

“Nine Syndrome”: A Rare Neuroophthalmological Syndrome Associated with HIV Vasculopathy

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Abstract

Keywords

- ▶ nine syndrome
- ▶ medullary infarct
- ▶ eight-and-a-half syndrome
- ▶ MLF
- ▶ PPRF
- ▶ HIV

There are many named brainstem syndromic variants of medullary infarcts among which “eight-and-a-half” syndrome is a rare condition that involves ipsilateral abducens nucleus or paramedian pontine reticular formation, ipsilateral medial longitudinal fasciculus, and adjacent facial colliculus/facial nerve. Here, we describe a new variant of brainstem syndrome, which includes the clinical features of “eight-and-a-half syndrome” with associated hemiparesis. This patient is a known case of HIV illness since 6 years on antiretroviral therapy, presented with features of “Nine” syndrome.

Introduction

Brainstem strokes present with varied symptomatology and pose diagnostic challenges to the clinicians. Apart from various known named brainstem stroke syndromes, pontine tegmental lesions usually present with gaze palsies, internuclear ophthalmoplegia (INO), nystagmus, and abducens palsy. A combination of horizontal gaze palsy in one eye and INO in the other eye caused by lesion in medial longitudinal fasciculus (MLF) or paramedian pontine reticular formation (PPRF) was first described by Freeman et al in 1943. Subsequently, Fisher in 1967¹ coined the term one-and-a-half syndrome to describe the same. The combination of one-and-a-half syndrome and ipsilateral seventh nerve palsy is known as eight-and-a-half syndrome, which was described by Eggenberger in 1998.² Eight-and-a-half syndrome with hemiataxia/hemisensory loss/hemiparesis fits into the description of a new neuro-ophthalmic syndrome often termed as nine syndrome. There are few case reports of nine syndrome published in literature. Here, we describe a patient with retroviral illness and nine syndrome with partial clinical improvement.

Case Report

A 50-year-old gentleman presented with 2 days history of sudden onset severe holocranial headache and brief loss of consciousness. He presented to us in altered sensorium with left-sided hemiplegia. There was no history of fever, vomiting, giddiness, and speech disturbance. He was a known diabetic on regular medication. He was a known patient of HIV since 6 years not on regular antiretroviral therapy. He was not a hypertensive. He presented to us in altered sensorium, with localizing to pain from right side with left-sided hemiplegia. Detailed neurological assessment revealed oculomotor abnormalities in the form of right horizontal gaze palsy with left INO. His vertical gaze was preserved. He had left facial lower motor neuron paresis with left hemiplegia (▶ **Figs. 1** and **2**). He had a poor gag reflex. He had chest infection and his chest X-ray revealed patchy airspace changes at both lung bases, suggestive of edema with infection. His CD 4 counts revealed 149 cells/ μ L ($N = 441$ – $1,295$ cells/ μ L) and CD 8 counts, 360 cells/ μ L (326–763 cells/ μ L). He was intubated in the emergency in view of low Glasgow coma scale and aspiration. He was started on antiplatelets, antibiotics, statins, and antihypertensives. He was continued on antiretroviral



Fig. 1 (A–E) Range of eye movements in all directions; Ipsilateral horizontal gaze palsy with internuclear ophthalmoplegia (*right*). (F) Facial weakness.

therapy. He underwent tracheostomy in view of poor gag reflex. He had residual deficits, mild residual symptoms = 4 at 90-days follow-up.

Discussion

Nine syndrome is a new variant of eight-and-a-half syndrome which involves ipsilateral abducens nucleus or PPRF, ipsilateral MLF, and adjacent facial colliculus/facial nerve. Clinically, patient presents with complete ipsilateral horizontal gaze palsy with partial contralateral paresis (abduction preserved).³ There is also VII nerve paresis due to the involvement of facial nerve. Pontomedullary lesions are of varied etiology including infarct, hemorrhage, brainstem demyelination, brain stem tumor, and arteriovenous malformation. A possible “nine” syndrome was described by Rosini et al in 2013⁴ which comprised eight-and-a-half syndrome with hemiparesis and hemihypesthesia due to additional involvement of corticospinal tract and medial lemniscus by lacunar pontine infarction. Two cases of nine syndrome were reported by Mahale et al in 2015⁵ with features of

eight-and-a-half syndrome and hemiataxia or hemiparesis in association with neuromyelitis optica and hypertension.

In present case report, patient had clinical feature of nine syndrome in the form of eight-and-a-half syndrome with lesions in medulla and pontine regions with left-sided hemiplegia. He had association with HIV which has a predilection toward small arteries in the brain and to the best of our knowledge, this is the first case report of nine syndrome in a patient with HIV illness.

