CASE REPORTS

“Nine” syndrome and its MRI brain findings

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Abstract

We describe a rare presentation of pontine infarction in a lady who was on dual antiplatelet therapy. Her presentation includes one and a half syndrome, left facial nerve palsy and contralateral hemiataxia. Magnetic resonance imaging (MRI) of the brain revealed acute infarction of the dorsal pons. A diagnosis of “nine” syndrome secondary to lacunar stroke was made. She continued dual antiplatelet therapy and her symptoms resolved completely over three months.

Keywords: Stroke; facial nerve palsy; one and a half syndrome; eight and a half syndrome; nine syndrome; caudal pontine tegmental syndrome

INTRODUCTION

One and a half syndrome is characterized by combined unilateral conjugate gaze palsy and internuclear ophthalmoplegia (INO) due to lesions affecting abducens nucleus or paramedian pontine reticular formation (PPRF) and the adjacent medial longitudinal fasciculus (MLF). Facial nerve palsy, in addition to one and a half syndrome, is called eight and a half syndrome (7 + 1.5 = 8.5); whereas a further addition of hemiparesis or hemihypesthesia would constitute a “nine” syndrome (0.5 + 8.5 = 9). This is a case report describing the clinical presentation and features of the MRI of the brain of a patient with this syndrome.

CASE REPORT

A 68-year-old Indian lady with underlying type 2 diabetes, hypertension and ischemic heart disease presented with sudden onset double vision, dizziness, vomiting and unsteadiness for two days. She has been on dual antiplatelet therapy (DAPT) (Cardiprin 100 mg daily and clopidogrel 75 mg daily) and simvastatin 40mg daily after percutaneous intervention of her right coronary artery five months ago. On examination, her Glasgow Coma Scale (GCS) was full, blood pressure was 186/83 mmHg and pulse rate was 60 beats per min. Neurological examination revealed one and a half syndrome (Figure 1), lower motor neuron type of left facial nerve palsy (Figure 2) and contralateral hemiataxia. No carotid bruit was auscultated. Electrocardiograms revealed normal sinus rhythm. Laboratory tests were unremarkable. Computed tomography of the brain, however, did not show significant abnormality. MRI of brain on day four of admission showed hyperintense T2/FLAIR lesion in the dorsal portion of the pons with restricted diffusion, involving the MLF, PPRF, facial genu, abducens nucleus, and medial lemniscus (Figure 3). Magnetic resonance angiogram of the brain was normal. Echocardiogram showed ejection fraction of 64.1%, normal left atrial size (dimension: 3.5 cm) and no mural thrombus. A constellation of her symptoms corresponded to “nine” syndrome. She continued DAPT and achieved complete resolution of the symptoms after three months.

DISCUSSION

Combined ipsilateral horizontal gaze palsy and INO was termed one and a half syndrome by Fisher et al. in 1967. If the lesion extends laterally to involve the genu of the facial nerve, it gives rise to ipsilateral LMN facial nerve palsy and was described as eight and a half syndrome. A further concomitant involvement of the corticospinal tract or medial lemniscus might cause hemiparesis or hemihypesthesia, which was first coined as “nine” syndrome by Rosini et al. in 2013. Ischemia, demyelination, tumour, hemorrhage, trauma and infection are potential causes but

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Figure 1. One and a half syndrome. (A) Neutral position; (B) Right gaze; (C) Left gaze

Figure 2. Lower motor neuron type of left facial nerve palsy. (A) Flattening of the left nasolabial fold; (B) Frontalis and upper orbicularis oculi of the left side were affected
the common etiologies are ischemic stroke and multiple sclerosis.1

Dorsal pons is usually supplied by two paramedian pontine perforating branches of the basilar artery, although it can be only one that bifurcates at the level of abducens nucleus due to anatomical variation. Occlusion of the perforators result in ipsilateral paramedian pontine syndrome. Proximal occlusion causes infarction of the paramedian pontine basis and the tegmentum,
whereas distal occlusion causes dorsal pontine tegmental syndrome. In the present case, the MRI findings of acute infarction of the dorsal pons suggested distal occlusion of her left paramedian pontine perforators. Figure 4 shows the schematic diagram of the pontine structures involved.

Despite the efficacy of aspirin has been well proven in primary and secondary ischemic stroke prevention, the role of combination therapy was not clear. The landmark SPS3 trial found no benefit of DAPT in secondary prevention of lacunar stroke. On the contrary, CHANCE trial demonstrated benefit of short-term (≤3 months) DAPT in those with minor stroke or transient ischemic attack. Lacunar stroke results predominantly from an intrinsic vascular pathology, whereby small vessel disease is the commonest etiology. The use of antiplatelet therapy in this group of patients has been a topic of controversy since the mechanism might not be of atherothrombotic origin. A recent case report indicated that DAPT resulted in favorable outcomes in a patient with bilateral pontine tegmental infarction. However, meta-analysis of randomized controlled trials suggested negative results. Notably, our patient was on DAPT yet she suffered a lacunar stroke.

In conclusion, identifying clinical signs of this syndrome guides precise anatomic localization of the lesion to the ipsilateral lower pontine tegmentum. Lacunar stroke is one of the commonest etiologies but there is insufficient data to suggest DAPT use.

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DISCLOSURE
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REFERENCES