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A Case Report of Rare Syndrome-One and a Half Syndrome

Amulya P[1], Sumanth Tangudu[2], Tanu Kashyap[3]

[1]Senior Resident, Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bengaluru, Dr.M.G.R. Educational and Research Institute, Chennai, India

[2, 3]Junior Resident, Department of Ophthalmology, Rajarajeswari Medical College and Hospital, Bengaluru, Dr.M.G.R. Educational and Research Institute, Chennai, India

Abstract: ***This case report describes a 52 year old male who presented with acute headache, giddiness, vomiting, and diplopia, later identified as one and a half syndrome linked to a pontine hemorrhage in the setting of poorly controlled hypertension. Ocular findings showed horizontal gaze palsy with preserved vertical movements and convergence, supported by neuroimaging that revealed chronic infarcts with superimposed brainstem hemorrhage and vascular variation on angiography. The clinical course highlights how a unilateral pontine lesion affecting the paramedian pontine reticular formation, abducens nucleus, and medial longitudinal fasciculus produces the classic eye movement pattern seen in this syndrome. Early blood pressure control led to gradual improvement in symptoms, underscoring the role of timely recognition and focused management in cerebrovascular causes of complex gaze disorders.***

Keywords: One and a half syndrome, pontine hemorrhage, conjugate gaze palsy, hypertensive stroke, diplopia

1. Case Report

A 52-year-old male presented to our hospital's emergency department with complaints of headache, giddiness, vomiting, and diplopia for the past three days. He was apparently in good health until three days ago when he initially developed headache and giddiness, which were later followed by non-projectile vomiting and diplopia. The patient had a seizure episode two years ago, which was not accompanied by paresis, palsy, paraesthesia, deafness, disturbed micturition, mouth deviation, or slurred speech. He is an occasional smoker and consumes alcohol. He has a history of hypertension for four years managed with irregular treatment. There is no history of diabetes.

On examination, the patient's blood pressure was 180/135 mmHg, which was stabilized using intravenous mannitol (100 mL) and lasix (10 mg), followed by oral antihypertensive medications. Ocular examination revealed that the right eye was abducted (15° exotropia), while the left eye remained at the midline. During extraocular movement assessment, the right eye could abduct with end-point nystagmus, but the left eye showed no horizontal movements. Vertical movements were preserved, indicating conjugate gaze palsy consistent with one-and-a-half syndrome. Visual acuity was 6/60 in both eyes, improving to 6/18 with a pinhole. Anterior segment examination revealed nuclear sclerosis grade 2 with posterior subcapsular changes. Fundus examination showed grade 3 hypertensive retinopathy.

NCCT of the brain revealed a large chronic infarct in the right basal ganglia, along with chronic lacunar infarcts in the left basal ganglia, bilateral thalami, and left hemipons. Additionally, small intraparenchymal hemorrhages were noted in the brainstem and left thalamus.

MR angiography of the brain demonstrated a hypoplastic A1 segment of the right anterior cerebral artery.

