A Rare Case of Nine Syndrome

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Disclosure: The authors report no conflict of interest.

ABSTRACT

Objective: To present a rare case of nine syndrome in a middle-aged patient with stage IV chronic kidney disease presenting with hypertensive emergency at a tertiary government hospital, and to discuss its clinical presentation, diagnostic approach, and management within the realities of limited-resource settings.

Method: This is a case report.

Result: A 45-year-old woman with uncontrolled hypertension and stage IV chronic kidney disease came to the emergency department with acute dizziness and vomiting. On neurologic examination, she had left horizontal conjugate gaze palsy and an adduction deficit of the left eye with intact abduction and an abducting nystagmus of the right eye consistent with one-and-a-half syndrome. In addition, she had left-sided facial weakness and right lower limb weakness suggesting nine syndrome. Brain imaging confirmed a left posterior pontine infarct. Treatment for neuroprotection and blood pressure control led to significant improvement in ocular symptoms and muscle strength.

Conclusion: This report describes a rare case of nine syndrome. Careful bedside neuro-ophthalmologic examination, supported by MRI, enabled accurate localization to the paramedian pons. Early recognition allowed timely stroke management and initiation of secondary management, even in a resource-limited setting.

Keywords: Nine syndrome, one-and-a-half syndrome, hypertensive emergency, neuro-ophthalmology, dorsal pons

Philipp J Ophthalmol 2025;50:108-111

Nine syndrome is a rare neuro-ophthalmologic condition that combines features of one-and-a-half syndrome (OHS) with additional neurologic deficits, specifically ipsilateral facial muscle weakness and either contralateral hemiparesis or ipsilateral hemiataxia. OHS is characterized by conjugate horizontal gaze palsy in one direction and an ipsilateral internuclear ophthalmoplegia (INO). It is usually caused by a lesion in the brainstem affecting the paramedian pontine reticular formation (PPRF) and/or the abducens nucleus and the medial longitudinal fasciculus (MLF) on one side. When combined with an ipsilateral fascicular facial or seventh cranial nerve palsy, the condition is sometimes referred to as eight-and-a-half syndrome. Additional finding of contralateral hemiparesis or ipsilateral hemiataxia due to involvement of the adjacent corticospinal tract or pontocerebellar fibers, respectively, gives rise to the nine syndrome. The rarity and complex neuro-ophthalmologic presentation of nine syndrome requires high clinical suspicion.

In this paper, we describe a rare case of nine syndrome in an adult Filipino female with stage IV chronic kidney disease and uncontrolled hypertension who presented at the emergency department of a tertiary government hospital.

CASE PRESENTATION

We present a 45-year-old female who consulted at the emergency room for acute-onset dizziness and vomiting. Past medical history was positive for stage IV chronic kidney disease and hypertension. On physical examination, patient was conscious, oriented and not in respiratory distress, the blood pressure was 230/120 mmHg and creatinine level was 354.19 umol/L with estimated glomerular filtration rate (eGFR) of 13 mL/min/1.73m^2. On neuro-ophthalmologic examination, visual acuity was 20/20 in each eye. She had full peripheral visual fields on confrontation testing and normal color vision in both eyes. Ocular motility testing showed left horizontal conjugate gaze palsy. On right gaze, the left eye showed limited adduction while the right eye showed full abduction with associated abducting nystagmus (**Figure 1**). The rest of the neurologic exam was unremarkable. One day after the onset of the ocular symptoms, the patient developed a left-sided facial weakness. Motor strength was 5/5 on both upper and lower extremities, and no sensory deficit was noted. A diagnosis of eight-and-a-half syndrome was made. Magnetic resonance imaging (MRI) of the brain revealed hyperintense signals in the left dorsal paramedian pontine tegmentum, extending to the brachium pontis and medulla, on diffusion weighted imaging (DWI) and non-contrast T2-weighted images, consistent with acute infarction (**Figure 2**). The patient was subsequently admitted to the intensive care unit under the neurology service where she received medical management with enoxaparin, citicoline, piracetam, erythropoietin, and nicardipine drip. Monocular occlusion was advised to relieve the diplopia. On the second day of hospitalization, or two days after the onset of ocular symptoms, the patient developed unsteady gait and weakness in the right lower limb, with the muscle strength graded at 4/5. Sensory examination and physiologic reflexes were intact. The new findings supported the diagnosis of nine syndrome. The patient's condition was closely monitored, and antihypertensive medications were maintained to control blood pressure. By the third hospital day, the patient's blood pressure had stabilized and no new symptoms were observed. The patient was subsequently transferred to the regular ward and discharged after five days with maintenance medications.

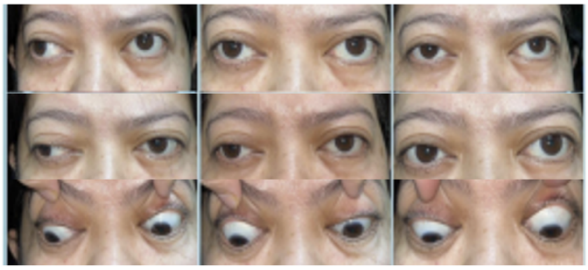

Figure 1. Composite image of the nine cardinal positions of gaze demonstrating left conjugate horizontal gaze palsy and left adduction deficit, with preserved vertical eye movements, consistent with one-and-a-half syndrome.

