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Case Report

Eight-and-a Half Syndrome: A Rare Presentation of Gaze Palsy in Ischemic Stroke.

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Abstract:

Eight- and-a- Half –syndrome is a rare clinical manifestation of stroke, where involvement of ocular motor movement manifested by horizontal gaze palsy to one direction, internuclearophthalmoplegia in the other and ipsilateral lower motor neuron seventh cranial nerve palsy. We report a case of eight-a- half syndrome. A 55 years old female known to have Type 2 Diabetes Mellitus, ischemic heart disease presented with double vision, dizziness, deviation of the mouth to the right for 2 days. Her vision was normal, no symptoms of raised intracranial pressure, no limb weakness, slurring of speech. At presentation, patient had poorly controlled diabetes. There were left eye limitation in abduction with contralateral right abducting eye nystagmus, left eye limited abduction consistent with right one and half syndrome. There was also left lower motor neuron facial nerve (7th) palsy. Eight –and-a –half syndrome is a combination of ipsilateral one-and-half syndrome and lower motor neuron facial(7th) nerve palsy. Brainstem conjugate gaze palsy is an important clinical finding, help in diagnosis even a small pontine lesion.

Keywords: Internuclearophthalmoplegia, one and a half syndrome, eight and a half syndrome.

Introduction

Horizontal gaze palsy in one eye and internuclearophthalmolplegia in the other eye, combination of both was first described by Freeman et al, in 1943.[1] Later, C Miller fisher introduced the term as one and a half syndrome.[2] The syndrome was more commonly found in stroke, but rarely in demyelinating and neoplastic lesion.This syndrome is produced by involving the medial longitudinal fasciculus (MLF) and paramedian pontine reticular formation. In 1998 Eggenberger came out with the term eight-and-a half syndrome, a combination one and a half syndrome and ipsilateral lower motor neuron type facial nerve palsy.[3] Although it is a rare syndrome, help the neurologist or any clinician to localize the lesion. Herein, we report a patient with Eight and a Half syndrome.

Case Report

A 55years old female presented with double vision, dizziness, deviation of the mouth to the right for 2 days. It was sudden in onset and non progressive. She was known case of diabetes and ischemic heart disease for 2 years and had regular follow up and medication. Patient gave no history of trauma, she was afebrile, no history of visual disturbance and ocular surgery.

On general examination, she was well and alert, conscious, oriented about time, place, person with Glasgow Coma Scale 15/15, blood pressure 120/80, random blood glucose 19.1 mmol/l and afebrile. Neurological examination revealed left horizontal paresis (figure-1) and limitation of left eye adduction with abducting nystagmus of the right eye (left internuclearophthalmoplegia) (figure-2). There was also left lower motor neuron type seventh cranial nerve palsy characterized by deviation of the angle of the mouth to the right and loss of left nasolabial fold. (figure-5) Bell's reflexes was prominent. Vertical gaze was intact. (figure-3 and 4) Horizontal and vertical vestibulo-ocular reflexes were intact. There were no cerebellar signs, motor and sensory function of upper and lower limbs were normal.Other cranial nerves were intact. Ocular examinations were unremarkable. Computed tomography of brain showed right frontoparietal lobe as well as both thalamic infarct. Magnetic resonance imaging of brain revealed hyperintensity in the right pontine tegmentum just anterior to the fourth ventricle on diffusion-weighted imaging sequence suggestive of an acute ischemic stroke. Patient was treated with antiplatelet.

Figure-1 : Left horizontal gaze paresis (Patient can't move the eyeball to the left)

Figure-2 : Internuclearophthalmoplegia on the right gaze shown by limitation of adduction on left eye and adducting nystagmus on the right eye.

