

A case of lateral medullary syndrome with unilateral selective thermoanaesthesia

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DOI: <https://doi.org/10.17511/ijmrr.2019.i01.09>


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A 42-year-old female was admitted to tertiary care teaching hospital with history of headache, vertigo, difficulty in swallowing both liquids and solids, vomiting, gait ataxia, drooping of left eyelid, inability to feel hot and cold on right side of body and diplopia of forty-fivedays' duration. Clinical examination and neuroimaging were suggestive a posterior circulation stroke, with lateral medullary syndrome. The patient had selective thermoanaesthesia on the right side, including the face, which is an atypical finding, given the clinical setting.

Keywords: Stroke, CVA, Lateral medullary syndrome, Hypoplasia, Vertebral, Thermoanaesthesia

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Arun Tyagi, Professor & HOD, Department of Medicine, DVVPF's Medical College, Ahmed Nagar, Maharashtra, India. Email: aruntyagidr@gmail.com	Mujeeb VR, Tyagi A, Jambunathan P. A case of lateral medullary syndrome with unilateral selective thermoanaesthesia. Int J Med Res Rev. 2019;7(1):50-53. Available From https://ijmrr.medresearch.in/index.php/ijmrr/article/view/1035	

Manuscript Received
2019-02-04

Review Round 1
2019-02-14

Review Round 2
2019-02-18

Review Round 3

Accepted
2019-02-23

Conflict of Interest
No

Funding
Nil

Ethical Approval
Yes

Plagiarism X-checker
6%

Note



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Introduction

Selective thermoanaesthesia is a rare manifestation in clinical neurology, per se and are generally found in thalamic strokes. However, when present in the context of a lateral medullary syndrome, is rarer still. We report an unusual case of lateral medullary syndrome presenting with thermoanaesthesia. A forty-two years old female patient, with a 10-year history of left sided unilateral headache, averaging one to two episodes per month, presented with multiple neurological deficits of recent onset. Evaluation through examination and neuroimaging revealed a posterior circulation stroke in the PICA territory.

Case Report

The patient had been apparently asymptomatic a month and a half back when she developed headache, localised to right occipital region, radiating to right frontal region. Ten days later, the headache increased in intensity and was associated with projectile vomiting, vertical diplopia, swallowing difficulty and inability to feel hot and cold on right side of the body. The patient drew a line through her midline, demonstrating that she was unable to appreciate hot or cold on the right side.

There was also a tendency to fall on the left side. The patient had been having intermittent headache since 2003 and had been taking over the counter medication. Examination revealed absent radial and brachial pulses on the left side, with a normal radial pulse on right side. Dorsalis pedis on both sides were feeble. Motor examination revealed reduced tone on the left side. However, the power was grade 5/5 in all limbs. The patient had cerebellar signs on the left side including dysdiadochokinesia. Motor ataxia with tendency to fall on the left was present. Nystagmus was absent and ocular examination was normal. CNS examination revealed a left sided Horner's deviated uvula (towards right), impaired gag reflex, hemisensory loss involving only temperature on the right half of body, including face.

Fine touch, crude touch, vibration and pain were preserved. Urgent computed tomography (CT) of head showed a wedge-shaped hypodense area with prominent adjacent folia and fourth ventricle and the left cerebellar hemisphere.

CT angiography revealed a hypoplastic left vertebral artery, with right vertebral artery dominance. Multiple collaterals were visualised. The anterior circulation showed no significant abnormality. Magnetic resonance imaging (MRI) showed wedge shaped area of attenuated signal intensity in left dorsolateral medulla, which appeared hypointense in T1 and hyperintense in T2 and FLAIR images (Fig. 1 and 2).

Colour Doppler flow imaging (CDFI) of upper extremity showed intraluminal arterial thrombi in left brachial and left radial artery. An impression of chronic infarct in left dorsolateral medulla involving left cerebellar hemisphere was made. ESR and CRP were within normal limits. Evaluation for vasculitis (ANA, cANCA and p-ANCA) and other prothrombotic conditions, such as protein C and S deficiency, prothrombin G20210 mutation and MTHFR mutation was negative.

