

Hypertrophic olivary degeneration following cerebral trauma managed with rehabilitation — A case report

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Abstract

Hypertrophic olivary degeneration is a kind of trans-synaptic degeneration, caused by the interruption of dentato rubro olivary pathway. Magnetic resonance imaging (MRI) has been the best modality to show the signals of olivary nucleus hypertrophy. It appears on T2-weighted magnetic resonance imaging as hyper-intensities. Here we present a unique case of a 27-year-old male with traumatic brain injury causing multiple intracranial haemorrhages and functional impairment, which was revealed on computerised tomography. Tracheotomy and thoracic drainage were performed immediately. Anti-infection therapy, brain protection, and comprehensive arousal therapy were part of the intervention along with a comprehensive rehabilitation programme including occupational therapy, balance training, coordination, bed mobility training, and strengthening exercises. A holistic diagnostic approach can reduce the chances of misdiagnosing post-traumatic Hypertrophic Olivary Degeneration cases.

We concluded that a comprehensive physical rehabilitation programme and medical treatment works best to heal the lesion and its resulting traumatic symptoms in cases of Hypertrophic Olivary Degeneration cases.

Keywords: MRI, Neurological rehabilitation, Olivary nucleus, Olivopontocerebellar atrophies.

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Introduction

Disruption of the Guillain-Mollaret triangle (GMT) results in different clinical manifestations, including palatal tremor, dentato-rubral tremors, together with ophthalmic myoclonus.^{1,2} Surgery, haemorrhage, tumour, trauma, inflammation, demyelination, degeneration, and radiation injury together with other factors can damage the dentate nucleus rubro olivary pathway.³

The treatment options for hypertrophic olivary degeneration (HOD) are limited, but there is some evidence that Gabapentin, Memantine, Botulinum toxin

injections, and deep brain stimulation can be used.⁴

In this article, we present the case of a HOD patient who developed head and left upper extremity rhythmic tremors. This is the first case of post-traumatic HOD dealt with a rehabilitation perspective and emphasis on rehabilitation. This case can be used as a guide for a combined medical and physical rehabilitation treatment of post-traumatic HOD.

Publishing this unique case will help other health professionals to reduce chances of misdiagnosing cases of post traumatic HOD, and give them mental stimulation for considering other diagnoses, such as post-traumatic HOD instead of misdiagnosing it as epilepsy and hypertrophy.

Case Report

A 27-year-old man presented to Tongji Hospital(Wuhan, Hubei Province, China) in August 2018, with a history of head trauma due to a traffic accident. Before managing the patient, the family's consent was taken for anonymous publication of this case report.

The patient had no history of hypertension, type 2 diabetes mellitus, hyperlipidaemia, obstructive or sleep apnoea and denied smoking, drinking (wine, alcohol), and using any recreational drugs.

Intracranial haemorrhage and tremors were the main findings and treatment concerns. The patient was directly admitted to our hospital after acquiring the trauma and no intervention was performed earlier, at any other hospital.

The patient's computerised tomography (CT) scan showed multiple intracranial haemorrhages. The MRI performed later, showed multiple old haemorrhagic foci in the bilateral fronto-parietal lobes, lateral ventricle, right temporal and occipital lobe, left thalamus, cerebellum, and brainstem. On detailed reassessment and physical examination, it was observed that the ventral vertebral body of medulla oblongata and inferior olivary nucleus region was shrinking; there was high signal on T2WI, poor overall control of the trunk and head tremors, fine postural tremors in the left upper extremity, lower limb bilateral spasticity and weakness, disconjugate eye movements with horizontal gaze nystagmus, ataxia, slurred speech, dysmetria, spasticity, exaggerated DTRs (deep

