"Nine" syndrome: A new neuro-ophthalmologic syndrome: Report of two cases

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Abstract

"Eight-and-a-half" syndrome is a rare condition involving the ipsilateral abducens nucleus or paramedian pontine reticular formation (PPRF), the ipsilateral medial longitudinal fasciculus (MLF), and the adjacent facial colliculus/facial nerve fascicle. The condition is often caused by a lesion (vascular or demyelinating) in the dorsal tegmentum of the caudal pons. There are new variants of this syndrome caused by extension of lesion to involve new adjacent structures in pontine tegmentum. We report two patients with different etiology presenting with clinical features suggestive of eight-and-a-half syndrome associated with hemiataxia representing "nine" syndrome (8½ + ½ = 9) adding new dimension to "eight-and-a-half" syndrome.

Key Words

Demyelination, eight-and-a-half syndrome, hemorrhage, pons, medial longitudinal fasciculus, paramedian pontine reticular formation, tegmentum

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Introduction

Pontine tegmental lesions usually present with gaze palsies, internuclear ophthalmoplegia (INO), nystagmus, and abducens palsy.[1] A combination of horizontal gaze palsy in one eye and INO in the other eye caused by a lesion in medial longitudinal fasciculus (MLF) or paramedian pontine reticular formation (PPRF) was first described by Freeman *et al*., in 1943.[2] Subsequently, C Miller Fisher in 1967 coined the term one-and-a-half syndrome to describe this combination.[3] The combination of one-and-a-half syndrome and ipsilateral cranial nerve seventh palsy is known aseight-and-a-half syndrome (1½+ 7 = 8½) and was described by Eggenbergerin 1998.[4] Here, we present two patients who presented with clinical features suggestive of eight-and-a-half syndrome along with hemiataxia. This combination represents probable "nine" syndrome.

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Case Reports

Case 1

A 62-year-old lady presented with giddiness, vomiting, gait unsteadiness of 10 days duration. Symptoms were sudden in onset and persistent. Giddiness was in the form of spinning sensation of the head, present continuously with no postural variation with vomiting. She used to sway on left side while walking. No history of fever, motor weakness, truncal imbalance, speech disturbance, or sensory disturbance in limbs. She was hypertensive on medication. On examination, she was alert, fully oriented. Pupils were equal and reactive to light. Oculomotor abnormalities were noted in the form of left horizontal gaze palsy (left eye abduction and right eye adduction) and limitation of adduction in left eye with abducting nystagmus of right eye (suggestive of left INO). Vertical eye movements and convergence were preserved. Horizontal and vertical vestibuloocular reflex (VOR) was preserved. Left lower motor neuron facial palsy was observed [Figure 1]. Rest of the cranial nerves were normal. Motor and sensory examination was normal. She had impaired left finger-nose and knee-heel coordination with swaying to left on tandem walking [Video 1]. Magnetic resonance imaging (MRI) brain showed hyper intensity in left half of tegmentum of the caudal pons, left dorsolateral rostral medulla, and inferior cerebellar peduncle on fluid-attenuated inversion recovery (FLAIR) sequence with diffusion-weighted imaging (DWI)/ apparent diffusion coefficient (ADC) showing no restriction. Magnetic resonance angiography (MRA) brain was normal [Figure 2]. Serum vasculitis profile and antineuromyelitis optica

(NMO) antibodies were negative. There was mildly raised protein (68 mg/dl) with no pleocytosis on lumbar puncture cerebrospinal fluid (CSF) examination. CSF oligoclonal band was negative. She received intravenous methylprednisolone 1 g for 5 days followed by tapering dose of oral Wysolone. She made slow but remarkable improvement in her symptoms over next 6 weeks. Repeat brain MRI showed reduction in the size of lesion in the pons.