2. Clinical Diagnosis

Figure 1: Right eye shows 15degree exotropia on hirschberg test

Figure 2: Horizontally – only right eye abduction preserved

Figure 3: Eye movements on depression

Figure 4: Eye movements on elevation

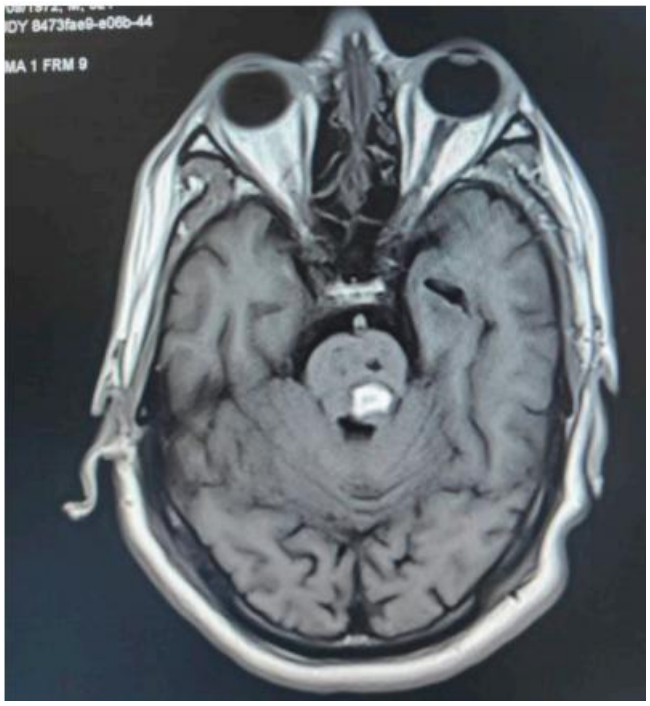

Figure 5: NCCT BRAIN showing hemorrhage in brain stem

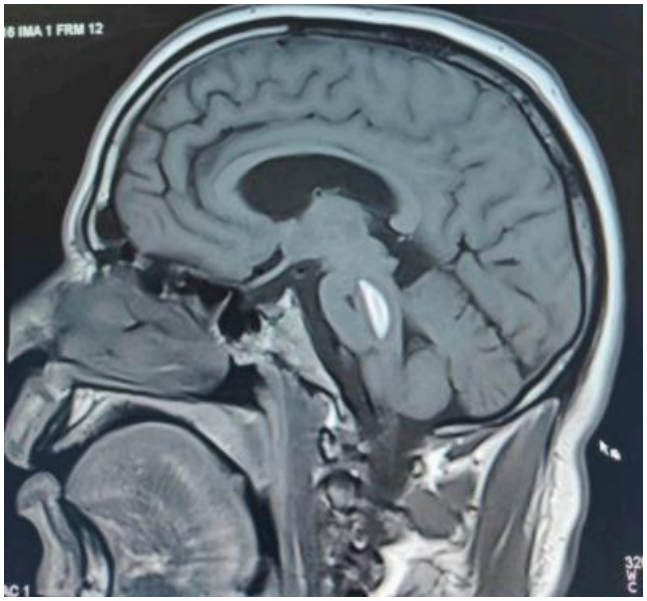

Figure 6: NCCT BRAIN showing hemorrhage in brain stem

Pathological Discussion

One-and-a-half syndrome results from a lesion in the unilateral tegmentum of the pons or a dorsal pontine lesion, which damages the paramedian pontine reticular formation (PPRF), abducens nucleus, and medial longitudinal fasciculus (MLF). Cerebrovascular events, such as pontine hemorrhages, brainstem infarcts, and basilar artery aneurysms, are the most common causes of this syndrome. Other causes include multiple sclerosis,[3] neurocysticercosis, brainstem encephalitis, trauma, brainstem malignancies, and arteriovenous malformations[4][5][6].

Patients with one-and-a-half syndrome typically present with complaints of diplopia, blurred vision, and gaze-evoked nystagmus. In this case, a hemorrhage in the hemipons affected the ipsilateral medial longitudinal fasciculus (MLF), paramedian pontine reticular formation (PPRF), and abducens nucleus, leading to the development of the syndrome[6]. This resulted in one eye remaining in a fixed position without horizontal movement, while the other eye remained abducted.

When horizontal extraocular movements are tested, the affected eye does not cross the midline, while vertical movements remain unaffected. Since the motor fibers and nuclei of the third cranial nerve are intact, convergence is preserved. Supranuclear ocular movements primarily involve vertical and horizontal movements, with horizontal movements controlled by subcortical centers at the pontine level and vertical movements by centers in the rostral midbrain. In this syndrome, the preservation of normal vertical eye movements and convergence indicates that the motor fibers and nuclei of the third cranial nerve are unaffected.

The degree and duration of horizontal gaze impairment caused by unilateral brain lesions above the level of the oculomotor nucleus depend on the lesion's size and location. The paramedian pontine reticular formation (PPRF) and the abducens nucleus serve as the final pre-nuclear and nuclear centers for horizontal eye movements, integrating nerve impulses from the frontal and parietal visual areas. [6] The

abducens nucleus projects two sets of horizontal gaze fibers: one controlling the ipsilateral lateral rectus muscle and the other activating the contralateral medial longitudinal fasciculus (MLF) [6] . The MLF transmits internuclear fibers that travel to the midbrain, where they end in the medial rectus area of the oculomotor nuclear complex.

OAHS is characterized by a combination of ipsilateral horizontal gaze palsy and ipsilateral internuclear ophthalmoplegia (INO). The horizontal gaze palsy in OAHS can result from:

1) Damage to both the ipsilateral abducens nucleus and the paramedian pontine reticular formation (PPRF).
2) Damage to either the ipsilateral abducens nucleus or the PPRF alone[6].
3) Damage to the ipsilateral abducens nerve root fibers combined with damage to the contralateral medial longitudinal fasciculus (MLF) in cases involving two distinct lesions.[6]

In this case, the diagnosis of one-and-a-half syndrome (OAHS) was established based on clinical and radiological findings. OAHS typically results from a single unilateral lesion involving the paramedian pontine reticular formation (PPRF) and the abducens nucleus on one side, leading to conjugate gaze palsy. Additionally, there is disruption of the internuclear fibers of the ipsilateral medial longitudinal fasciculus (MLF) after they cross the midline from their origin in the contralateral abducens nucleus, causing failure of adduction in the ipsilateral eye.

