Clinical Case Report

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Atypical "nine" syndrome in bilateral pontine infarction

A case report

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Abstract

Rationale: "Nine" syndrome, that is "eight-and-a-half" syndrome associated with hemiplegia and hemidysesthesia, is a rare disorder. This study aimed to report a Chinese patient with acute bilateral pontine infarction manifesting as eight-and-a-half syndrome plus hemiplegia (atypical nine syndrome), and also the clinical and neuroimaging findings were explained and discussed with review of the literature.

Patient concerns: A 79-year-old woman experienced sudden vertigo, nausea, vomiting, and weakness at her left arm and leg. The neurological examination disclosed her right horizontal gaze palsy, internuclear ophtalmoplegia (INO), and right-sided peripheral facial paralysis combined with slight left-sided hemiplegia, which were consistent with atypical nine syndrome.

Diagnoses: Cranial magnetic resonance imaging (MRI) displayed acute multiple ischemic infarction, involving bilateral pontine tegmentum, basilar part of right paramedian pontine, and left cerebellar hemisphere. Intracranial MR angiography (MRA) revealed right middle cerebral artery occlusion, no clear visualization of bilateral vertebral arteries, and basilar artery hypoplasia with stenotic segments.

Interventions: Thrombolysis could not be performed due to the time window. The patient was given low molecular weight heparin for anticoagulation because of posterior circulation and progressive stroke.

Outcomes: The vertigo disappeared, and a notable improvement with minimal restriction in the right horizontal gaze and partial relief of her facial paralysis were found at discharge, while her left hemiparesis was fully resolved. No recurrence of cerebral infarction was observed during follow-up as well.

Lessons: This case report with atypical nine syndrome is fairly rare. Nine syndrome may refer to the lesion located in unilateral tegmentum of the caudal pontine plus paramedian pontine, with an important localization value.

Abbreviations: DWI = diffusion-weighted imaging, INO = internuclear ophtalmoplegia, MLF = medial longitudinal fasciculus, MRA = magnetic resonance angiography, MRC = Medical Research Council, NIHSS = National Institutes of Health Stroke Scale, MRI = magnetic resonance imaging, PPRF = paramedian pontine reticular formation.

Keywords: diffusion-weighted imaging, eight-and-a-half syndrome, one-and-a-half syndrome, pontine infarction, stroke

1. Introduction

The so-called "eight-and-a-half" syndrome, originally proposed by Eggenberger in 1998, refers to "one-and-a-half" syndrome combined with ipsilateral fascicular seventh cranial nerve palsy, which is caused by circumscribed lesions of the pontine tegmentum involving the abducens nucleus, the adjacent facial colliculus, and the ipsilateral medial longitudinal fasciculus (MLF).[1] Rosini et al proposed the concept of "nine" syndrome, which showed eight-and-a-half syndrome plus hemiplegia and hemidysesthesia ($8^{1/2} + {}^{1/2} = 9$).[2] Since then, few variants of nine syndrome have been reported. Mahale et al presented two patients with a variant of nine syndrome who manifested as eight-and-a-half syndrome plus hemiataxia.[3] Maas and Verrips reported a typical "nine syndrome" patient in Neurology.[4] Here, we reported a Chinese patient with acute bilateral pontine infarction presenting as atypical nine syndrome (eight-and-a-half syndrome plus hemiplegia), and discussed the clinical and neuroimaging findings with review of the limited literature.

2. Case presentation

A 79-year-old Chinese woman was admitted to our inpatient department due to sudden vertigo, nausea, vomiting, and weakness at her left arm and leg approximately 6 h after onset. In terms of medical history, she had a history of obesity and

