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CORRESPONDENCE

Vertical and horizontal one and a half syndrome with ipsiversive ocular tilt reaction in unilateral rostral mesencephalic infarct: A rare entity

Supranuclear ocular movements comprise mainly vertical and horizontal movements. Vertical movements are controlled by the centres located mainly at the rostral midbrain and horizontal movements at the level of the pons.[1] Pontine tegmental lesions usually present with gaze palsies, internuclear ophthalmoplegia (INO), abducens palsy and one and a half syndrome. Usually, one and a half syndrome is produced by a unilateral caudal pontine tegmental lesion that includes the paramedian pontine reticular formation (PPRF) and medial longitudinal fasciculus (MLF) on the same side causing horizontal gaze palsy in one eye and INO in the other eye.[2] Similarly, vertical one and a half syndrome has also been described. The literature on co-existence of horizontal and vertical one and a half syndrome is few. The co-existence of horizontal and vertical one and a half syndrome with ocular tilt reaction (OTR) has not been reported so far. Here, we report a patient who presented with left horizontal one and a half syndrome along with bilateral conjugate upgaze palsy and right downward palsy suggestive of vertical one and a half syndrome and left ocular tilt reaction. Magnetic resonance imaging (MRI) brain revealed infarct in left rostral midbrain with sparing of pons.

A 64-year-old male was brought with history of giddiness, weakness of right upper limb and double vision on looking to the left of 7 days duration. The symptoms were sudden in onset and persistent. Giddiness was in the form of spinning sensation of the head, present continuously with no postural variation. He had impaired dexterity of right upper limb. There was no history of facial weakness, dysphagia and dysarthria, sensory disturbance in limbs or unsteadiness in gait. He was hypertensive and diabetic on medication. General physical examination and other system examination were unremarkable. His blood pressure was 146/88 mm Hg. On examination, he was alert, fully orientated. Higher mental functions were normal. Pupils were symmetric (3 mm) and reactive to light. There was slight head tilt to the left and incomplete left eye ptosis. Oculomotor abnormalities were noted in the form of hypertropia of right eye with incyclotorsion and left eye hypotropia with excyclotorsion suggestive of skew deviation and ocular torsion, left horizontal, conjugate 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) indicating left horizontal one and a half syndrome. Diplopia on abduction of the right eye was seen. For vertical ocular movement, only the left eye could gaze downward. There was bilateral conjugate upgaze palsy and difficulty in downward gaze of the right eye indicating right vertical one and a half syndrome. Eyes were unable to converge (Figure 1). Doll's eye response (oculocephalic reflex) was present. Rest of the cranial nerves were normal. There was pronator drift in right upper limb. Right plantar response was extensor. Remainder of examination was normal. Complete hemogram, renal, hepatic and thyroid functions were normal. MRI brain showed infarct in left rostral midbrain involving left cerebral peduncle (Figure 2a-c). MR angiography of intracranial arteries was normal (Figure 2d). Two-dimensional (2D) echocardiography showed concentric left ventricular hypertrophy. Serum homocysteine levels were high. He was treated with anti-platelets and statins. The symptoms remained the same at the time of discharge.

Horizontal 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 paramedian pontine reticular formation (PPRF) and an ipsilateral internuclear ophthalmoplegia (half) due to a lesion in the medial longitudinal fasciculus (MLF).[2] The most common causes are brain stem infarcts or haemorrhage, multiple sclerosis or brainstem demyelination, brain stem tumours, and arteriovenous malformations. In the present case, left horizontal one-and half syndrome was due to involvement of left MLF at rostral mesencephalic level causing left eye adduction deficit and right eye abducting nystagmus. Left horizontal gaze palsy (right eye adduction and left abduction deficit) can be explained due to involvement of descending fibres from right frontal eye fields to left PPRF and abducens nucleus decussating at rostral midbrain (after decussation).[3] The involvement of left intraaxial third nerve fascicles may be responsible for incomplete left eye ptosis.

