

## ONE AND A HALF SYNDROME (OAHS) - A CASE REPORT

Shruti K. Nair<sup>1</sup>, L. Subha<sup>2</sup>, K. Mohan Raj<sup>3</sup>

<sup>1</sup>Postgraduate Student, Department of Ophthalmology, Sree Balaji Medical College and Hospital, Chromepet, Chennai.

<sup>2</sup>Associate Professor, Department of Ophthalmology, Sree Balaji Medical College and Hospital, Chromepet, Chennai.

<sup>3</sup>Professor and HOD, Department of Ophthalmology, Sree Balaji Medical College and Hospital, Chromepet, Chennai.

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### PRESENTATION OF CASE

It is characterised by gaze palsy in ipsilateral side and internuclear ophthalmoplegia on gaze to contralateral side. This case is reported for its rarity. A 65-year-old male reported to our hospital with complaints of double vision for 5 days. He is a known case of diabetes and hypertension for 10 years. On clinical examination, anterior segment was normal. Fundus was normal in both eyes. Vertical eye movements were normal. He had exotropia in right eye. Left eye horizontal movements were absent. Only abduction with nystagmus was present in right eye. MRI showed an exophytic haemorrhagic lesion measuring 2.4 x 1.7 cm size involving pons. In this case, left horizontal gaze palsy is due to parapontine reticular formation lesion and restriction of adduction in left eye was due to internuclear ophthalmoplegia from lesion in medial longitudinal fasciculus. Apart from vertical movements, only abduction was possible in right eye. Hence, this case was diagnosed as one-and-a-half syndrome (OAHS).

Parapontine Reticular Formation (PPRF) and Medial Longitudinal Fasciculus (MLF) combined lesions on the same side characterised by-

- Ipsilateral gaze palsy.
- Ipsilateral restriction of adduction (internuclear ophthalmoplegia (INO)).
- Presence of abduction in contralateral eye with nystagmus.
- Normal vertical movements.

Main causes of this rare syndrome are stroke and multiple sclerosis. Other causes include tumours, AV malformations, basilar artery aneurysms and rarely vasculitis, brainstem tuberculoma and neurocysticercosis.

A 65-year-old male came to our outpatient department. He was apparently alright when he developed binocular diplopia since 5 days. He denied any history of weakness of any part of the body, paraesthesias or numbness of limbs or face, urinary incontinence, deafness, tinnitus or any slurring

of speech. He was a nonsmoker, diabetic and hypertensive for past 10 years.

On admission, his pulse was regular, his blood pressure was 130/80 mmHg. The ocular position of left eye on forward gaze was fixed at the midline, while the right eye was abducted. For horizontal ocular movements, only the right eye could abduct with monocular horizontal nystagmus. Also, there was conjugate gaze palsy to left indicating horizontal left One And a Half Syndrome (OAHS), i.e. on leftward gaze, neither eye could pass the midline and on rightward gaze, horizontal jerky nystagmus in the abducting right eye was noticed. Vertical and rotatory nystagmus was absent. Vertical eye movements were preserved. Visual acuity was 6/24 with pin hole 6/12. Pupils 3 mm and normally acting. Fundus was within normal limits.

MRI of the brain was taken, which revealed an exophytic haemorrhagic lesion measuring 2.4 x 1.7 cm noted involving the pons.

### DIFFERENTIAL DIAGNOSES

1. **Internuclear Ophthalmoplegia (INO)**- Restriction of adduction in ipsilateral eye and nystagmus in contralateral eye on abduction, but in one-and-a-half syndrome. There is also restriction of adduction in contralateral eye and abduction in ipsilateral eye. In both, vertical movements and convergence are normal.
2. **Oculomotor Nerve Palsy**- Restriction of adduction, vertical movements and convergence are affected.

### CLINICAL DIAGNOSIS

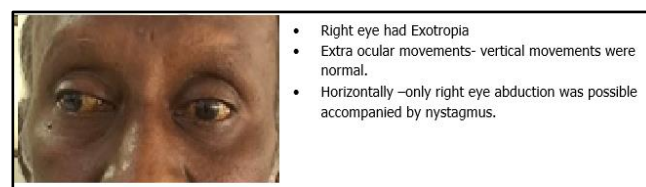


Figure 1

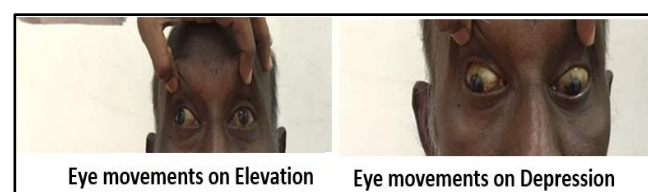


Figure 2

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Corresponding Author:

Dr. K. Mohan Raj,

Professor and HOD, Department of Ophthalmology,

Sree Balaji Medical College and Hospital,

Chromepet, Chennai- 600044.

