

Isolated Eight and Half Syndrome, a Rare Neuro-ophthalmic Presentation: A Case Report

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Abstract:

Eight and half syndrome is one of the rare neurological disorders, which is usually overlooked in our clinical practice. Here I report 58 years-old hypertensive Indian gentleman presented with horizontal gaze palsy and facial weakness.

Key Words: Brainstem lesion, Eight and Half Syndrome, Horizontal Gaze palsy, Ischemic Stroke

Isolated brain stem syndromes are rare clinical presentations which are tricky to identify. One and half syndrome with facial nerve palsy together represents eight and half syndrome.

2. Case Description:

A right-handed 58-year-old hypertensive non diabetic gentleman from Darjeeling presented in outpatient clinic with sudden onset of dizziness and visual obscuration for last 3 and half months. His family members noticed facial drooping in left side and drooling of saliva. He developed double vision, food regurgitation more on taking liquids. He then noticed less movement of his left eyeball as compared to other side. He was consulted with ophthalmologist and ENT specialist in local hospital but no clue was identified. He had been diagnosed with hypertension on irregular medication since last one year. He denied limb weakness on either side or no alteration in sensation. His bowel and bladder habits were normal. On examination his vitals were within normal limit. He had normal higher mental function with normal motor power in all four

limbs. Conjugate horizontal gaze palsy in left eye was evident. Conjugate medial horizontal gaze palsy (**Figure 1**) and conjugate lateral horizontal gaze palsy were present (**Figure 2**).



Figure 1(A): Conjugate medial horizontal gaze palsy



Figure 1(B): Conjugate lateral horizontal gaze palsy was present



Figure 2: Lower motor neuron type facial nerve palsy in the left side

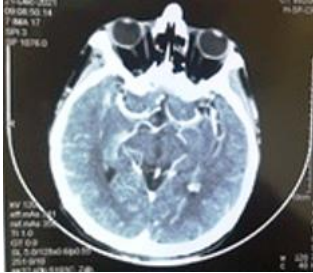


Figure 3: Right paramedian pontine tegmentum (dorsal Pons) chronic infarction

There was lower motor neuron type facial nerve palsy in the left side. Vertical eye movements were normally present. There was no history of decrease hearing, facial numbness, and imbalance or gait abnormality. He had no headache vomiting. He was urgently sent for contrast CT head (**Figure 3**) which showed marked hypodensity in right paramedian pontine tegmentum (dorsal Pons) suggestive of chronic pontine infarction giving a picture of one and half plus seven syndrome.

3. Discussion

Isolated brain stem syndromes are very tricky to identify clinically. One and half syndrome with facial nerve involvement together represents eight and half syndrome. One and half syndrome is characterized by conjugate horizontal gaze palsy in one direction with internuclear ophthalmoplegia (INO).¹ INO is characterized by horizontal gaze abnormality with weak adduction of affected eye and presence of abduction nystagmus of contralateral eye. INO is caused by the lesion in medial longitudinal fasciculus (MLF).² It receives neuronal signals from the contralateral paramedian pontine reticular formation (PPRF) and sends signals back to ipsilateral oculomotor nerve for conducting conjugate eye movement.³ PPRF is also known as Para Abducens nucleus that involves for

coordinating horizontal eye movement and saccades.⁴ When the lesion affects both MLF and PPRF it represents one and half syndrome. One represents PPRF lesion and half represents MLF lesion with INO giving rise to ipsilateral conjugate horizontal gaze palsy. When lesion affects PPRF, MLF and fascicles of 7th CN, it signifies eight and half syndrome. Abducens nucleus is located in the facial colliculus, adjacent to the internal genu of the facial nerve. Lateral rectus motor neurons come together with internuclear neurons and send their axons across to opposite MLF and to the contralateral oculomotor nerve. Abducens nerve exits the ventral Pons near the midline.⁵ When genu of facial nerve fibers that loops around the Abducens nerve nucleus at the level of paramedian pontine tegmentum is involved, lower motor neuron type ipsilateral facial palsy is detected clinically.⁶ CT head findings clearly corresponds the typical neuro-ophthalmic features of my patient. Neuro-ophthalmic entities are usually caused by brain stem ischemic stroke (vascular), demyelination disease like multiple sclerosis.⁷ Some space occupying lesions like tuberculoma, cavernoma, and tumors have also been reported representing one and half or eight and half syndrome.⁸

Conclusion:

Neurological knowledge and understanding is essential for precise localization and diagnosis of brain stem lesion.

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