Nine syndrome

Somarajan Anandan[1], Sajeesh S Rajendran[1], Sisira Sree Rajan[2], Joesni Joy[2], Jyothish P Kumar[2]

From [1]Neurologist, [2]Resident, Department of Neurology, St Joseph Hospital, Anchal, Kerala, India

A 65-year-old male with a 3-year history of diabetesmellitus presented to the outpatient department with a 3-day history of swaying to the right while walking and paraesthesia of the right upper limb and face. He developed a deviation of the angle of the mouth to the right with difficulty in spitting for the past 1 day. He is hypertensive and has a history of coronary artery disease for which he underwent stenting 9 years back. He was on double antiplatelets and statins but defaulted for a few weeks. He is a non-smoker. On examination, his blood pressure was 140/80 mm of Hg and his pulse rate was 80/min and was regular. Neurological examination showed an alert person who is oriented in time, place, and person. Extra-ocular movement examination showed an adduction deficit in the left eye with nystagmus in the abducting eye (left INO). There was gaze palsy to the left. There was no ptosis. Pupils were equal and reactive to light. Convergence was preserved. The oculocephalic reflex was impaired. There was no upbeat nystagmus. Vertical eye movements were normal. There was left lower motor neuron type facial palsy. Other cranial nerves were normal. Motor system examination showed right-hand grip weakness. There was no arm drift or lower limb weakness. Corneal reflexes were normal. Deep tendon reflexes were normal bilaterally. Plantars were flexor. Sensory system examination showed impaired fine touch over the right side of the face, right upper limb, and right side of the thorax. Vibration sense was reduced in the right upper limb. Joint position sense and pain sensation were normal in both upper and lower limbs. Pain sensation over the face was intact. There was no finger-nose incoordination, toe-finger incoordination, or tandem ataxia.

The evaluation showed fasting blood glucose of 158 mg%. His HbA1c was 11.2%. He had polycythemia. Other investigations are given in Table 1. Magnetic resonance imaging of the brain showed acute infarct in the left lower paramedian pontine tegmentum (Fig. 1). Magnetic resonance angiography of the brain and neck was normal. His echocardiogram and 72-h Holter study were normal. He was treated with double antiplatelets, statin, and one session of venesection. His symptoms improved partially by the time of discharge.

Access this article online	
Received - 16 October 2024 Initial Review - 28 October 2024 Accepted - 27 November 2024	Quick Response code
DOI: 10.32677/ijcr.v11i1.4866	

Table 1: Blood investigations of the patient

Blood parameters	Result	Normal values
Hemoglobin (%)	17.4 gm	11–14 gm
Packed cell volume (%)	50.7	38.3–48.6
Red cell count	6.01 million/mm^3	4–6 million/mm^3
Total count	9440 cells/mm^3	4000–11000 cells/mm^3
Platelet count	2.14 lakhs/mm^3	1.5–3.5 lakhs/mm^3
ESR	4 mm/h	<33 mm/h
Low-density lipoprotein	106 mg/dL	<100 mg/dL
Triglycerides	230 mg/dL	<150 mg/dL
High-density lipoprotein	35 mg/dL	30–70 mg/dL
Erythropoietin	14.4mIU/mL	5.4–31 mIU/mL

A combination of one-and-a-half syndrome (1½), ipsilateral lower motor neuron (LMN) type facial palsy (7), and contralateral hemiparesis, hemiataxia or hemi-hypoesthesia/hemi anesthesia (1/2) constitutes nine syndrome. It is one of the one-and-a-half spectrum disorders [1]. It's a very rare brainstem syndrome with only a few case reports in the literature [2-4]. It is usually due to stroke and has been reported with infarct and hemorrhage. It has been reported with demyelination and tumors involving the brainstem.

Brainstem syndromes and their eponyms are well known. Unlike these "human" eponyms dedicated to famous neurologists, "numerical" eponyms that describe various disorders at the anatomical level of the brainstem, are less known and are rarely used in clinical practice. In literature, we can find 24 brainstem syndromes that carry human eponyms. There are 4 midbrain-related eponyms, 9 pontine eponyms, and 11 medulla oblongata-related eponyms. The first described brainstem syndrome with a "human" eponym dates back to 1856 (Millard Gubler syndrome). There are many brainstem syndromes with numerical eponyms [5].

