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ABSTRACT

Background: “Eight-and-a-half” syndrome is an unusual neuro-ophthalmic clinical entity which comprises of “one-and-a-half” syndrome (conjugated horizontal gaze palsy and internuclear ophthalmoplegia) with ipsilateral fascicular seventh cranial nerve palsy. This condition is caused by circumscribed lesions of the pontine tegmentum involving the abducens nucleus, the ipsilateral medial longitudinal fasciculus, and the adjacent fascial colliculus. Here, we report a case of “eight-and-a-half” syndrome.

Materials and Methods: Case Report

Result: A 62-year old gentleman with hypertension, diabetes mellitus and paroxysmal atrial fibrillation presented with sudden onset of binocular diplopia and facial asymmetry. Examination showed right eye exotropia at primary gaze. He had conjugate horizontal gaze palsy to the left. On right gaze, the patient had restricted left eye adduction with retained ability to abduct the right eye, which evoked a right horizontal nystagmus. Convergence with near fixation were spared. Vertical eye movements were also normal. In addition, he had left lower motor neuron facial weakness with no other neurological deficit. Computed tomography scan showed a left paramedian pontine infarct. A diagnosis of eight-and-a-half syndrome was made. The patient was referred to neuromedical team for treatment of acute stroke in which he was treated conservatively. Evaluation after 4 weeks showed slight improvement in adduction lag of the right eye.

Conclusion: Our patient presented with the unique combination of left sided horizontal one-and-a-half syndrome and lower motor neuron seventh cranial nerve palsy. Combination of these signs (seven plus one-and-a-half) is known as eight-and-a-half syndrome. Recognition of the spectrum of eight-an-a-half syndrome allows precise anatomic localisation of the lesion to pontine tegmentum region.

Keywords: eight-and-a-half syndrome, one-and-a-half syndrome, internuclear ophthalmoplegia, pontine infarct, pontine tegmentum
1.0 Introduction

One-and-a-half syndrome is a clinical condition characterised by an ipsilateral conjugate horizontal gaze palsy and an ipsilateral internuclear ophthalmoplegia (INO) with sparing of contralateral abduction. However, the restriction of adduction can be overcome by convergence. This condition is caused by a lesion that affects the ipsilateral paramedian pontine reticular formation (PPRF) or the abducens (sixth cranial nerve) nucleus and the ipsilateral medial longitudinal fasciculus (MLF). When this lesion also affects the fasciculus of the ipsilateral facial nerve (seventh cranial nerve) in the region of the facial colliculus at it wraps around the abducens nucleus, it produces a lower motor neuron facial nerve palsy. When this occurs, it is termed an eight-and-a-half syndrome (a one-and-a-half syndrome plus a seventh cranial nerve palsy). [1,2].

2.0 Materials and Methods

Case Report

3.0 Result

A 62-year old gentleman with underlying hypertension, diabetes mellitus and paroxysmal atrial fibrillation presented with one day history of sudden onset of binocular diplopia with left sided facial numbness and asymmetry. Symptoms were followed by slurring of speech and subsequently generalised headache, dizziness and vomiting.

On examination, there was dysarthria with left sided lower motor neuron facial nerve palsy as evident by loss of wrinkling of left forehead, loss of left nasolabial fold and left lagophthalmos. (Fig. 1)

Ocular examination showed right eye exotropia on primary gaze. There was conjugate horizontal gaze palsy to the left. On looking to the right, left eye adduction is restricted but abduction of the right eye is intact evoking a right horizontal nystagmus, a condition termed internuclear ophthalmoplegia. The combination of a horizontal gaze palsy with an internuclear ophthalmoplegia is called “one-and-a-half” syndrome. (Fig. 2)
Fig. 2  (A-C) Combination of left gaze paresis along with restricted adduction in left eye and intact abduction in right eye (with nystagmus) suggestive of left horizontal one-and-half syndrome. Note the normal vertical eye movements from primary position of gaze (D, E)

Otherwise, other cranial nerve examinations were normal and there were no other neurological deficits. In view of findings of a one-and-a-half syndrome with an ipsilateral lower motor neuron facial nerve palsy, a clinical diagnosis of “eight-and-a-half” syndrome is made. Acute onset of presentation would suggest a vascular origin.

A computed tomography scan of the brain showed a left paramedian pontine hypodensity suggestive of an infarct which further supports the diagnosis (Fig.3). A magnetic resonance imaging was not done in view patient had a cardiac pacemaker.

Fig.3  CT scan showing a left paramedian pontine hypodensity

During admission, patient developed an episode of atrial fibrillation and was started on T. propranolol 40mg BD and T. amiodarone 200mg OD after which the heart rate was controlled with no signs of failure. ECHO done showed no valvular lesions.
Patient was started on T. aspirin 150mg OD and T. atorvastatin 20mg ON for the treatment of acute ischemic stroke. He was also referred to speech therapist, occupational and physio therapist for rehabilitation.

Subsequent eye clinic follow-up showed gradual improvement of eye movement with resolved restricted left eye adduction with residual minimal horizontal gaze palsy to the left at six month post infarct.

4.0 Discussion

Fischer et al. first introduced one-and-a-half-syndrome in 1967 as a combination of horizontal gaze palsy and internuclear ophthalmoplegia [3]. It is defined as a horizontal gaze palsy due to damage to the parapontine reticular formation (PPRF), and an ipsilateral internuclear ophthalmoplegia (INO) which leads to a deficit in adduction of the ipsilateral bulbus due to an interruption of internuclear fibres of the ipsilateral medial longitudinal fasciculus (MLF) after crossing the midline.

Eggenberger et al later described eight-and-a-half syndrome as one-and-a-half-syndrome in combination with a peripheral facial nerve palsy. [4], usually caused by a vascular or demyelinating lesion [5].

The close anatomical proximity of PPRF, MLF, abducens nucleus and facial nerve in the dorsal pontine area is responsible for the presentation of eight-and-a-half syndrome in which there is simultaneous occurrence of horizontal gaze palsy, internuclear ophthalmoplegia as well as peripheral facial palsy [6].

5.0 Conclusion and recommendation

Our patient presented with the combination of left lateral gaze palsy, internuclear ophthalmoplegia and left lower motor neuron facial nerve palsy (collectively called eight-and-a-half syndrome) secondary to left paramedian pontine infarct. In our patient, the most likely cause of infarct is an embolus secondary to paroxysmal atrial fibrillation.

Recognising the specific constellation of findings allows for precise localisation to the pontine tegmentum, which is mostly supplied by the inferior cerebellar artery or paramedian pontine perforators of the basilar artery, and also for planning of appropriate investigations to identify the possible causes.

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Consent was obtained by all participants in this study.
Declaration

Author(s) declare that there is no conflict of interest, no financial support was received from any organisation for the submitted work and no other relationships or activities that could appear to have influenced the submitted work.

Authors contribution

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