

One and a Half Syndrome: A Manifestation of Right Pontine Hemorrhage

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ABSTRACT

One-and-a-half syndrome is a syndrome of horizontal movement disorder of both eyes. It presents as a combination of conjugate horizontal gaze palsy in one eye (one) and internuclear ophthalmoplegia (INO) of the contralateral eye (half). Our case is interesting as this syndrome is one of the most localizing brainstem syndromes. Being a rare entity with low prevalence worldwide, this case is the first to be witnessed at our institute with one and a half syndrome being the presenting symptom of a right pontine infarct with hemorrhagic transformation.

Keywords: Internuclear ophthalmoplegia, One-and-a-half syndrome, Hemorrhage

INTRODUCTION

One and a half syndrome encompasses gaze palsy in one direction with an INO on horizontal gaze in the opposite direction, hence on attempted horizontal gaze, only abduction of the contralateral eye is present. On neutral gaze, the contralateral eye is slightly abducted (so-called paralytic pontine exotropia). Convergence is spared. This syndrome is caused by affection of paramedian pontine reticular formation (PPRF) and/or abducens nucleus and medial longitudinal fasciculus (MLF) on the same side [1]. Usually it is of vascular origin and it can be a sign of lateral pontine hemorrhage [2]. INO is a gaze abnormality characterized by impaired horizontal eye movements with weak adduction of the affected eye, and abduction nystagmus of the contralateral eye resulting from a lesion in the MLF in the dorsomedial brainstem tegmentum of either the pons or the midbrain [1]. The most common cause of one and a half syndrome in older patients is ischemic infarction, risk factors for the latter is advanced age, hypertension, diabetes and smoking.

CASE REPORT

A 57 year old female with a past medical history of long standing hypertension- not compliant with medications- and anemia, presented to the emergency department for one day history of diplopia associated with headache, nausea and vomiting. The patient's initial blood pressure was 240/140 mm Hg. The patient woke up with left sided eye deviation and diplopia. She denied aphasia, dysarthria and mentioned the Emergency Medicine Service (EMS) noted left sided facial droop. In physical exam, in neutral gaze, the right eye globe was central in position, the left

eye was abducted. On horizontal gaze examination, the left eye adducts to the midline only and not further and abducts with no nystagmus, while the right eye can neither adduct nor abduct. Brain CT showed small right pontine hemorrhage (**Figure 1**). The patient was started on nicardipine drip and admitted to the Medicine Intensive Care Unit for further management with the diagnosis of one and half syndrome status post hypertensive right pontine hemorrhage.

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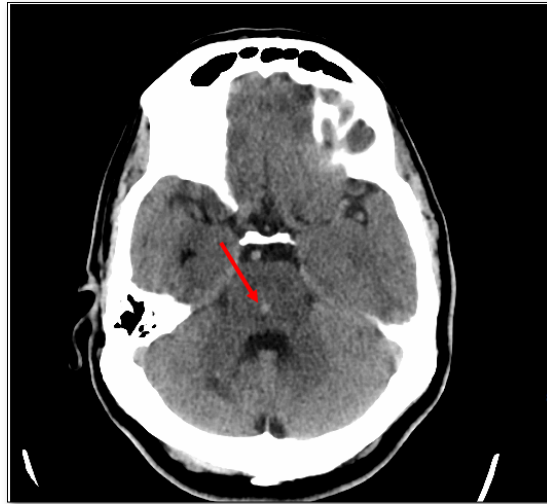


Figure 1. Axial CT scan of the brain showing a tiny focus of pontine hemorrhage (red arrow).

Repeated brain CT scan in 24 h later showed stable changes. Neurology was consulted and recommended MRI of the brain which showed acute to early sub-acute right paramedian pontine lacunar infarct with small amount of acute hemorrhage at the anterior margin (**Figure 2**). The patient remained neurologically stable and was transferred to the floors where she reported

feeling well. She reported persisting double vision in her left eye, an eye patch was prescribed. The patient was discharged home with a follow up visit scheduled at Neurology clinic where she showed improvement, she had only mild dysconjugacy on nearly normal horizontal Extraocular muscle (EOM).

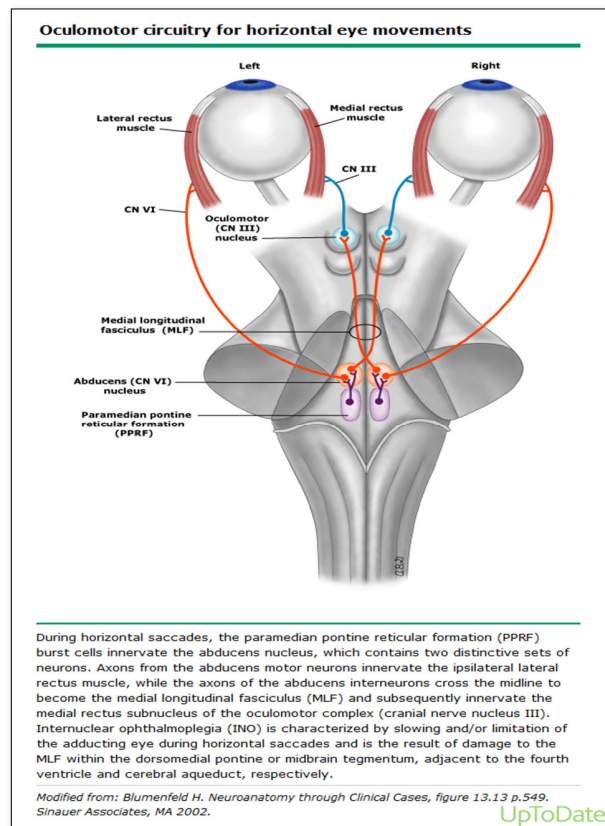


Figure 2. Illustrating the circuitry for horizontal eye movements.