Horizontal One-and-a-half syndrome due to involvement of medial longitudinal fasciculus and paramedian pontine reticular formation/abducens nucleus in pons is characterized by ipsilateral gaze palsy and internuclear ophthalmoplegia. Vertical one-and-half syndrome is usually due to thalamo-mesencephalic lesions [6]. Eight-and-a-half syndrome is not an uncommon brainstem syndrome characterized by one-and-a-half syndrome with ipsilateral LMN facial palsy. It is due to the involvement

Correspondence to: Somarajan Anandan, Department of Neurology, St Joseph Hospital, Anchal - 691306, Kerala, India. E-mail: drsomarajan@yahoo.co.in

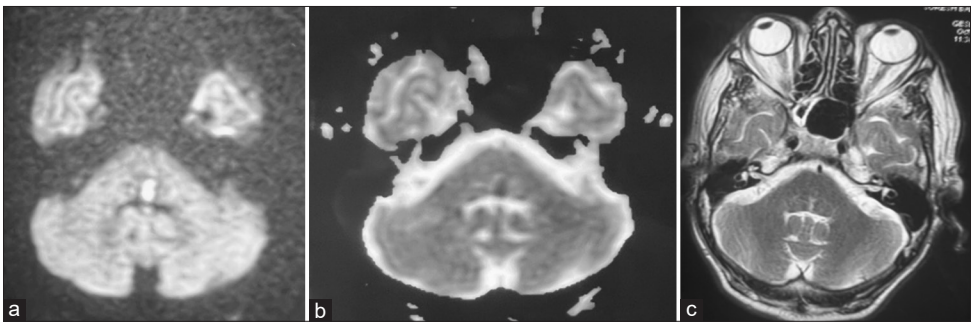

Figure 1: (a) MRI brain axial diffusion-weighted image showing hyperintensity (diffusion restriction) in left lower paramedian pontine tegmentum suggestive of acute infarct; (b) MRI brain axial apparent diffusion coefficient image showing corresponding hypointensity in left lower paramedian pontine tegmentum; and (c) MRI brain axial T2 weighted image showing corresponding hyperintensity

of medial longitudinal fasciculus, paramedian pontine reticular formation (PPRF) or abducens nucleus, and facial nerve fasciculus or facial colliculus. When the lesion extends to involve adjacent structures such as the medial lemniscus or corticospinal tract with resultant contralateral hemi-anesthesia or hemiparesis (1/2), results in Nine syndrome. Mahale *et al*. described two patients with nine syndromes characterized by eight-and-a-half syndrome with hemiataxia [7].

In our case, there was involvement left abducens nucleus, left facial nerve fascicle, left medial longitudinal fasciculus, partial involvement of corticospinal tract and medial lemniscus, and trigeminal lemniscus. Unlike other reported cases, our case had only contralateral upper limb and face involvement.

REFERENCES

1. Xue F, Zhang L, Zhang L, Ying Z, Sha O, Ding Y. One-and-a-half syndrome with its spectrum disorders. Quant Imaging Med Surg 2017;7:691-7.
2. Rosini F, Pretegiani E, Guideri F, Cerase A, Rufa A. Eight and a half syndrome with hemiparesis and hemihypesthesia: The nine syndrome? J Stroke Cerebrovasc Dis 2013;22:e637-8.
3. Singhdev J, Asranna A, Sureshbabu S, Mittal GK, Singla S, Peter S, *et al*. Nine syndrome: Case report and review of clinical signs in internuclear ophthalmoplegia. Ann Indian Acad Neurol 2018;21:325-7.
4. Jesuthasan J, Getheeswaran S, Pirasath S. An unusual presentation of hemiparesis with complex ophthalmoplegia: A "nine" syndrome. SAGE Open Med Case Rep 2022;10:2050313X221135599.
5. Čerimagić D, Bilić E. What's the time?-Numerical eponyms and brainstem syndromes. RAD CASA Med Sci 2021;547=54-5:94-9.
6. Anandan S, Rajendran SS, Kumar JP, Shajee DS, Padmanabhan R. Vertical one-and-a-half syndrome in artery of Percheron infarct. Indian J Case Rep 2024;10:377-8.
7. Mahale RR, Mehta A, John AA, Javali M, Abbas MM, Rangasetty S. "Nine" syndrome: A new neuro-ophthalmologic syndrome: Report of two cases. Ann Indian Acad Neurol 2015;18:335-7.

Funding: Nil; Conflicts of interest: Nil.

How to cite this article: Anandan S, Rajendran SS, Rajan SS, Joy J, Kumar JP. Nine syndrome. Indian J Case Reports. 2025; 11(1):45-46.