

One-and-a-half syndrome with facial diplegia: A case report

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Abstract

One-and-a-half syndrome with facial diplegia, also referred to as the fifteen-and-a-half syndrome, is an extremely rare clinical entity caused by involvement of bilateral tegmentum of pons. Herein, we report a 52-year-old male who presented with one-and-a-half syndrome with left facial paralysis, which was consistent with the so-called eight-and-a-half syndrome. Brain magnetic resonance imaging showed pontine infarction. Five days after initiation of antiplatelet therapy, the patient developed right facial paralysis, a diagnosis of fifteen-and-a-half syndrome was made, repeat MR imaging revealed bilateral pontine tegmentum infarction. Fifteen-and-a-half syndrome is a newly proposed concept associated with pontine infarction. The clinicoradiological features of this specific disease are as yet unclear due to its extreme rarity. The current case would help advance the current understanding of the disease spectrum of pontine infarction.

Keywords: one-and-a-half syndrome; facial diplegia, pontine infarction; fifteen-and-a-half syndrome; eight-and-a-half syndrome

INTRODUCTION

“One-and-a-half” syndrome refers to a gaze disorder characterized by ipsilateral gaze paresis or palsy, and internuclear ophthalmoplegia on contralateral gaze. The syndrome was first described by Fisher in 1967.¹ 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.² In 2005, Bae described a new clinical entity, “fifteen-and-a-half” syndrome, i.e., one-and-a-half syndrome with facial diplegia.³ Due to its extreme rarity, the clinical course, radiological characteristics and outcomes of this specific disease are not well-characterized. Herein, we report a case of fifteen-and-a-half syndrome.

CASE REPORT

A 52-year-old male presented with a 3-day history of dizziness, nausea, and diplopia. He had a history of hypertension and poorly-controlled diabetes mellitus. On admission, his blood pressure was 154/87 mmHg, and the heart rate was 73 beats per minute. Neurological examination revealed

complete left horizontal conjugate gaze palsy and a right adduction deficit. Vertical ocular movements were normal and there was no nystagmus or ptosis. The pupils were equal in size and normally reactive to light. Additionally, left peripheral facial palsy was noted. The physical examination findings were otherwise normal. Laboratory examination showed an elevated fasting blood-glucose level (16 mmol/L) and glycosylated hemoglobin percentage (12.3%), and a slightly increased low-density lipoprotein cholesterol level (3.82 mmol/L). The blood routine examination, erythrocyte sedimentation rate, blood biochemistry including serum electrolytes, homocysteine level and tumor markers were all within the normal reference range. Magnetic resonance (MR) diffusion-weighted imaging (DWI) revealed a pontine tegmentum infarct (Figure 1A). Magnetic resonance angiography (MRA) showed stenosis of posterior cerebral vessels (Figure 2). Five days later, the patient developed right peripheral facial paralysis and bilateral pyramid sign. Repeat MR imaging showed aggravation of the pontine infarct (Figure 1B). A diagnosis of fifteen-and-a-half syndrome was made, and dual-antiplatelet, hypocholesterolemic, antihypertensive and antidiabetic therapy was added. Half a month later, there was slight improvement of diplopia, whilst the facial diplegia continued to persist. The

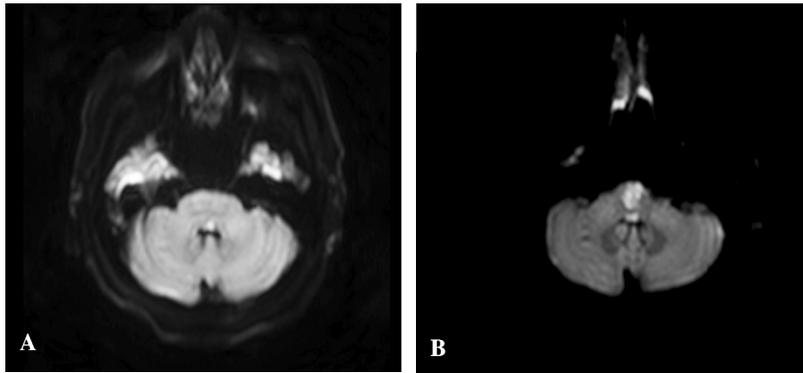


Figure 1. Brain magnetic resonance (MR) diffusion-weighted imaging showing hyperintense lesion in the pontine tegmentum (A); Repeat MR imaging five days later showed extension of the pontine infarct (B)

patient continued to undergo neuro-rehabilitation on the stroke ward.

DISCUSSION

one-and-a-half syndrome is usually due to a single unilateral lesion of the paramedian pontine reticular formation or the abducens nucleus on one side (causing the conjugate gaze palsy), with interruption of internuclear fibers of the ipsilateral medial longitudinal fasciculus after it has crossed the midline from its site of origin in the contralateral abducens nucleus (causing failure of adduction of the ipsilateral eye). Most frequent causes include multiple sclerosis and brainstem infarction.⁴ The isolated form of one-and-a-half syndrome is uncommon, and this syndrome is usually accompanied by cranial nerve palsies, hemiplegia, or hemihypesthesia.

Fifteen-and-a-half syndrome is attributed to a bilateral tegmental pontine lesion which involves the right abducens nucleus or fasciculus, bilateral or right medial longitudinal fasciculus,

and bilateral intraaxial fasciculi of the facial nerve. Pontine infarction is largely attributed to stenosis or occlusion of vertebrobasilar arteries as well as atherosclerotic plaque formation or occlusion of perforating arteries in the context of hypertension and/or diabetes. Bilateral pontine tegmentum infarction may be associated with variation in vascular anatomy. In some cases, the bilateral pontine tegmentum was supplied by unilateral pontine paramedian artery, and the dysfunction of this artery results in bilateral infarction. The clinical manifestations of pontine infarction include horizontal gaze palsy, ipsilateral miosis, peripheral facial paralysis, contralateral hemisensory loss, and dysarthria. In the present case, the patient presented with diplopia, horizontal gaze palsy, and facial palsy, which is consistent with fifteen-and-a-half syndrome.

The clinical course of fifteen-and-a-half syndrome is not well understood due to its extreme rarity, and the treatment is yet to be established. For this patient with many risk factors and atherosclerotic posterior circulation, we gave the



Figure 2. Magnetic resonance angiogram (MRA) showing stenosis of bilateral vertebral arteries and basilar arteries.

dual-antiplatelet therapy and achieved favorable outcomes. It appeared that single antiplatelet alone was not sufficient to prevent the second stroke, and timely dual-antiplatelet therapy may have been effective in arresting further progression of the disease.

In conclusion, the fifteen-and-a-half syndrome is a rare clinical entity caused by bilateral pontine tegmental lesion.

DISCLOSURE

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Conflict of Interest: None

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