Eight-and-a-Half Syndrome: A Rare Presentation of Gaze Palsy Following Cerebrovascular Event

INTRODUCTION

A combination of horizontal gaze palsy in one eye and internuclear ophthalmoplegia in the other was first described by Freeman et al., in 1943 [1]. Later, C Miller Fisher introduced the term one-and-a-half syndrome referring to a combination of horizontal gaze palsy and internuclear ophthalmoplegia that was described earlier by Freeman [2]. The syndrome was attributed by vascular etiology particularly, rarely demyelinating and neoplastic lesions involving the medial longitudinal fasciculus (MLF) and paramedian pontine reticular formation (PPRF). In 1998 Eggenberger came out with the term eight-and-a-half syndrome, recognizing the signs help guide the clinician to precisely localize the lesion. Herein, we report a patient with eight-and-a-half syndrome.

CASE REPORT

A 52-year old gentleman presented with dizziness and double vision of 1-week duration. It was sudden onset but does not progressive. He has been diagnosed with diabetes mellitus and hypertension 3 years ago however he did not have regular follow up and medication. There was no history of trauma, no fever, no reduced vision, and no past ocular surgery. On general examination, he was alert and conscious to time, place and person with Glasgow Coma Scale (GCS) 15/15, blood pressure 168/78 mmHg, random blood sugar 20.9mm/L and afebrile. Neurological examination revealed right horizontal paresis (Figure 1) and limitation of right eye adduction with abducting nystagmus of left eye (Right internuclear ophthalmoplegia) (Figure 2). There were also right lower motor seventh cranial nerve palsy. Eight-and-a-half syndrome is a combination of ipsilateral one-and-a-half syndrome and lower motor neuron 7th nerve palsy, Brainstem conjugate gaze palsy is an important clinical diagnosis, aid in diagnosis in even a small pontine lesion. Keywords: internuclear ophthalmoplegia, one and a half syndrome, eight and a half syndrome, cerebrovascular event

Abstract: Eight-and-a-half-syndrome is a rare clinical syndrome characterized impairment of oculomotor movement; a horizontal gaze palsy to one direction, internuclear ophthalmoplegia in the other, and ipsilateral lower motor neuron seventh cranial nerve palsy. We report a case of eight-and-a-half syndrome. A 53-year-old gentleman known to have non-insulin dependent diabetes mellitus, hypertension and hypercholesterolaemia presented with double vision and dizziness of 1-week duration. There were not associated with reduced vision, no symptoms of raised intracranial pressure, not associated with limb weakness and abnormal speech. At presentation, patient had elevated blood pressure and poorly controlled blood sugar. There were right eye limitation in adduction with contralateral abducting nystagmus, right eye limited abduction consistent with right one and a half syndrome. There was also right lower motor neuron 7th nerve palsy. Eight-and-a-half syndrome is a combination of ipsilateral one-and-a-half syndrome and lower motor neuron 7th nerve palsy, Brainstem conjugate gaze palsy is an important clinical diagnosis, aid in diagnosis in even a small pontine lesion.

Keywords: internuclear ophthalmoplegia, one and a half syndrome, eight and a half syndrome, cerebrovascular event

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Fig-1: Right horizontal gaze paresis.

Fig-2: Internuclear ophthalmoplegia on left gaze shown by limitation of adduction on right eye and abducting nystagmus on left eye

Fig-3: Intact upgazed

Fig-4: Intact downgaze

Fig-5: Right lower motor neuron seventh cranial nerve palsy; loss of right nasolabial fold and drooping of the right angle of mouth.
Fig-6: Hyperintensity on right inferior pons in MRI (FLAIR)

Fig-7: Narrow of the right cerebral artery shown in MRA
DISCUSSION

Eight-and-a-half syndrome is characterized by clinical syndrome of internuclear ophthalmoplegia, horizontal gaze paresis and ipsilateral lower motor neuron seventh cranial nerve palsy, a combination of one-and-a-half syndrome with lower motor neuron seventh cranial nerve palsy [3]. These clinical oculomotor findings were resulted from 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]. Proximity of these structures makes it vulnerable to a vascular event and demyelination [2]. Blood supply of the dorsal pontine tegmentum derived from paramedian pontine arteries, branches of basilar artery. In previous case report demonstrated that unremarkable high-quality MRI in patients 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 period a period of 3 to 6 months. Other etiology that contributes to lesion in lower pontine tegmentum includes multiple sclerosis, vasculitis and brainstem tuberculoma [4].

CONCLUSION

Diagnosis of eight-and-a-half syndrome mainly by clinical signs, thus although the incidence is rare, one should be able to recognize the features. The imaging modalities which available such as magnetic resonance imaging of brain and magnetic resonance angiography of cerebral circulation supplement the clinical diagnosis made earlier. The recognition of the syndrome made precise anatomical localization and ensure appropriate treatment being given to the patient.

REFERENCES


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