Case 2

A 54-year-old male presented with headache with decrease in the level of consciousness of 1-day duration. Headache was sudden in onset, holocranial associated with projectile vomiting. He lapsed into altered state of consciousness and was brought to hospital. No seizures, fever, or weakness of limbs. He was known hypertensive with poor drug compliance. On examination, his blood pressure (BP) was 220/120 mmHg, he was drowsy but arousable on verbal commands. Fundus examination revealed blurring of disc margins. There was no lateralizing deficit on the day of admission. There was improvement in his level of sensorium as he became conscious the next day with reduction in the BP. Oculomotor abnormalities were noted in the form of right horizontal gaze palsy (right eye abduction and left eye adduction) and limitation of adduction in right eye with abducting nystagmus of left eye (suggestive of right INO). Vertical eye movements and convergence were preserved. Horizontal and vertical VOR was preserved. Right lower motor neuron facial palsy was observed [Figure 3]. Rest of the cranial nerves were normal. Motor and sensory examination was normal. He had impaired left finger-nose and knee-heel coordination with swaying to left on standing [Video 2]. Computed tomography (CT) brain showed bleed in the right caudal pontine tegmentum with extension into right basis pontis and midbrain tegmentum [Figure 4]. He was treated with antihypertensives and antiedema measures. There was no further worsening at the time of discharge.

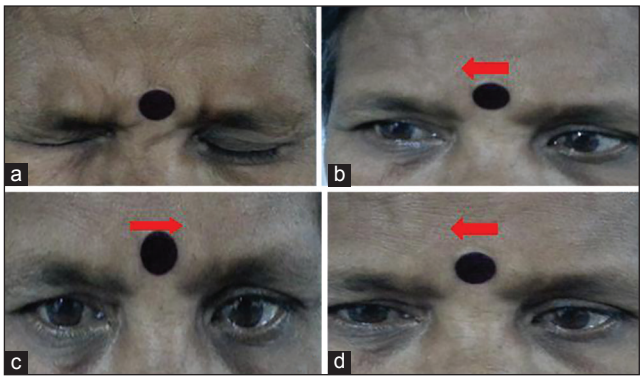

Figure 1: (a) Weakness of left eye closure; (b) Right eye abduction nystagmus; (c) Left horizontal gaze palsy (impaired left eye abduction and right eye adduction); and (d) Right eye adduction paresis

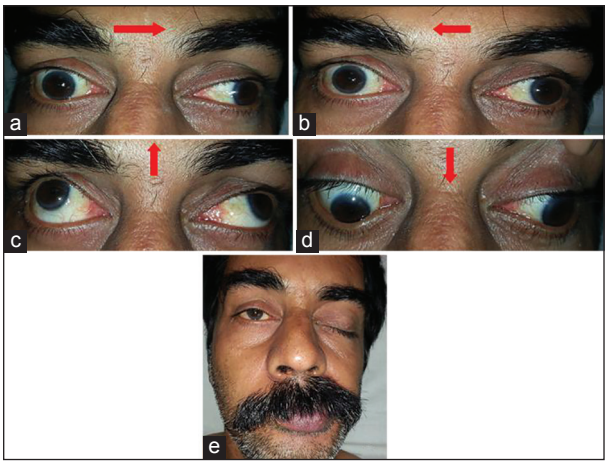

Figure 3: (a) Horizontal gaze to left shows restriction of right eye adduction and left eye abducting nystagmus; (b) Horizontal gaze to right shows restriction of right eye abduction and left eye adduction; (c and d) Vertical up and down movements are preserved; and (e) Right lower motor neuron facial palsy

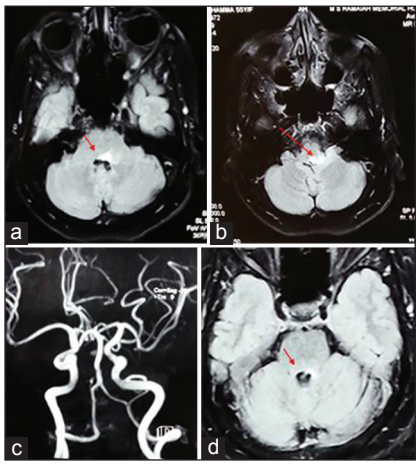

Figure 2: (a and b) MRI brain FLAIR sequence axial view showing hyperintense lesion in caudal left paramedian pontine tegmentum and left rostral dorso-lateral medulla involving inferior cerebellar peduncle (red arrow) respectively; (c) MRA brain showing no abnormality; (d) FLAIR sequence axial view showing resolution of lesion in pontine tegmentum. MRI = Magnetic resonance imaging, FLAIR = Fluid-attenuated inversion recovery, MRA = Magnetic resonance angiography