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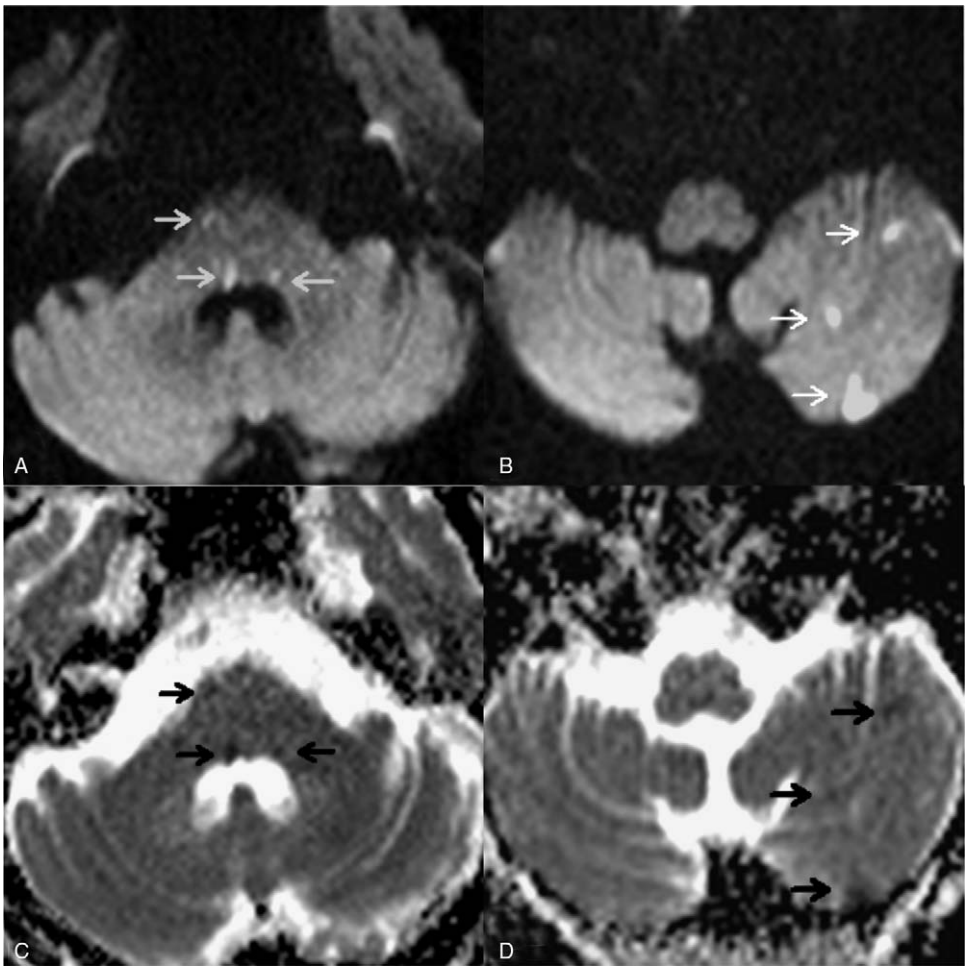

Figure 1. (A–D) Cranial MRI. Axial diffusion-weighted imaging (DWI) showed a high-intensity lesion comprising bilateral pontine tegmentum, basilar part of right paramedian pontine, and left cerebellar hemisphere (A and B, white arrows), with low apparent diffusion coefficient signals in corresponding regions, indicating an ischemic lesion (C and D, black arrows).

hypertension. She was diagnosed as bilateral cerebellar infarction 1 month before her admission, and she was given regular secondary prevention. The findings of physical examination were listed as follows: temperature (*T*): 36.7°C, pulse rate (*P*): 75 times/min, respiration (*R*): 18 times/min, and blood pressure (BP): 150/100 mm Hg; there were no abnormal findings in the lungs, heart, and abdomen. On neurological examination, she was alert, and her cognitive function was also normal. She presented with right horizontal gaze palsy, internuclear ophthalmoplegia (INO), and right-sided peripheral facial paralysis combined with slight left-sided hemiplegia (Medical Research Council (MRC) scale, V^{-}). She achieved stable and accurate results in finger-nose test, while failed to complete the inspection of other coordinate movements. The Babinski sign was negative bilaterally. There was no paresthesia as well. Her score of National Institutes of Health Stroke Scale (NIHSS) was 3.

The results of laboratory test were as follows: coagulation test showed that D-dimer level was 1.000 μg/mL (normal range, 0–0.500 μg/mL), and biochemical test demonstrated that triglyceride level was 7.20 mmol/L (normal range, 0.45–1.81 mmol/L). In addition to those abnormal findings, the remaining test results were normal. Echocardiography showed left ventricular diastolic dysfunction. Carotid color ultrasonography disclosed bilateral carotid atherosclerosis with plaque in the right common carotid artery. Electrocardiogram showed sinus rhythm, left anterior hemiblock, and abnormal ST segment depression. Cranial magnetic resonance imaging (MRI) displayed acute multiple ischemic infarction, involving bilateral pontine tegmentum, basilar part of right paramedian pontine, and left cerebellar hemisphere (Fig. 1a–d). Intracranial MR angiography (MRA) revealed the right middle cerebral artery occlusion, no clear visualization of bilateral vertebral arteries, and basilar artery hypoplasia (the maximum diameter of basilar artery is 1.89 mm) with stenotic segments.

