

Twenty-Four Syndrome: An Untold Presentation of Pontine Hemorrhage

Ummer Karadan, DM (Neurology), Ramesh N. Supreeth, MBBS,
Robin George Manappallil, MD, and Chellenton Jayakrishnan, DM (Neurology)

Pontine hemorrhages are relatively uncommon. Various atypical manifestations of pontine stroke like eight-and-a-half syndrome, fifteen-and-a-half syndrome, and sixteen syndrome have been described in the past. We came across a case of pontine bleed that presented with bilateral facial palsy, bilateral horizontal gaze palsy, and contralateral sensorineural hearing loss accounting to the hitherto not described “twenty-four syndrome” with Horner’s syndrome and left hemiparesis. **Key Words:** Pontine hemorrhage—facial diplegia—bilateral horizontal gaze palsy—contralateral sensorineural hearing loss.

© 2017 National Stroke Association. Published by Elsevier Inc. All rights reserved.

Introduction

Primary pontine hemorrhages are relatively rare when compared to supratentorial cerebral hemorrhage. Because of the close proximity of pons to the cranial nerve nuclei and vascular anomalies, patients with primary pontine hemorrhages show various atypical manifestations. Our patient presented with facial diplegia, bilateral horizontal gaze palsy, and contralateral sensorineural hearing deficit with Horner’s syndrome and left hemiparesis following pontine hemorrhage.

Case Report

A 75-year-old lady was brought with history of giddiness and fall, followed by slurred speech. She was hypertensive for the past 5 years, on amlodipine 5 mg/day. She was brought to our hospital within 1 hour of

the incident. On the way, she had 2 episodes of nonprojectile vomiting, with vomitus containing food particles. On presentation, she was conscious and oriented, with Glasgow Coma Scale of 15/15. Her heart rate was 100/minute and regular, blood pressure was 220/110 mmHg, respiratory rate 20/minute with saturation 96% in room air. Neurological examination revealed right eye ptosis, right pupil miosis, bilateral horizontal gaze palsy with vertical ocular bobbing and bilateral lower motor neuron facial palsy (right more than left side). Ocular pursuit and saccades were impaired bilaterally in vertical plane. Horner’s syndrome was present. Oculocephalic maneuver and calorie test failed to elicit horizontal ocular movements. Rinne’s test was positive bilaterally with poorer air and bone conduction on left side. Weber’s test showed lateralisation of sound to right ear suggesting left sensorineural hearing loss. She had dysphagia but no palatal palsy or deviation of uvula. Left upper and lower limb had decreased tone and power (4/5 by manual muscle test) with brisk deep tendon reflexes, along with loss of pain, temperature, vibration, and proprioceptive sensations. Left lower limb had extensor plantar reflex. Other systemic examinations were normal. Her blood investigations showed reactive leukocytosis. Her renal and liver functions, electrolytes, calcium, thyroid-stimulating hormone and hemoglobin A1c were normal. Echocardiography showed concentric left ventricular hypertrophy.

From the Baby Memorial Hospital, Calicut, Kerala 673004, India.
Received October 15, 2017; revision received November 22, 2017; accepted December 3, 2017.

Address correspondence to Robin George Manappallil, MD, Internal Medicine, Baby Memorial Hospital, Mini Bypass Road, Calicut, Kerala 673004, India. E-mail: drrobingeorgempl@gmail.com.

1052-3057/\$ - see front matter

© 2017 National Stroke Association. Published by Elsevier Inc. All rights reserved.

<https://doi.org/10.1016/j.jstrokecerebrovasdis.2017.12.007>



Figure 1. CT brain showing a hyperdense lesion in the pons suggestive of hemorrhage. Abbreviation: CT, computed tomography.

Computerized tomography (CT) of brain revealed an acute pontine bleed predominantly on the right and extending to the left (Fig 1). From the clinical presentation and imaging findings, a diagnosis of acute pontine hemorrhage with cranial nerve involvement presenting as a “twenty-four syndrome” along with Horner’s syndrome and left hemiparesis was made. On admission, her National Institutes of Health Stroke Scale (NIHSS) score was 18 and modified Rankin Scale (mRS) was 4. She was managed with antihypertensives, mannitol, and physiotherapy, and was discharged on day 12 of admission with National Institutes of Health Stroke Scale and modified Rankin Scale of 12 and 3, respectively.