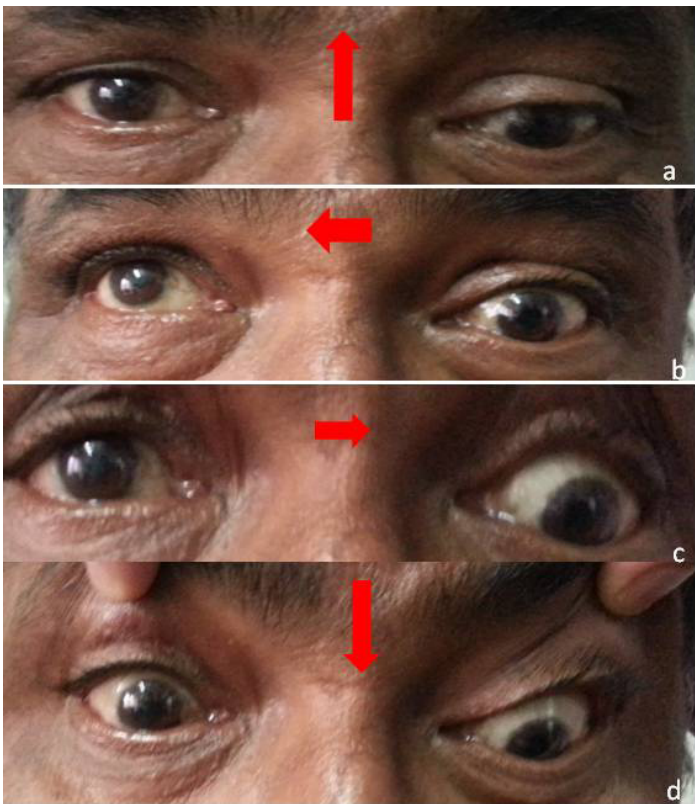

Figure 1 (a & d). For vertical movement, only the downward gaze of the left eye is possible. Downward palsy of the right eye and bilateral conjugated upward palsy were seen; (b & c) For horizontal movements, left eye abduction and adduction were restricted with right eye adduction restriction and abducting nystagmus.