After admission, thrombolysis could not be performed due to the time window. The symptoms of right facial paralysis and left hemiplegia were slightly aggravated (Her NIHSS score was 5), and the patient was given low molecular weight heparin for anticoagulation. Routine use of atorvastatin statin and symptomatic supportive treatment were given simultaneously. About 7 days later, the vertigo disappeared, and a notable improvement in her symptoms was found at discharge. The final neurological examination revealed minimal restriction in the right horizontal gaze and partial relief of her facial paralysis, while her left hemiparesis was fully resolved without dystaxia. During follow-up, the patient had no recurrence of cerebral infarction.

3. Discussion

In 1967, Fisher, for the first time, proposed the concept of one-and-a-half syndrome (conjugated horizontal gaze palsy and INO), in which the lesion involved the ipsilateral abducens nucleus or paramedian pontine reticular formation (PPRF), and the ipsilateral MLF.[5] Subsequently, Eggenberger put forward eight-and-a-half syndrome (i.e., one-and-a-half syndrome plus ipsilateral peripheral seventh cranial nerve palsy) in 1998.[1] Accordingly, some new variants of eight-and-a-half syndrome have been successively proposed, such as "fifteen-and-a-half" syndrome (i.e., one-and-a-half syndrome plus bilateral seventh cranial nerve palsy proposed by Bae et al [6]), nine syndrome (i.e., eight-and-a-half syndrome plus hemiplegia and hemidysesthesia presented by Rosini et al[2]), and "thirteen-and-a-half" syndrome (i.e., eight-and-a-half syndrome plus ipsilateral fifth cranial nerve palsy introduced by Allbon et al[7]). Although Rosini et al. first proposed the concept of nine syndrome,[2] Uysal et al had previously reported a patient with the right pontine infarction who manifested as eight-and-a-half syndrome combined with transient hemiparesis, which should belong to atypical nine syndrome.[8] In the present case, the patient exhibited one-and-a-half syndrome, peripheral right facial palsy, and mild left-sided hemiparesis, which were consistent with the characteristics of eight-and-a-half syndrome along with contralateral hemiparesis (i.e., atypical nine syndrome). Although the clinical manifestations on admission were very similar to the case reported by Uysal et al,[8] the neuroimaging findings were distinctly different.

The clinical manifestations of this patient could be explained by the anatomical location of the infarcts on DWI. The large lesion in the right paramedian pontine tegmentum might not only involve the ipsilateral abducens nucleus or horizontal gaze center in PPRF and MLF, but also involve the adjacent facial colliculus, configuring eight-and-a-half syndrome. The right lacunar paramedian pontine infarction, involving the corticospinal tract, might result in mild left-sided hemiplegia. However, the difference was the absence of contralateral sensory disturbance, which might be due to the lesion not extending the medial lemniscus. In brief, we presumed that the infarcts in the right pontine were responsible for the patient's atypical nine syndrome. The definite involvement of the corticospinal tract was consistent with the left-sided hemiplegia, which suggested a direct tissue damage caused by ischemia rather than a transient extension of the peri-infarct edema as recently postulated by Uysal et al.[8] In addition, although the patient had lesion in the left pontine tegmentum and right cerebellum, the corresponding focal neurological signs could not be detected in the examination.

Nine syndrome is rare in clinical practice. The etiology spectrum of several reported cases referred to vascular infarction or hemorrhage,[2–4,8] which was different from that of eight-and-a-half syndrome. Our case was the first nine syndrome reported in China. The etiology was also considered to be vascular infarction. However, it should be differentiated from multiple sclerosis, tumors located in the pontine, etc. Although our patient had two onsets of ischemic stroke, the prognosis was satisfactory, which might be related to basilar artery hypoplasia and possible presence of compensatory collateral circulation.[9]

In conclusion, we reported a Chinese patient with eight-and-a-half syndrome associated with hemiplegia, appearing with atypical nine syndrome as a consequence of pontine infarction. Such case report was fairly rare. Nine syndrome may refer to the lesion located in unilateral tegmentum of the caudal pontine plus paramedian pontine, with an important localization value for diagnosis.

4. Patient consent

The patient provided consent for the writing and publication of this case report.

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

Author contributions

Conceptualization: Shugang Cao, Rongfeng Wang, Mingwu Xia.
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Visualization: Shugang Cao.
Writing – original draft: Shugang Cao.
Writing – review & editing: Shugang Cao, Rongfeng Wang, Mingwu Xia, Wen'an Xu.

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