Introduction

Brainstem vascular events involving the pons often produce gaze palsies in combination with other clinical signs. Fisher first described a variant of brainstem stroke with the combination of horizontal gaze palsy in one eye associated with ipsilateral internuclear ophthalmoplegia (INO) and named it one and a half syndrome. When other cranial nerves are involved in combination with this, numbers denoting the nerves are added to this syndrome, resulting in new syndromes such as eight-and-a-half syndrome, which results from the combination of one and a half syndrome and ipsilateral lower motor neurone (LMN) type VII cranial nerve palsy. Rosini et al. first described a novel neuro-ophthalmic pontine syndrome in 2013 when eight-and-a-half syndrome was associated with other clinical signs such as hemiparesis and hemi-hypesthesia in contralateral limbs and coined the term as ‘nine’ syndrome. We report a rare case of brainstem stroke in a 56-year-old male who presented with clinical signs consistent with right-side eight-and-a-half syndrome with left hemiparesis and magnetic resonance imaging (MRI) of the brain confirming an acute infarction in the right side of the pons near to the midline. This report demonstrates the importance of careful examination for all neurological signs in stroke patients as this will lead to the precise localization of the lesion anatomically at the bedside and enable the institution of effective treatment.

Case history

A 56-year-old male presented with left upper and lower limb weakness and double vision on looking towards the right side of body, which were sudden in onset and persistent during the first week. Apart from being a smoker, he did not have any other risk factors for stroke. He had a past history of pulmonary tuberculosis for which he completed treatment 6 years previously. He has neither constitutional symptoms nor respiratory symptoms suggestive of active tuberculosis. Examination of the cranial nerves revealed right-side horizontal gaze palsy with right internuclear ophthalmoplegia and right lower motor neuron type VII nerve palsy constituting eight-and-a-half syndrome. With the additional involvement of left upper and lower limb upper motor neuron weakness, it revealed the lesion responsible for the ‘nine’ syndrome in magnetic resonance imaging of the brain. It is our purpose to highlight the genesis of this combination of clinical signs.

Keywords

Internuclear ophthalmoplegia, one and a half syndrome, horizontal gaze palsy, eight-and-a-half syndrome, nine syndrome

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VII nerve palsy (absence of wrinkling of right forehead, difficulty in closing right eyes, weakness of blowing of right cheek, deviation of mouth towards right side) (Figure 1(d)). All the other cranial nerves and cerebellar examination were normal. Moreover, he also had upper motor neuron (UMN) type weakness in the left upper limbs (exaggerated biceps jerk, triceps jerk, and supinator jerk and increased tone) and lower limbs (exaggerated knee jerk and ankle jerk, and upgoing planter and increased tone with ankle clonus). The rest of the systemic examinations were normal except for the higher blood pressure reading of 170/100 mmHg on admission.

All blood tests including plasma electrolytes, liver function tests, full blood count, and erythrocyte sedimentation rate (14 mm in the first hour) were normal. Non-contrast computed tomography of the brain was normal. Screening for tuberculosis did not reveal any evidence of active tuberculosis or reactivation. Cerebrospinal fluid (CSF) analysis was acellular with a total protein of 17 mg/dL. MRI of the brain confirmed an acute infarction in the right side of the pons near the midline (Figure 2). There were no areas of abnormal enhancement seen in the cerebral parenchyma or meninges.

The patient was treated with a combination of aspirin 300 mg at night for the initial 2 weeks, and then switched to clopidogrel 75 mg at night and atorvastatin 40 mg at night. Blood pressure was monitored carefully during the hospital stay and the patient did not require antihypertensive medication on discharge. Neuro-rehabilitation was arranged. After a month of follow-up, significant improvement in the horizontal gaze was observed, especially in the left eye.

**Discussion**

Eight-and-a-half syndrome is not an uncommon clinical entity among brainstem stroke patients. It is caused by a lesion in the dorsal tegmentum of pons due to the involvement of either abducens nucleus or parapontine reticular formation (PPRF) and medial longitudinal fasciculus (MLF) combined with adjacent facial nerve fascicle or colliculus. As these are LMN-type fibres of the VII nerve, a distinctive LMN type VII nerve palsy is observed in this brainstem stroke. The most common etiologies described for the above presentation are brainstem infarction and haemorrhage, multiple sclerosis, and tumours involving the brainstem and fourth ventricle. The majority (70%) are caused by brainstem infarction and multiple sclerosis. When eight-and-a-half syndrome extends to involve the adjacent structure such as corticospinal tract and medial lemniscus, it additionally produces hemiparesis and hemihypesthesia contralaterally. Transient hemiparesis has well been reported with eight-and-a-half syndrome due to the extension of transient perilesional oedema in the infarct site. Our patient had right horizontal gaze palsy with right INO and right LMN type VII nerve palsy constituting eight-and-a-half syndrome. Nine syndrome...
has also been named as an eight-and-a-half plus syndrome, and it can include hemiataxia.6

Nine syndrome is a rare entity and there have been few cases reported in the literature till date, which are shown in Table 1.3,7–11 This syndrome comprises eight-and-a-half syndrome associated with hemiparesis and hemianesthesia due to additional involvement of the corticospinal tract and medial lemniscus. A variation to nine syndrome was reported in two patients who had eight-and-a-half syndrome without hemiparesis/hemianesthesia but instead had contralateral hemiataxia. In our case, with the additional involvement of left upper and lower limb UMN weakness, the possibility of ‘nine’ syndrome was suspected clinically, and his MRI of the brain finding ultimately confirmed this diagnosis. Furthermore, the nine syndrome is confirmed clinically, not radiologically, and imaging, although useful, does not change the clinical impression, only the etiological workup.

## Conclusion

Since the first description of the classical eight-and-a-half syndrome 50 years ago, there have been several reports of new variants of this syndrome. The reported variants of the syndrome are due to the extension of the brainstem stroke lesion to other parts of the brainstem as has been documented in our case. Precise anatomical localization of the lesion is essential for the delivery of effective treatment. The awareness of these clinical signs helps in precise localization and

### Table 1. Case reports of nine syndrome in literature.

| Case report       | Clinical findings                                                   |
|-------------------|---------------------------------------------------------------------|
| Rosini et al.3    | Eight-and-a-half syndrome associated with contralateral hemiparesis and hemihypesthesia |
| Mahale et al.7    | A variation to nine syndrome in two patients who had eight-and-a-half syndrome without hemiparesis/hemianesthesia but instead had contralateral hemiataxia |
| Yong et al.8      | One and a half syndrome, left facial nerve palsy, and contralateral hemiataxia |
| Singhdev et al.9  | Internuclear ophthalmoplegia, ipsilateral horizontal gaze palsy, lower motor neuron type of facial palsy, contralateral hemiparesis, and hemianesthesia |
| Cao et al.10      | Eight-and-a-half syndrome plus hemiplegia (atypical nine syndrome) |
| Siam et al.11     | Combination of lower motor facial nerve lesion, one and a half syndrome, and hemiparesis or hemiataxia |

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Figure 2. Diffusion-weighted MRI of the brainstem shows a tiny area of restricted diffusion in the caudal right paramedian region of the pons.
consideration of relevant etiologies of nine syndrome. It is our purpose to highlight the genesis of this combination of clinical signs.

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**Informed consent**
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