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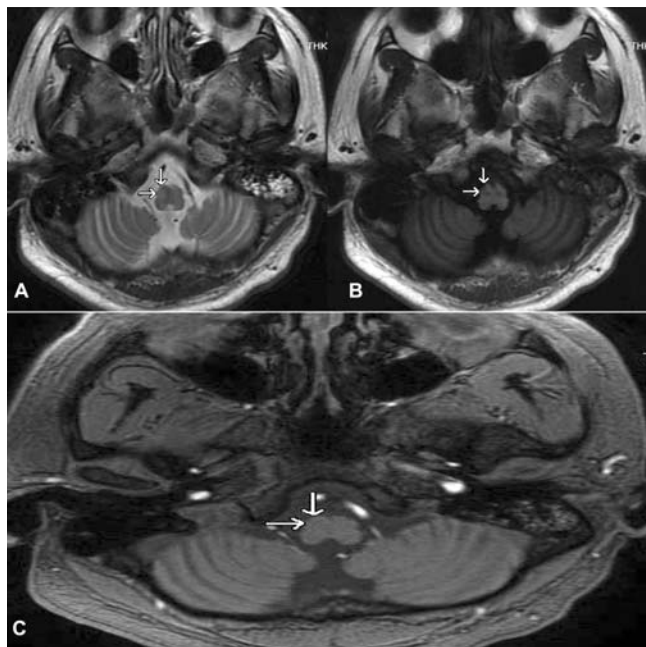


Figure-1: 2018-04-26, Axial T2 image (A) demonstrates the hyperintense signal in an enlarged right inferior olivary nucleus (arrows) that does not enhance (arrows) on post-contrast axial T1 (B). Axial DWI T2 (C) demonstrates no diffusion restriction signal.

tendon reflexes) in the left upper and lower extremity.

MRI of the head and diffusion-weighted imaging (DWI) showed the ventral vertebral body of the medulla oblongata and inferior olivary nucleus region shrinking, with a high signal on T2WI. The neuro-imaging findings and the clinical manifestations supported the diagnosis of HOD and traumatic successor, Wallerian degeneration. On T2 high-intensity non-contrast enhancing lesions, the presence of olivary nucleus enlargement, suggested hypertrophic olivary degeneration as the only diagnosis which explains all the imaging manifestations (Figure-1, 2).

MRI showed persistent HOD together with insensitivity whereas weighted imaging revealed left signal enhancement.

After tracheotomy, closed thoracic drainage was performed. The patient was further treated for dehydration, infection, prevention of epilepsy, and brain protection. Symptomatic supportive care and nutritional medical care were also provided. The patient was admitted to the rehabilitation department for further investigations and treatment. Interestingly, after comprehensive arousal therapy for six months, consisting of auxiliary drainage of phlegm, limbs physiotherapy, median nerve stimulation, facial irritation, physical agents, acupuncture and tuina, extraction of the air pipe, and stomach tube, the patient gradually gained consciousness. He could answer simple

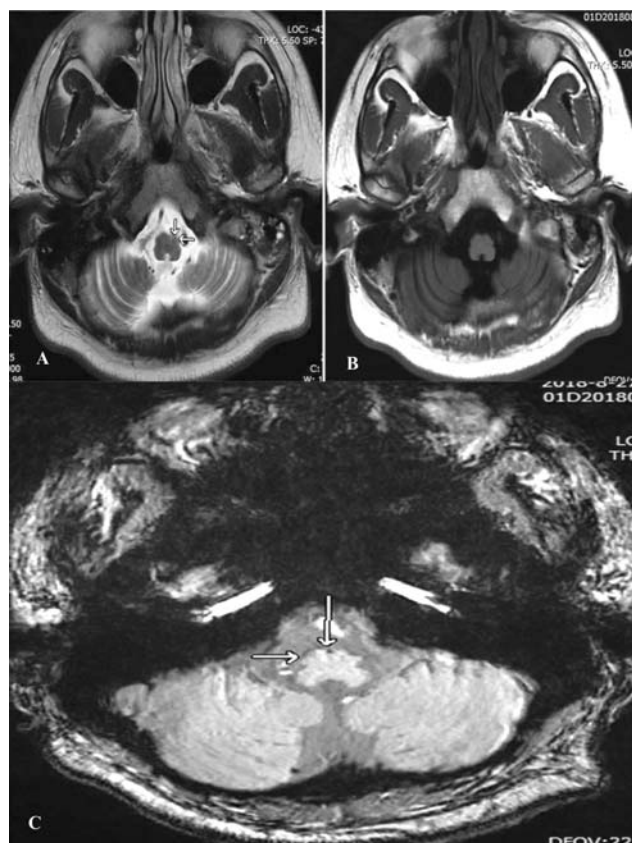


Figure-2: 2018-08-27 Axial T2 image (A) demonstrates the hyperintense signal in an enlarged left inferior olivary nucleus (arrows) that doesn't enhance (arrows) on post-contrast axial T1 (B). Axial SWI T2 (C) demonstrate signal increase.

questions, and grip actively by the left upper limb. On the mini-mental state examination (MMSE) scale, used to assess cognitive function, the patient scored 19. His MMSE score showed moderate dementia.

