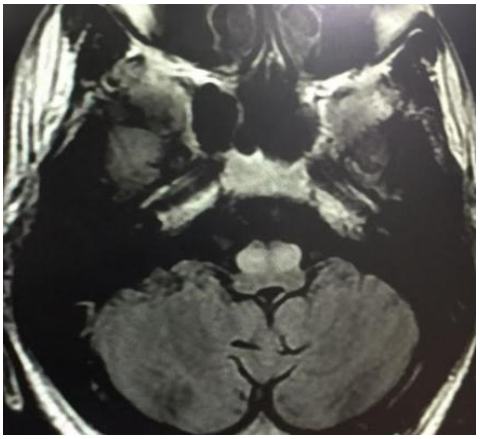

Fig 1 MRI Brain T2 Flair Image Axial View showing bilateral hyperintensities involving Inferior Olivary Nucleus

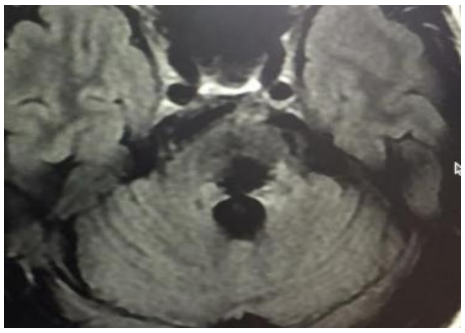

Fig 2 MRI Brain T2 Flair Axial view showing hypodense lesion without any edema or mass effect in pons suggestive of old bleed

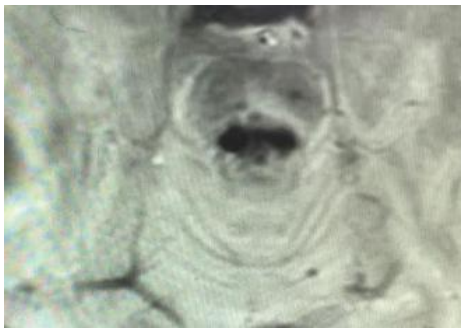

Fig 3 MRI Brain GRE Sequence axial view showing hemosiderin deposits in Pons suggestive of old hemmoraghe.

DISCUSSION

The anatomical triangle of Guillian and Mollaret consists of a central segmental tract connecting red nucleus to inferior olivary nucleus, dentatorubral tract (connecting dentate nucleus to contralateral red nucleus via superior cerebellar peduncle), and inferior cerebellar peduncle connecting inferior olivary nucleus to contralateral cerebellar cortex and dentate nucleus. The spectrum of lesions varies from ischemic infarction, demyelination, and haemorrhage of various etiologies or trauma [3, 5, 6].

Pathologically hypertrophic olivary degeneration is characterized by cell body enlargement along with vacuolation of the cytoplasm, astrocytic hyperplasia and proliferation, demyelination and fibrillary gliosis are also seen.[7]

These histopathological changes are reflected in the typical imaging appearance of hypertrophic olivary degeneration. In the initial stages there is increase in signal on T2 and an increase in size of the olivary nucleus .In acute stage the size is

normal and then hypertrophy sets in around 4-6 months after the initiating pathology and gradually resolves over 3-4 years and olivary atrophy develops.[3,8]. Even though radiological findings may revert, clinical abnormalities may persist.

The differential diagnosis of hyperintensities on T2 weighted images within the pontomedullary region includes tumours, demyelinating lesions, infarction, and inflammatory processes (tuberculosis, sarcoidosis, or encephalitis). Contrast MRI Images with diffusion sequences can help us in ruling out other etiologies. Signal changes confined to the olivary nucleus with or without enlargement, lack of contrast enhancement or diffusion restriction along with presence of an inciting lesion in the brain stem or cerebellum and sometimes contralateral cerebellar atrophy should point toward the diagnosis of HOD. Correct diagnosis of HOD can help in avoiding unnecessary investigations.[9] Disruption of Guillain and Mollaret triangle can result in unilateral, bilateral or contralateral hypertrophy of olivary Nucleus. Central tegmental tract affection causes unilateral involvement, while dentate nucleus or the superior cerebellar peduncle involvement causes contra lateral involvement. Bilateral ION degeneration occurs in paramedian pontine involvement due to simultaneous involvement of the central tegmental tract and the superior cerebellar peduncle [10,11] Our case has been presented as it is clinically relevant as the diagnosis of HOD in this patient explained the worsening of symptoms after 6 months of initial pontine bleed from which the patient had improved significantly .The degeneration was bilateral in our case however in previous reported cases of pontine bleed with HOD ,it was usually unilateral. Our patient also lacked the hallmark finding seen in HOD that is palatal myoclonus and had other features like ataxia, ocular myoclonus and tremors.

CONCLUSION

In patients with history of brainstem lesions, HOD should be kept in mind in patients presenting with new focal neurological symptoms like Ataxia, Rubral tremors and nystagmus even in the absence of palatal myoclonus. Early diagnosis with MRI Brain can help us avoid unnecessary investigations.

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