Discussion

Cerebrovascular diseases are at a rapid increase in developing countries, with more than 2-fold rise in their incidence between 1970 and 2008. An incidence of 117 per 1 lakh population has been noted in low- and middle-income countries. Intracerebral hemorrhage (ICH) is the second most common cause of stroke, following ischemia, accounting for about 10%-20% of all stroke cases.¹ About 80% of ICH cases are due to hypertensive atherosclerotic cerebrovascular disease and cerebral amyloid angiopathy.² The pathophysiology behind nontraumatic ICH has been attributed to age-related vessel wall degenerative changes, hypertension, and diabetes, resulting in the formation of Charcot-Bouchard micro aneurysms and lipohyalinosis of small arterioles.³

Pontine hemorrhage accounts for 5%-9% of ICH cases, with incidence being highest between 40 and 50 years of age. These patients present with acute onset coma, con-

stricted pupils, quadriplegia, and early onset respiratory arrest and sudden death.⁴

One-and-a-half syndrome is characterized by horizontal gaze palsy (1) with intranuclear ophthalmoplegia (1/2) due to a lesion of the paramedian pontine reticular formation and medial longitudinal fasciculus.⁵ Eight-and-a-half syndrome (7 + 1 + 1/2) was noticed when the lesion also affects the fascicle of ipsilateral facial nerve in the region of the facial colliculus, producing an ipsilateral lower motor neuron facial palsy.⁶ Such a case was reported by Nandhagopal and Krishnamoorthy.⁷ Bae and Song reported a case of fifteen-and-a-half syndrome where the patient had unilateral one-and-a-half syndrome and bilateral lower motor neuron facial palsy (1 + 1/2 + 7 + 7).⁸ A sixteen syndrome characterized by bilateral horizontal gaze palsies and facial diplegia (7 + 7 + 1 + 1) was observed by Connors et al in a case of pontine hemorrhage.⁹

In our case, the patient had predominantly right pontine hemorrhage with extension to the left resulting in facial diplegia, bilateral horizontal gaze palsy, and contralateral sensorineural hearing deficit, making it to a twenty-four syndrome (7 + 7 + 1 + 1 + 8) along with Horner’s syndrome and left hemiparesis. The contralateral sensorineural hearing loss was probably due to involvement of lateral lemniscus after its decussation at the mid pons. Expansion of the hemorrhage or perilesional edema crossing midline would have caused the lateral lemniscal involvement. To the best of our knowledge, such a scenario has not been reported yet.

References

1. Feigin VL, Lawes CM, Bennett DA, et al. Worldwide stroke incidence and early case fatality reported in 56 population-based studies: a systematic review. *Lancet Neurol* 2009;8:355-369.
2. Sutherland GR, Auer RN. Primary intracerebral hemorrhage. *J Clin Neurosci* 2006;13:511-517.
3. Fisher CM. Pathological observations in hypertensive cerebral hemorrhage. *J Neuropathol Exp Neurol* 1971;30:536-550.
4. Silverstein AJ. Primary pontine haemorrhage. In: Vinken PJ, Bruyn GW, eds. *Handbook of clinical neurology, vascular diseases of nervous system*. Amsterdam: North Holland Inc, 1972:37-53.
5. Fisher CM. Some neuro-ophthalmological observations. *J Neurol Neurosurg Psychiatry* 1967;30:383-392.
6. Eggenberger E. Eight-and-a-half syndrome: one-and-a-half syndrome plus cranial nerve VII palsy. *J Neuroophthalmol* 1998;18:114-116.
7. Nandhagopal R, Krishnamoorthy SG. Eight-and-a-half syndrome. *J Neurol Neurosurg Psychiatry* 2006;77:463.
8. Bae JS, Song HK. One-and-a-half syndrome with facial diplegia: the 15 1/2 syndrome? *J Neuroophthalmol* 2005;25:52-53.
9. Connors R, Ngan V, Howard J. A case of complete lateral gaze paralysis and facial diplegia: the 16 syndrome. *J Neuroophthalmol* 2013;33:69-70.