He was reassessed seven months after the trauma as he presented with worsening of functional movements. Examination showed poor trunk control, left upper extremity tremors, bilateral upper and lower extremity weakness, spasticity, visual and movement coordination difficulties.

The oral medications included Gabapentin, Nimodipine, Baclofen, Benzoisol, and Aidibenzoquinone. A 45-minute, 5 days/week rehabilitation programme was designed for four months, that included bed mobility training, upper extremity fine motor skills training, strengthening exercises, tone/spasticity reducing exercises, and exercises for nystagmus, coordination exercises, and balance training. After four months of systematic rehabilitation, the patient showed good sitting balance, bed mobility, and improvement in upper limb functional activities.

Discussion

Anatomically speaking, the dentato-rubro-olivary pathway, or the triangle of Guillain and Mollaret, consists of the ipsilateral red nucleus of the midbrain, the contralateral dentate nucleus of the cerebellum, and the ipsilateral inferior olivary nucleus of the medulla.⁵

HOD always occurs with the damage of dentate nucleus-red nucleus pathway or the tegmental central tract pathway instead of the pathway from the inferior olivary nucleus to the contralateral cerebellum.⁶

According to the location, together with the extent of infarction in the caudal midbrain, the clinical manifestations may be different. The symptoms on initial presentation are mainly bilateral; the reason is that the caudal collateral branch of the midbrain has the descending cerebellar tract, the central tegmental tract, and other important white matter bundles passing through the midline, which makes it hard for inexperienced eyes to locate the lesions in one vascular area.^{6,7}

The differential diagnosis of high signal intensity on T2 weighted imaging in pontine medullary area includes tumour, demyelinating disease, infarction, or inflammatory diseases (encephalitis, tuberculosis, sarcoidosis). However, the lack of contrast ensures the absence of tumours or the origin of infection, while the extra enlargement of the olivary nucleus is conducive to the chronic stage of infarction or multiple sclerosis. Therefore, in the background of T2 high-intensity non-contrast-enhanced lesions with olivary nucleus enlargement, hypertrophic olivary degeneration remains the only diagnosis to explain all imaging manifestations.⁸

HOD is currently lacking an ideal treatment for tremors. Most patients are alleviated by use drugs as Carbamazepine, Clonazepam, Diazepam, Levodopa, Benzene sea somatotherapy and others. The presented patient, in the past three months had mostly been lying or sitting and with no rehabilitation training leading to a declining physical strength. With no exercise for a long duration, his muscle strength and endurance of both the lower limbs had declined. To overcome these problems, drug treatment and systematic rehabilitation treatment was initiated. The principle "step by step" was followed to strengthen basic exercise training. Through the combination of drug treatment and intensive basic exercise training, tremors of the whole body were relieved.

Therefore, with the reduction of tremors, the left limb muscle strength increased from 3 to 4, and the right proximal limb

increased from 1 to 3. His sitting balance improved up to 2 levels and standing balance was on level one¹. The patient's daily living ability also significantly improved, and the Barthel index score increased from 10 points before treatment to 45 points. The etiology was intra-parenchymal pontine haemorrhage secondary to trauma and not hypertonia or epilepsy, which was very frequently encountered in the rehabilitation department. Thus, it is important to explore and recognise the HOD symptoms, particularly in young patients, to avoid misdiagnosis of this benign condition.⁹

Conclusion

This case shows neurological deterioration occurs after cerebellum and brainstem haemorrhage secondary to head trauma with interval development of HOD during recovery. Comprehensive rehabilitation along with medical treatment works best to improve tremors, spasticity, strength, and functional movements in patients with HOD.

Disclaimer: This case report has not been submitted to any other journal.

Conflict of Interest: The authors certify that there is no conflict of interest with any financial organisation regarding the material discussed in the manuscript.

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