E-mail: mohanrajdr@yahoo.com

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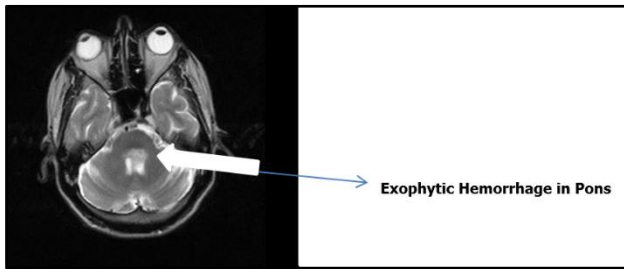


Figure 3

**PATHOLOGICAL DISCUSSION**

Supranuclear ocular movements comprise chiefly vertical and horizontal movements. Horizontal movements are controlled by the subcortical centres located mainly at the pontine level and vertical movements at the level of the rostral midbrain.

The final common pathway for all types of horizontal gaze movements is the Abducens Nucleus (AN)- extensive experimental data from cats and monkeys<sup>1,2,3</sup> suggest that the AN contains two types of cells, i.e. motoneurons, which innervate the ipsilateral external rectus muscle (magnocellular motor neurons) and excitatory internuclear neurons that decussate at the level of the nucleus, ascend in the contralateral MLF and terminate at the medial rectus subdivision of the oculomotor nuclear complex. The gaze motor command is prepared in the specialised areas of the reticular formation of the brainstem, which receives a variety of supranuclear inputs from different areas. The main premotor region for horizontal gaze is Paramedian Pontine Reticular Formation (PPRF), which is located on each side of the midline in the central paramedian part of the tegmentum extending from the pontomedullary junction to the pontipeduncular junction.

Complex forms of nystagmus and conjugate eye movement deficits are generated if the pontine paramedian structures are damaged. The combination of one sided horizontal gaze palsy and ipsilateral internuclear ophthalmoplegia known as one-and-a-half syndrome is most commonly caused by demyelination, vascular disease or tumours. Other known cause include arteriovenous malformation, basilar artery aneurysms and rarely vasculitis. Brainstem tuberculoma as a cause of one-and-a-half syndrome has been reported only twice before with complete resolution of symptoms after antitubercular treatment.<sup>4</sup> The one-and-a-half syndrome is usually due to a single unilateral lesion of the parapontine reticular formation and the abducens nucleus on one side (causing conjugate gaze palsy) with interruption of internuclear fibres of the ipsilateral medial longitudinal fasciculus after it has crossed midline from its origin in the contralateral abducens nucleus (causing failure of adduction of the ipsilateral eye).

1. MLF lesion (between ipsilateral 3<sup>rd</sup> and contra lateral 6<sup>th</sup> nerve).

2. Parapontine Reticular Formation (PPRF) to 6<sup>th</sup> nerve nucleus.
3. 6<sup>th</sup> nerve nucleus (maybe affected).

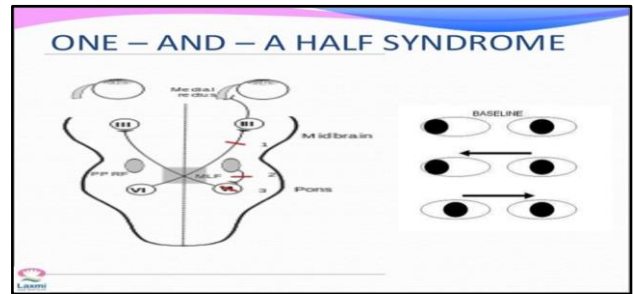


Figure 4

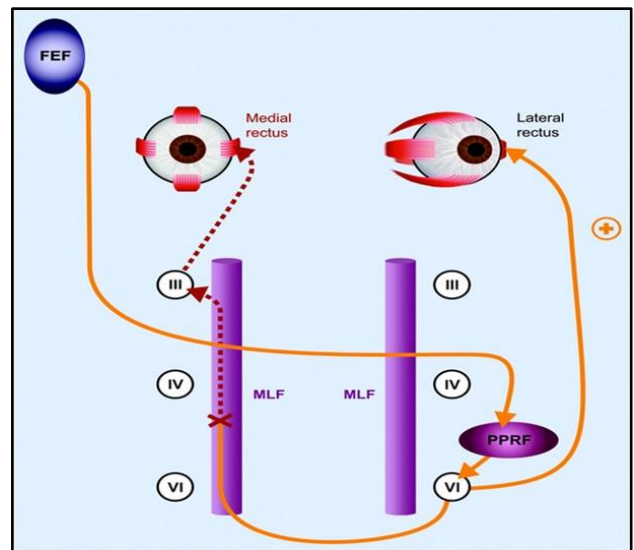


Figure 5. Showing Internuclear Ophthalmoplegia (Left Eye)

