

Unilateral asterixis, thalamic astasia and vertical one and half syndrome in a unilateral posterior thalamo-subthalamic paramedian infarct: An interesting case report

Subasree Ramakrishnan, Veera Rajkumar Narayanaswamy¹

Department of Neurology, National Institute of Mental Health and Neurosciences, Bangalore, Karnataka, ¹Department of Neurosciences, New Vikram Hospital, Madurai, Tamil Nadu, India

ABSTRACT

A 42-year-old young lady presented with acute onset of dizziness, drooping of left eye with binocular diplopia and inability to walk unassisted. She had past history of uncontrolled diabetes mellitus and hypertension. On examination, she had left fascicular type of third nerve palsy, vertical one and half syndrome (VOHS), left internuclear ophthalmoplegia and skew deviation with ipsilesional hypertropia. She also had thalamic astasia and right unilateral asterixis. Her MRI revealed T2 and Flair hyper intense signal changes with restricted diffusion in the left thalamus, subthalamus and left midbrain. MR Angiography was normal. Thalamic-subthalamic paramedian territory infarct is relatively uncommon. It can present with oculomotor abnormalities including vertical one and half syndrome, skew deviation, thalamic astasia and asterixis. This case is reported for the rarity of the presenting clinical findings in unilateral thalamo-mesencephalic infarcts.

Key words: Thalamic astasia, thalamo-mesencephalic infarct, unilateral asterixis, vertical one and a half syndrome

Introduction

The posterior thalamo-subthalamic paramedian artery is often unpaired and originates from P1 segment of posterior cerebral artery. It supplies the paramedian part of the upper midbrain and of the thalamus (intralaminar nuclei, dorsomedial nucleus and internal part of the ventral posterior nucleus). Unilateral thalamic and midbrain infarcts are uncommon.

Decreased consciousness, behavioral changes, memory impairment, motor neglect, facio brachial tactile hypoesthesia, up gaze palsy, VOHS, Weber and Claude third nerve fascicular syndromes, loss of convergence, pseudo-sixth nerve palsy, bilateral

internuclear ophthalmoplegia, miosis and bright light intolerance, asterixis and delayed movement disorders have been described in posterior thalamo-subthalamic paramedian arterial infarct.^[1] Unilateral asterixis though relatively uncommon are associated with acute structural lesions and focal cerebral lesions. Patients with thalamic infarct can manifest with Thalamic Astasia.

Case Report

A 42-year-old young lady presented with 3 days history of acute onset of dizziness, transient loss of consciousness, sudden drooping of left eyelid and binocular diplopia with difficulty in walking unassisted. She also had hypersomnolence. There was history of hypertension and uncontrolled diabetes mellitus of 2 years duration. Her admission BP was 210/120 mm hg and her blood sugar was 283 mg/dl with urine acetone positive at admission. She had past history of migraine for several years. Systemic examination was normal. Higher mental functions were normal. She had complete ptosis with dilated 5 mm nonreactive pupil on left side. There was left internuclear ophthalmoplegia and skew

Access this article online

Quick Response Code:



Website:
www.ruralneuropractice.com

DOI:
10.4103/0976-3147.112775

Address for correspondence:

Dr. Subasree Ramakrishnan, Department of Neurology, NIMHANS, Bangalore, Karnataka, India. E-mail: subasree.ramakrishnan@gmail.com

deviation with left hypertropia. There was also bilateral up gaze palsy with monocular left down gaze palsy and the only possible vertical movement was downward movement of right eye suggestive of vertical one and half syndrome. There was bilateral horizontal gaze evoked nystagmus. Convergence was impaired. Bell's phenomenon and vertical oculocephalic movement was absent. The muscle strength, sensory system, finger nose and knee heel coordination were normal. Reflexes were normal and plantars flexors. She was not able to sit or stand without support. She had thalamic astasia, was leaning back while sitting and required support to walk. On outstretched prone arm, dorsiflexed wrist, she had right asterix which comprised of semi rhythmic flexion movements at wrist and fingers.

On evaluation, biochemistry, lipid profile and blood counts were normal. Cardiac evaluation was normal. MRI imaging showed evidence of T1 hypo intense and T2, Flair hyper intense signal changes in left anterior thalamus, subthalamus, left midbrain including periaqueductal region [Figure 1]. There were also bilateral frontal caps, bilateral asymmetric fronto parietal sub cortical white matter and symmetric peritrigonal white matter signal changes. Magnetic Resonance angiogram was normal and diffusion studies showed restriction [Figures 2 and 3]. There were no hemorrhage or micro bleeds. Patient was started on anti-platelets, anti-edema measures, antihypertensive and insulin. Ptosis, diplopia, dizziness and ataxia started improving gradually over next 2 days.