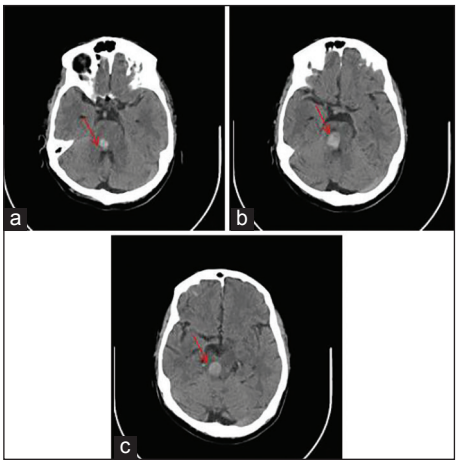

Figure 4: Bleed in right caudal pontine tegmentum (red arrow); (a) Extension into right basis pontis; (b) And right midbrain tegmentum (c)

Discussion

One-and-a-half syndrome represents ipsilateral conjugate horizontal gaze palsy (one) due to a lesion in the abducens nucleus or horizontal gaze center in the PPRF and an ipsilateral INO (half) due to a lesion in the MLF.[3] In brief, there is complete ipsilateral horizontal gaze palsy and partial contralateral horizontal gaze paresis (abduction preserved). Additional involvement of intraaxial fasciculus of the facial nerve results in palsy of cranial nerve VII and this along with one-and-a-half syndrome constitute the eight-and-a-half syndrome.[4] The anatomical localization is in the ipsilateral dorsal tegmentum of the caudal pons in one-and-a-half and eight-and-a-half syndrome. The prime etiologies for both syndromes are brain stem infarcts/hemorrhage, multiple sclerosis/brainstem demyelination,[5] brain stem tumors, and arteriovenous malformations.[6] There are few new variants of eight-and-a-half syndrome. An elderly patient with acute dorsal pontine infarction developed bilateral horizontal gaze palsy due to involvement of bilateral abducens nucleus/PPRF with unilateral peripheral facial paralysis due to anterior extension of the vascular lesion reaching facial nerve fascicle.[6] Lee *et al*., (2007) described a new variant in an elderlypatient with bilateral facial palsy and complete loss of vertical saccades and pursuit with bilateral horizontal gaze palsy.[7] A case of eight-and-a-half syndrome, combined with ipsilateral vertical gaze palsy was reported by Marquart *et al*., (2013) due to involvement of midbrain reticular formation apart from dorsal pontine tegmentum.[8] A possible "nine" syndrome was described by Rosini *et al*., (2013) which comprised of eight-and-a-half syndrome with hemiparesis and hemihypesthesia due to additional involvement of corticospinal tract and medial lemniscus by lacunar pontine infarction.[9]

In the present case series, the first patient had clinical features suggestive of left eight-and-a-half syndrome due to caudal pontine tegmental demyelinating lesion with additional involvement of inferior cerebellar peduncle in left rostral dorsolateral medulla responsible for left hemiataxia. Whereas, the second patient had clinical features suggestive of right eight-and-a-half syndrome due to bleed in right caudal pontine tegmentum with extension into midbrain tegmentum/red nucleus responsible for contralesional hemiataxia. These features further widen the spectrum of eight-and-a-half syndrome, configuring a possible "nine" syndrome.

Conclusion

Apart from the classical eight-and-a-half syndrome described in the literature, there are few new variants of this syndrome that has been frequently reported of late. They represent the extension of lesion either anteriorly in the pontine tegmentum or inferiorly to rostral medulla or superiorly to midbrain as depicted in the present cases. Recognition of this syndrome helps in localization of the lesion to ipsilateral lower pontine tegmentum.

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