DISCUSSION

The paramedian pontine reticular formation (PPRF) is the conjugate gaze center for horizontal eye movements. During horizontal eye movement, the PPRF cells innervate the abducens nucleus, which contains two distinctive sets of neurons: a) Axons of the abducens motor neurons directly innervate the ipsilateral lateral rectus muscle. b) Axons of the abducens interneurons cross the midline to become the Medial Longitudinal Fasciculus (MLF) and subsequently innervate the medial rectus sub nucleus of the oculomotor complex. Motor neurons from this sub nucleus innervate the medial rectus muscle (ipsilateral to the medial rectus sub nucleus and contralateral to the abducens nucleus) [3,4].

The MLF exists as a pair of white matter fiber tracts that lie near the midline just under the fourth ventricle and cerebral aqueduct and extend through the dorsomedial

pontine and midbrain tegmentum (**Figure 3**). The MLF is involved in coordinating synchronous horizontal eye movements. Because of their close physical proximity near the midline, bilateral injury is common [2,3]. Through the MLF, the actions of the oculomotor and the abducens nuclei are coordinated, generating conjugate horizontal eye movements. The disorders of the horizontal eye movement that are caused by brainstem lesions are classified into three groups: a) lateral gaze palsy; b) INO; and c) one and a half syndrome. The pathologic lesions depicted on magnetic resonance images were topographically well correlated with the brainstem pathways and each type of horizontal eye movement disorder. Most of the lesions were tiny acute infarctions and were found in the most posterior region of the pons, which corresponded to the location of the brainstem pathways [5].

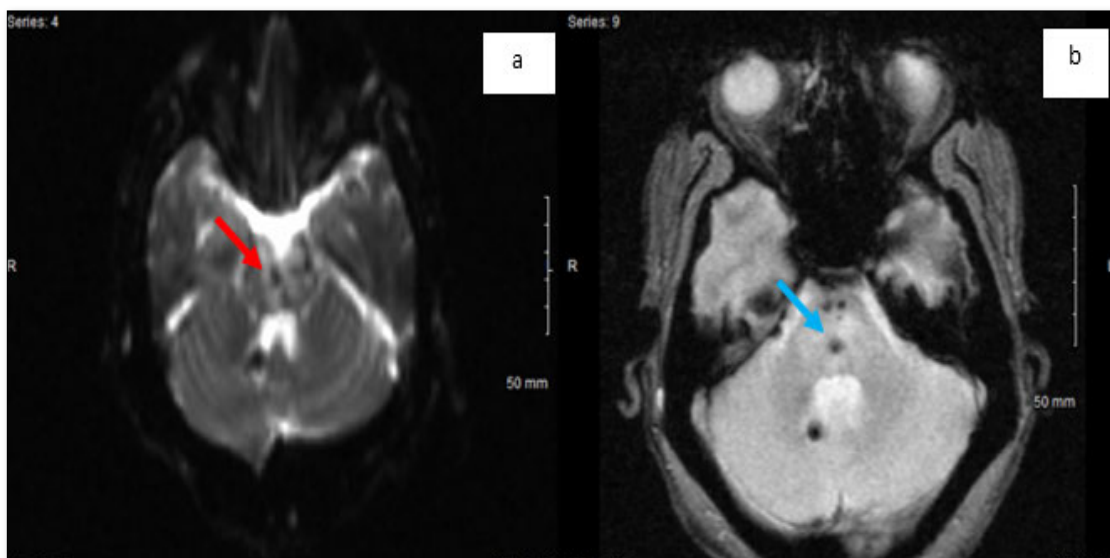


Figure 3. MRI of the brain. a) Diffusion weighted image of an axial brain cut showing right pontine diffusion restriction (red arrow). b) Susceptibility weighted image on an axial brain acute showing the right pontine paramedian focus of hemorrhage (blue arrow).

An INO results from injury to the MLF. The side of the INO is named by the side of the adduction deficit, which is ipsilateral to the MLF lesion. In our case, being the left eye with incomplete adduction deficit due to damage of the left MLF. Damage to the right PPRF will cause bilateral conjugate eye paralysis to the left. Our case is interesting since the patient has an additional affection of the right abducens nerve resulting in right eye abduction paralysis. Along with clinical examination, patients who present with an INO require brain magnetic resonance imaging (MRI). The deficits associated with INO often resolve over a few to several months. Patients may be treated with patching of one eye for symptomatic relief of diplopia.

CONCLUSION

One and a half syndrome consists of gaze palsy in one direction with an INO on horizontal gaze in the opposite direction. With attempted horizontal gaze, only abduction of the contralateral eye remains. Convergence is spared. In primary (neutral gaze), the contralateral eye is slightly abducted (so-called paralytic pontine exotropia). This syndrome is produced by damage to the paramedian pontine reticular formation (PPRF) and/or abducens nucleus and MLF on the same side, all of which are affected in our case owing to pontine infarction with reperfusion hemorrhage. Awareness of the brainstem pathways controlling horizontal eye movement is important to avoid missing a small pontine lesion [5].

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