Figure-3 : Intact down gaze

Figure-4 : Intact upgaze

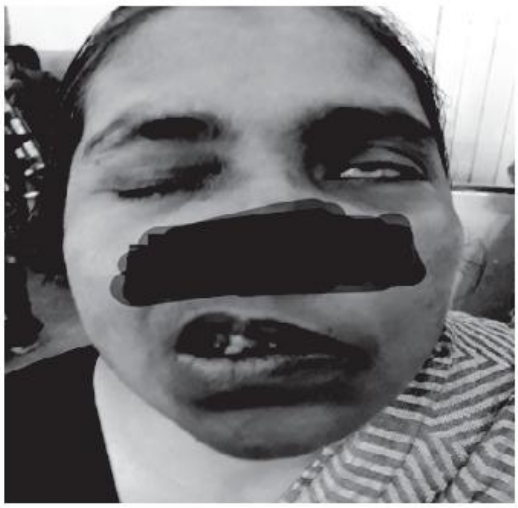

Figure-5 : Left lower motor neuron seventh cranial nerve palsy

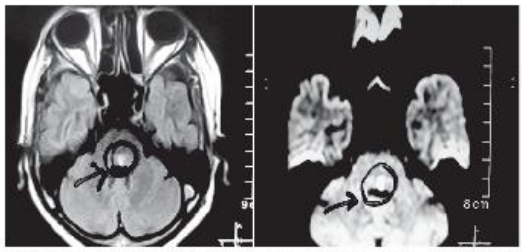

Figure-6 : Hyperintensity on FLAIR and restricted diffusion area on Diffusion-weighted imaging sequence, in the left pontine tegmentum just anterior to the forth ventricle.

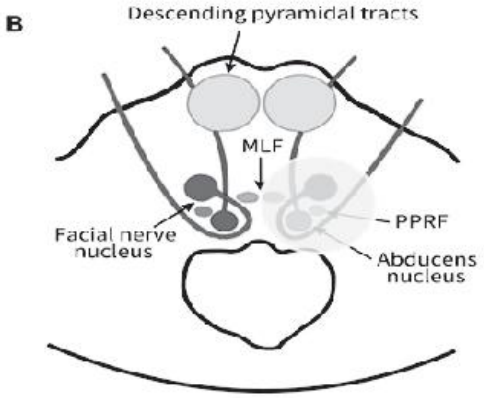

Figure-7 : Axial section through the caudal pons, showing the genu of the facial nerve (red) looping around the abducent nucleus(blue) in the pontine tegmentum and location of the lesion (yellow) with relevant structures affected. MLF: Medial longitudinal fasciculus, PPRF: Paramedian pontine reticular formation.

Discussion

Eight –and –a half eye syndrome is aclinical syndrome of internuclearophthalmoplegia, horizontal gaze paresis and ipsilateral lower motor neuron seventh cranial nerve palsy, a combination of one and half eye syndrome with lower motor neuron seventh cranial nerve palsy.[3] These clinical feature were due to lesion affecting the medial longitudinal fasciculus, paramedian pontine reticular formation or abducens nucleus and adjacent facial nucleus/fascicle at the level of dorsal tegmentum of caudal pons.[5] Those mentioned structures remain very closely in dorsal aspect of the pons and makes it vulnerable to a vascular event and demyelination.[2] Blood supply of the dorsal pontine tegmentum derived from paramedian pontine arteries, branches of basial artery. In previous case report demonstrated that

unremarkable high-quality MRI in patient with eight –and-a half syndrome highlighted the importance of clinical recognition of the syndrome.[3] On the other hand MRA is recommended as it is not only valuable to demonstrate the vascular pathology but also assisting in therapeutic management. In a case of ischemic stroke, treatment with anti-platelet and rehabilitation has been shown to improve the neurological deficit over a period of 3-6 months.Other etiology that contributes to lesion in the lower pontine tegmentum includes multiple sclerosis, vasculitis and brainstem tuberculoma.[4]

Conclusion

Diagnosis of eight-and-a-half syndrome mainly by clinical signs. It is a rare presentation and we should be capable to recognize the sign. The imaging modalities like MRI brain can help in localization of the lesion and occasionally give some clues for the etiology of that lesion. The diagnosis of the syndrome made precise anatomical localization and ensure proper treatment being given to the patient.

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