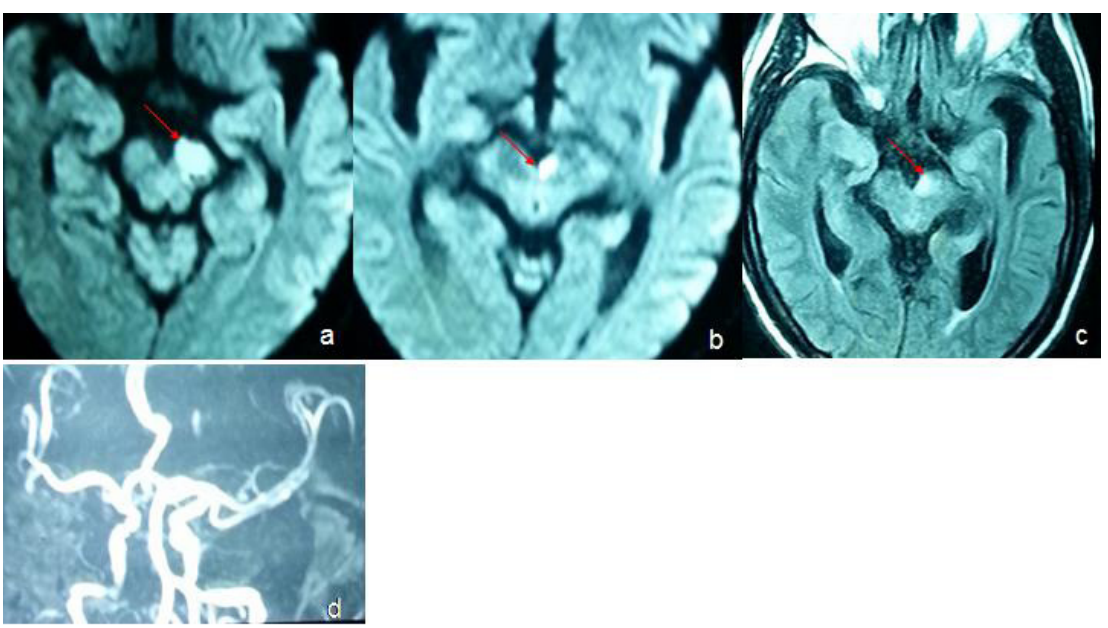

Figure 2. MRI brain diffusion-weighted imaging (DWI) axial view shows hyperintense signal change in left cerebral peduncle and left rostral midbrain (1-c). MR angiography of intracranial arteries was normal (2d).

The rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF), interstitial nucleus of Cajal (INC), and posterior commissure (PC), all located in the tegmentum of the midbrain are the brainstem centres for vertical eye movement.[1] Vertical one and a half syndrome consists of either bilateral conjugate upgaze palsy and a unilateral downward palsy[4] or a bilateral conjugate downward palsy and monocular upgaze palsy.[5] Brain MRI showed unilateral left rostral mesencephalic infarct involving left riMLF and INC. It can explain bilateral supranuclear upgaze palsy (fibres mediating upgaze traverses through opposite riMLF before innervating bilateral elevator muscles). So, unilateral lesion in riMLF caused bilateral supranuclear upgaze palsy. Fibres mediating downgaze innervating ipsilateral downgaze muscles traverse through opposite riMLF. In our patient, fibres from right riMLF innervating right depressor muscles were affected when it traversed through left riMLF. Left riMLF lesion explains the vertical one and half syndrome in our patient. A patient reported by Deleu *et al*. with vertical one and a half syndrome had bilateral riMLF lesions causing bilateral conjugate downward palsy.[5] Terao *et al*. reported a rare occurrence of co-existing vertical and horizontal one and a half syndrome in a patient with infarcts in the right medial thalamus, left dorsal portion of the upper midbrain, and left upper cerebellum. Their patient had bilateral conjugate upgaze palsy and a right unilateral downward palsy suggestive of vertical one and a half syndrome with left horizontal one and a half syndrome. Patient also had left OTR due to cerebellar involvement. There was no lesion in the pontine tegmentum.[3] Our patient had similar presentation of bilateral conjugate upgaze palsy and a right unilateral downward palsy with left horizontal one and a half syndrome and ipsiversive OTR with infarct in left rostral mesencephalon.

Left ocular tilt reaction (OTR) in our patient was due to involvement of left MLF that interrupted the graviceptive pathways from right vestibular nucleus to left INC and riMLF. Lesions of INC usually causes contraversive OTR (contralesional eye hypotropia and ipsilesional eye hypertropia).[6] However, in our patient, OTR was ipsiversive .

OTR constitutes a triad of skew deviation, ocular torsion, and head tilt. Unilateral deficit of otolith input or a unilateral lesion in the graviceptive pathways from the vestibular nuclei to the INC is responsible for OTR. The graviceptive pathway crosses at the caudal pons. The OTR is ipsiversive if lesion occurs below the decussation, as in peripheral or pontomedullary lesions indicating the involvement of the medial and/or superior vestibular nuclei. Here, the ipsilesional eye is on a lower plane with head tilt and ocular torsion toward the side of lesion in ipsiversive OTR. OTR is contraversive if lesion occurs above the decussation as in unilateral pontomesencephalic brainstem lesions above the decussation, indicating the involvement of the MLF or INC and riMLF. The ipsilesional eye is on a higher plane, with head tilt and ocular torsion away from the side of lesion in contraversive OTR.[7]

The present patient represents a rare case of the coexistence of two distinct syndromes, vertical and horizontal one and a half syndromes along with ipsiversive OTR. The exact anatomical and physiological mechanism underlying vertical gaze still remains obscure. Like horizontal one and a half syndrome, vertical one and a half syndrome represents an important neurological signs suggestive of a lesion affecting the rostral midbrain.

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