Discussion

Midbrain lesions may give rise to the most complex eye movement disorders observed in clinical neurology. The neural structures known to be involved in the control of vertical gaze includes the nucleus of Darkschewitsch, the interstitial nucleus of Cajal, rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) and the posterior commissure. Various Neuro-ophthalmic findings like vertical gaze palsy, VOHS, Weber syndrome, nuclear third nerve palsy, seesaw nystagmus, skew deviation are seen due to involvement of these midbrain centres.^[1]

The arterial supply of the structures involved in the supranuclear control of vertical gaze depends on the posterior thalamo-subthalamic arteries and the paramedian peduncular arteries. The frequent coexistence of both midbrain and paramedian thalamic infarction is related to their vascular supply; a single vessel arising near the top of the basilar may branch to supply both the paramedian region of the thalamus and the rostral

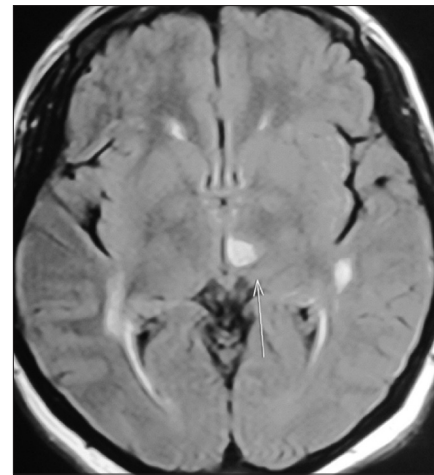


Figure 1: FLAIR Image of left thalamic infarct



Figure 2: Normal MR Angiography

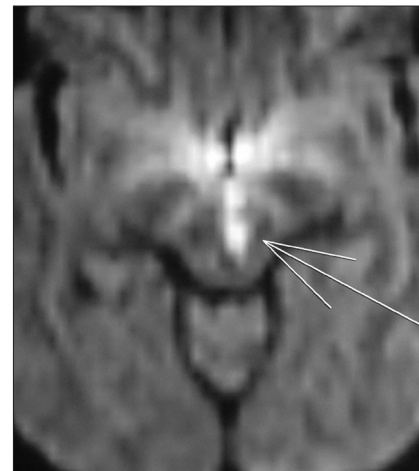


Figure 3: Diffusion image of left midbrain infarct

medial mesencephalon. Vertical gaze palsies are common manifestations of unilateral or bilateral paramedian thalamic infarctions. These gaze palsies have been attributed to lesions in the rostral midbrain rather than the thalamic injury. Unilateral lesion of the upper midbrain

tegmentum is sufficient to generate an up gaze palsy or a combined up and down gaze palsy, whereas an isolated down gaze palsy requires bilateral lesions.^[2,3]

Dysconjugate vertical gaze syndromes includes VOHS, skew deviation and variants, monocular up gaze palsy, ocular tilt reaction, slowly alternating skew deviation, see-saw nystagmus, and "V" pattern pseudo bobbing.^[4] The VOHS consists of paralysis of upward vertical conjugated gaze and monocular paralysis of downward gaze or vice-versa. It occurs as a consequence of a mesencephalodiencephalic lesion, either unilaterally or bilaterally. In a case report of thalamo-mesencephalic infarct with VOHS which evolved subsequently into vertical gaze palsy, it was suggested that in VOHS, pathway to contralateral down gaze neurons could have been damaged due to the unilateral dorsal midbrain lesion before its decussation with the unilateral interstitial nucleus of Cajal, the oculomotor nucleus and the riMLF.^[5] Unilateral lesion destroy the fibres of the posterior commissure and the descending fibres to the ipsilateral subnucleus of the inferior rectus and contralateral subnucleus of the superior oblique just after they decussate, probably above the level of the third nerve nucleus.^[6]

A similar case of unilateral thalamo-mesencephalic infarct was reported with VOHS, Weber syndrome, fascicular third nerve palsy and pseudo-abducens palsy. But there was no astasia and asterixis.^[7] Three cases of posterior thalamo-subthalamic paramedian artery infarct with vertical ophthalmoplegia, impaired convergence and altered mental status were reported. None had VOHS.^[8] In a series of 38 patients of paramedian thalamic artery infarcts, posterior thalamo-subthalamic paramedian artery territory involvement was reported in 89%. Neurological manifestations were somnolence (87%), hemi syndromes (79%), cognitive deficits (58%), oculomotor nerve palsies (53%) and vertical gaze palsy (39%).^[9] In a report of 11 patients with vertical gaze palsy in unilateral midbrain stroke, 3 had VOHS. None had associated astasia and asterixis.^[10]

Thalamic astasia which is inability to stand unassisted in absence of motor weakness and cerebellar dysfunction is documented after unilateral thalamic lesions.^[11] It is attributed to supero posterolateral thalamic involvement. Fastigial fibers of the vestibulocerebellar pathway project to the medial ventrolateral nucleus of the thalamus and damage to this pathway may be responsible for thalamic astasia.^[12] Patient cannot maintain a stationary sitting or standing position and has no other conditions that could account for contralateral pulsion such as pyramidal or sensory deficits, kinaesthesia, abnormal proprioceptive sensation, cerebellar ataxia, psychosis or Parkinsonism.