- Lesion is in MLF (which connects 3<sup>rd</sup> nerve with the contralateral 6<sup>th</sup> nerve).
- The affected eye shows impairment of adduction when an attempt is made to gaze contralaterally.

Convergence maybe normal.

**Internuclear Ophthalmoplegia (INO)**- The INO is characterised by paresis or paralysis of the ipsilateral eye adduction- on attempted horizontal gaze to the contralateral side- and horizontal jerk nystagmus in the contralateral abducting eye. Typically, convergence is preserved if the lesion does not extend to the mesencephalon. Bilateral lesions cause bilateral defects in adduction and nystagmus in the abducting eye. In addition, there are nystagmus on upward gaze as well as bilateral partial ptosis. Lesions of the MLF do not cause paralysis of conjugate horizontal or vertical gaze.<sup>5</sup>

Wall M et al<sup>6</sup> studied 20 cases of one-and-a-half syndrome and reported 14 to have multiple sclerosis. De Seze J et al<sup>7</sup> examined the clinicoradiological correlations in six patients with one-and-a-half syndrome due to stroke. Out of these 6 patients, 4 had associated facial nerve palsy, 3 had hemiparesis and 1 had unilateral hemihypoesthesia. In

all patients, MRI revealed infarcts in the pons. Cause of infarct was a basilar artery dissection in 1 patient, bilateral vertebral artery dissection in a second and unknown in the other four. This study showed a good correlation between the site of the lesion and clinical deficits. Satoshi Katoaka et al<sup>8</sup> analysed the clinical signs and their association with MRI findings in 49 patients with acute paramedian pontine infarcts to clarify the clinicotopographical correlation and their prognosis. Twenty-seven patients had basal infarcts, fifteen patients basal tegmental infarcts and seven patients had tegmental infarcts. The study revealed that paramedian pontine infarcts, which are usually due to thrombosis of perforating arteries presented with faciobrachial dominant hemiparesis with dysarthria, somatosensory disturbances and horizontal gaze abnormalities including abducens nerve palsy, Internuclear Ophthalmoplegia (INO), horizontal gaze palsy and one-and-a-half syndrome.

Pontine haemorrhages represent approximately 5% of the cases of intracranial haemorrhage.<sup>9</sup> Basal tegmental variety produces the classic picture of coma, quadriplegia, decerebrate posturing, horizontal ophthalmoplegia, ocular bobbing, pinpoint reactive pupils, abnormalities of respiratory rhythm and preterminal hyperthermia.

In our case, since the patient reported earlier, he did not develop other systemic features. In our patient, MRI showed an exophytic haemorrhagic lesion measuring 2.4 x 1.7 cm noted involving the pons.

Clinical examination findings correlate with the interruption of function limited to ipsilateral (right) parapontine reticular formation and medial longitudinal fasciculus in the pons resulting in ipsilateral horizontal gaze palsy and internuclear ophthalmoplegia. The involvement of abducens nerve nucleus caused failure of the adduction of the ipsilateral (right eye) with exotropia. Thus, one-and-a-half syndrome has also been called Paralytic Pontine Exotropia (PPE)- the presence of exotropia in OAHS was first observed by Fisher.<sup>10</sup> In the acute phase, the exotropic eye shows abduction nystagmus during attempts to move it further laterally and there is extreme slowness of adduction when the eye moved to the midline. The other eye remains straight ahead, unable to move right or left. PPE is attributed to an acute brainstem lesion involving the PPRF and the MLF on the side opposite the deviated eye.<sup>11</sup>

## DISCUSSION OF MANAGEMENT

Patient was given injection methylcobalamin for 3 days followed by oral medications for 10 days. He is on regular neurology follow up.

## FINAL DIAGNOSIS

ONE-Left horizontal gaze palsy (PPRF lesion).

HALF-INO (restriction of adduction in left eye-lesion is in MLF) Tonic abduction of right eye.

Hence, this case is diagnosed as one-and-a-half syndrome.

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