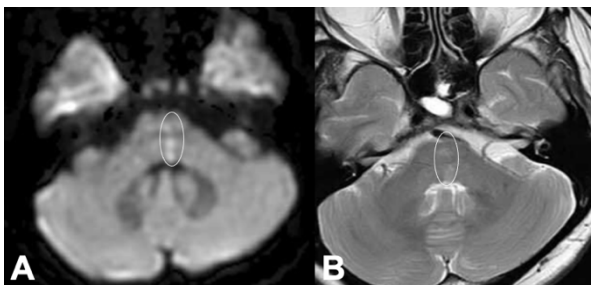

Figure 2. MRI demonstrates hyperintense signals (encircled) in the left dorsal paramedian pontine tegmentum at the level of the brachium pontis, seen on (A) DWI and (B) non-contrast T2-weighted imaging.

DISCUSSION

Nine syndrome is a rare and complex neurologic condition characterized by a constellation of symptoms involving ocular motility, facial nerve function, and motor control. It combines the features of OHS, ipsilateral peripheral facial nerve palsy, and either contralateral hemiparesis or ipsilateral hemiataxia. This clinical triad localizes the lesion to the dorsal paramedian pontine tegmentum, involving the abducens nucleus, medial longitudinal fasciculus (MLF), facial nerve fascicle, corticospinal tract, and pontocerebellar fibers.

In this report, we presented a patient with left conjugate horizontal gaze palsy and a left internuclear ophthalmoplegia, consistent with a left OHS syndrome. Additionally, she had a left peripheral facial nerve palsy, and right-sided motor deficits, collectively consistent with a nine syndrome. Brain MRI confirmed an acute infarct involving the left dorsal paramedian pontine tegmentum at the level of the brachium pontis.

The brainstem is a densely compact structure containing several neurologic pathways in close proximity; thus, pontine strokes frequently affect multiple adjacent structures and produce a combination of neurologic deficits. In our case, involvement of the left abducens nucleus and the adjacent facial nerve fascicle in the dorsal pons resulted in left horizontal conjugate gaze palsy and left facial weakness, respectively. Involvement of the ipsilateral MLF caused the left INO, manifested as loss of adduction of the left eye. The right-sided lower limb weakness was attributable to the involvement of the left corticospinal tract. This clinicoradiologic correlation is similar to previously documented cases of nine syndrome.[1-5] Several case reports have described nine syndrome in the context of pontine infarction. Uysal *et al.* reported a patient who developed transient hemiparesis in the setting of eight-and-a-half syndrome after a pontine infarct.[2] Rosini *et al.* described right horizontal conjugate gaze palsy, internuclear ophthalmoplegia, ipsilateral facial palsy, and contralateral hemiparesis from a lacunar pontine infarct. Similarly, Shen *et al.* and Asham Al Salkhadi *et al.* reported patients with horizontal conjugate gaze palsy, internuclear ophthalmoplegia, ipsilateral peripheral facial nerve palsy, and contralateral limb weakness with ataxia, all attributable to acute ischemic lesions involving the dorsal pontine tegmentum.[4,5]

MRI is the imaging modality of choice for detecting small pontine infarcts and was crucial in confirming the diagnosis in this case. However, in resource-limited settings such as the Philippines, where MRI is not readily available in all centers, alternative imaging modalities which are less costly, such as non-contrast cranial tomography (CT) may aid in the diagnosis.[6]

While ischemic infarction is the most common cause of pontine lesions, other etiologies include hemorrhagic infarction, vascular malformation, demyelinating disorder such as multiple sclerosis, and intrinsic pontine tumors.[5] A complete history, thorough examination, and careful clinical localization are important to guide clinicians in selecting appropriate diagnostic tools and ensuring timely management.

Our patient was managed as a case of acute posterior circulation stroke. She received antithrombotic therapy, a nicardipine infusion for blood pressure control, and neuroprotective agents including citicoline and piracetam, and erythropoietin for anemia. Monocular occlusion was advised to alleviate diplopia. Secondary stroke prevention measures were initiated before discharge, including lipid-lowering medication, strict blood pressure control, optimization of renal function, and patient education on lifestyle changes. These interventions are consistent with current evidence showing that short-term dual antiplatelet therapy combined with intensive medical management lowers the risk of early recurrence in posterior circulation strokes, whereas long-term prevention requires single antiplatelet therapy plus strict risk factor control.[7]

In summary, we reported a rare case of nine syndrome from a posterior circulation ischemic stroke in a patient with chronic kidney disease and hypertensive emergency. A detailed bedside neuro-ophthalmologic examination, supported by MRI findings, localized the lesion to the left dorsal paramedian pontine tegmentum and confirmed the diagnosis. Early recognition allowed timely initiation of acute stroke management and secondary prevention, which are essential to improving functional outcomes. Nine syndrome, though rare, presents with a distinct clinical pattern that should be promptly recognized, even in resource-limited settings, because a careful clinical assessment can guide accurate diagnosis and appropriate treatment.

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