The gait differs from cerebellar ataxia and there is no wide base or lurching. The Romberg sign is negative and finger nose, knee heel and shin tap tests are normal.

Asterixis manifests as bilateral flapping tremor at the wrist, metacarpophalangeal, and hip joints. It is characterized by brief, arrhythmic interruptions of sustained voluntary muscle contraction usually due to brief lapse of posture. Unilateral asterixis can be seen due to focal brain lesions in the genu and anterior portion of the internal capsule, midbrain, medial frontal cortex, parietal cortex or ventrolateral thalamus.^[13-16] Interruption of the posture pathway in the rostral reticular formation and abnormal joint proprioception are suggested.^[13] The postural stability of the extremities is related to multiple brainstem-spinal pathways. The ventro-lateral nucleus of the thalamus is the area in which cerebello-rubral or vestibulocerebellar fibres converge.^[11] The exact mechanism underlying asterixis remains unclear. Diffuse, widespread derangement of CNS function, abnormal activity in the motor field in the cerebral cortex demonstrated using silent period locked averaging method, dysfunction of the sensorimotor integration occurring in the contralateral parietal lobe and midbrain, a failure in arm posture maintenance that is comparable to failure in leg posture control in patients with astasia are all postulated.^[17]

Conclusion

Unilateral posterior thalamic-subthalamic paramedian infarct is rare. It can present with complex neuro-ophthalmic findings including vertical one half syndrome, thalamic astasia and unilateral asterixis. This case is reported for the rarity and unique nature of the presenting clinical features in unilateral thalamo-mesencephalic infarct. This association of findings is not reported in literature.

References

1. Bogousslavsky J, Regli F, Assal G. The syndrome of unilateral tuberothalamic artery territory infarction. *Stroke* 1986;17:434-41.
2. Pierrot-Deseilligny C. Nuclear, internuclear, and supranuclear ocular motor disorders. *Handb Clin Neurol* 2011;102:319-31.
3. Alemdar M, Kamaci S, Budak F. Unilateral midbrain infarction causing upward and downward gaze palsy. *J Neuro Ophthalmol* 2006;26:173-6.
4. Bogousslavsky J. Oculomotor syndromes resulting from mesencephalic lesions in man. *Rev Neurol* 1989;145:546-59.
5. Suzuki K, Odaka M, Tatsumoto M, Miyamoto T, Takamatsu K, Hirata K. Case of unilateral thalamo-mesencephalic infarction with enlargement to bilateral vertical gaze palsy due to vertical one-and-a-half syndrome. *Brain Nerve* 2008;60:92-6.
6. Bogousslavsky J, Regli F. Up gaze palsy and monocular paresis of downward gaze from ipsilateral thalamo-mesencephalic infarction: A vertical "one-and-a-half" syndrome. *J Neurol* 1984;231:43-5.
7. Deleu D, Imam YZ, Mesraoua B, Salem KY. Vertical one-and-a-half

- syndrome with contralesional pseudo-abducens palsy in a patient with thalamo mesencephalic stroke. *J Neurol Sci* 2012;312:180-3.
8. Wall M, Slamovits TL, Weisberg LA, Trufant SA. Vertical gaze ophthalmoplegia from infarction in the area of the posterior thalamo-subthalamic paramedian artery. *Stroke* 1986;17:546-55.
 9. Weidauer S, Nichtweiss M, Zanella FE, Lanfermann H. Assessment of paramedian thalamic infarcts: MR imaging, clinical features and prognosis. *Eur Radiol* 2004;14:1615-26.
 10. Hommel M, Bogousslavsky J. The spectrum of vertical gaze palsy following unilateral brainstem stroke. *Neurology* 1991;41:1229-34.
 11. Masdeu JC, Gorelick PB. Thalamic astasia: Inability to stand after unilateral thalamic lesions. *Ann Neurol* 1988;23:596-603.
 12. Solomon DH, Barohn RJ, Bazan C, Grissom J. The thalamic ataxia syndrome. *Neurology* 1994;44:810.
 13. Young RR, Shahani BT. Asterixis: One type of negative myoclonus. *Adv Neurol* 1986;43:137-56.
 14. Stell R, Davis S, Carroll WM. Unilateral asterixis due to a lesion of the ventrolateral thalamus. *J Neurol Neurosurg Psychiatry* 1994;57:116-8.
 15. Wee AS. Unilateral asterixis: Case report and comments. *Eur Neurol* 1986;25:208-11.
 16. Klos KJ, Wijdicks EF. Unilateral asterixis after thalamic hemorrhage. *Neurology* 2006;66:E11.
 17. Gokula RM, Khasnis A. Asterixis. *J Postgraduate Med* 2003;49:272-5.

How to cite this article: Ramakrishnan S, Narayanaswamy VR. Unilateral asterixis, thalamic astasia and vertical one and half syndrome in a unilateral posterior thalamo-subthalamic paramedian infarct: An interesting case report. *J Neurosci Rural Pract* 2013;4:220-3.
Source of Support: Nil. **Conflict of Interest:** None declared.