The key components involved in OAHS include:

1) Lesion of the MLF (between the ipsilateral third nerve and contralateral sixth nerve).
2) Damage to the PPRF affecting the sixth nerve nucleus.
3) Potential involvement of the sixth nerve nucleus itself.

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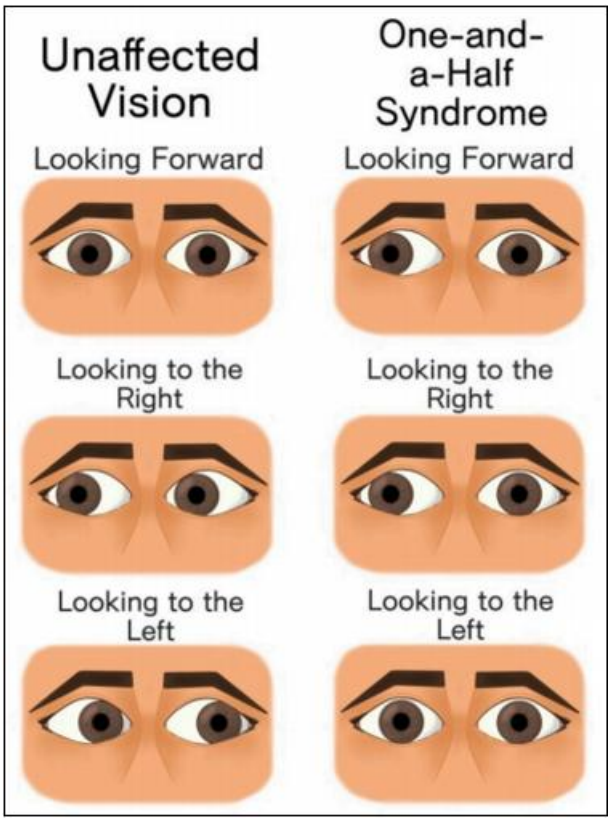

Figure 7

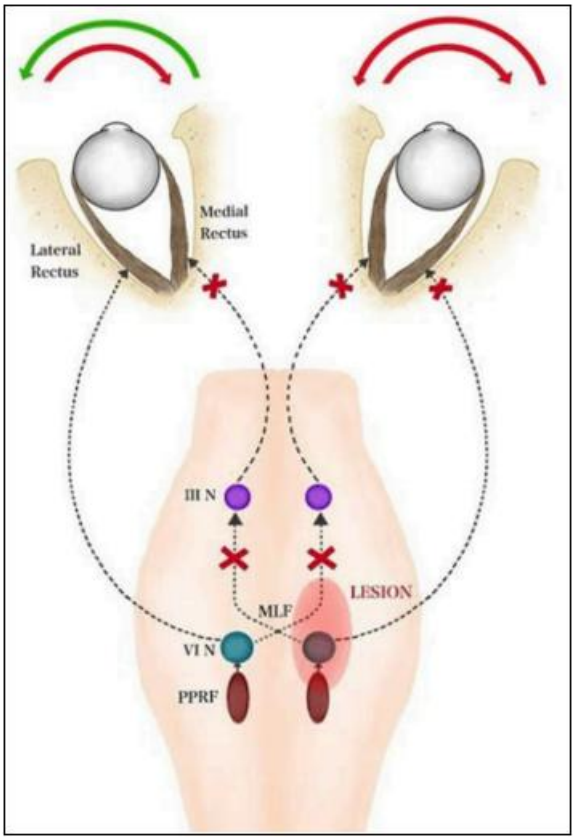

Figure 8

Key components affected include:

1) The MLF lesion (between the ipsilateral third nerve and the contralateral sixth nerve).
2) The PPRF to the sixth nerve nucleus.
3) The sixth nerve nucleus itself (which may also be involved).

The lesion in the MLF disrupts the connection between the third nerve and the contralateral sixth nerve, contributing to the clinical presentation.

The affected eye demonstrates impaired adduction when attempting to gaze contralaterally, while convergence may remain normal.[7][8]

Diagnosis and Management

The case was diagnosed as one-and-a-half syndrome secondary to a hemorrhage in the hemipons. Immediate management involved controlling blood pressure with intravenous mannitol, injection lasix, and oral antihypertensive medications. The patient was regularly followed up, and after four days, there was slight improvement in diplopia and extraocular movements.

3. Conclusion

The diagnosis of one-and-a-half syndrome (OAHS) is clinical while radiological findings confirm pons involvement. Early diagnosis and active management of cerebrovascular accidents can result in a favorable prognosis for one-and-a-half syndrome. Regular medications, consistent blood pressure monitoring, a healthy diet, identification of risk factors, and timely hospitalization play crucial roles in improving outcomes in cerebrovascular accidents and one-and-a-half syndrome.

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