Conclusion

Nine syndrome is a new variant of eight-and-a-half syndrome which has been diagnosed and reported recently in the literature. Till date, only two case reports are published in English literature. It may have an association with HIV which causes microvasculopathy affecting small vessels. This syndrome represents extension in anterior pontine tegmentum or in medullary region. Early diagnosis of this rare syndrome helps in the localization of lesion to ipsilateral pons and medulla and better clinical outcomes.

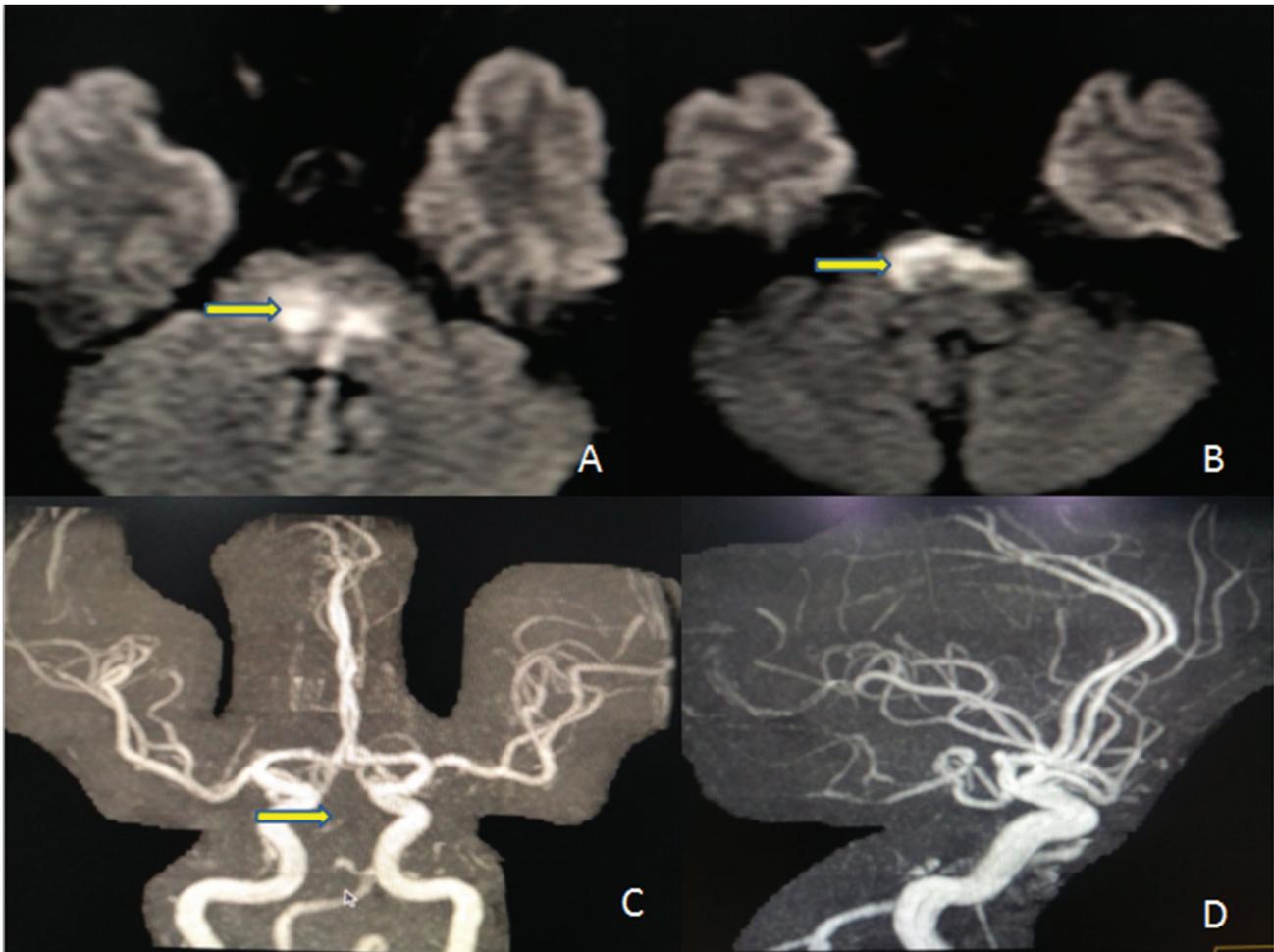


Fig. 2 (A and B) MRI DWI images reveal infarct in pontomedullary region. (C and D) TOF images depicting absent basilar artery with well-preserved PCA, MCA, ACA. ACA, anterior cerebral artery; DWA, diffusion-weighted imaging; MCA, middle cerebral artery; MRI, magnetic resonance imaging; PCA, posterior cerebral artery; TOF, time of flight.

Conflict of Interest

None declared.

References

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