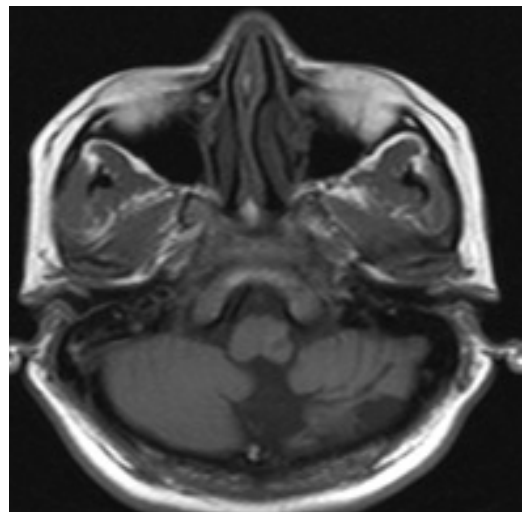


Fig-1

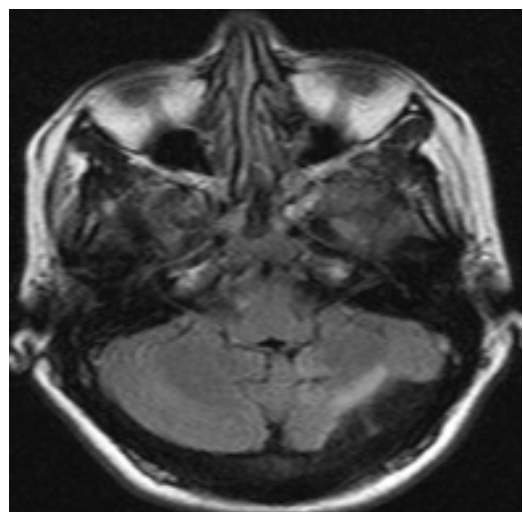


Fig-2

Fi-1: T1 weighted showing atrophy of left cerebellar hemisphere. Hypointense areas are seen in the left medulla and left cerebellar hemisphere.

Fig-2: FLAIR image showing atrophy of left cerebellar hemisphere. Hyperintense areas are seen in the left medulla and left cerebellar hemisphere.

A clinical diagnosis of posterior circulation stroke and lateral medullary syndrome in a patient with migraine without aura, was made. The patient was managed by a multi-disciplinary team comprising of physician, neurologist and haematologist with anti-platelet measures and statins

Discussion

Selective thermoanaesthesia is a rare manifestation in clinical neurology, per se and are generally found in thalamic strokes [1-3]. However, when present in the context of a lateral medullary syndrome, is rarer still. Rare and atypical presentations and associations of lateral medullary syndrome have been reported in literature.

These include a AV malformation with an extra-cranial PICA [4], lateral medullary stroke with epicrania fugax [5], aneurysmal lateral medullary stroke [6], lateral medullary stroke with vestibulopathy [7], sarcoidosis presenting as lateral medullary stroke [8] and central hypoventilation with a lateral medullary stroke [9].

In our case, the MRI suggested no pathology in the thalamus. In view of multiple thrombi in the vasculature, a clinical thermo-specific variant of *Dejerine and Roussy* is possible [10-12]. However, such conjecture is refuted by normal FLAIR imaging, which would have suggested some involvement, through occlusive infarct, in the area supplied by the thalamogeniculate arteries. The multifocal nature of the patient's lesion is nearly certain.

These range from the CN III papillary sympathetic to lower cranial nerves as part of the nucleus ambiguus in the dorsolateral medulla and from the vestibulospinal tracts to the arch-cerebellum and its connections.

This widespread area of involvement is seen to correlate both clinically and radiographically.

However, in the absence of an obvious thalamic focus, unilateral selective thermal anaesthesia remains an enigma.

Conclusion

A rare presentation in an otherwise typical case of lateral medullary syndrome has re-introduced the enigmatic concept of hemi-sensory loss. In traditional neurological practice, this phenomenon continues to remain sparse and under reported. Since the event was vascular and sudden in onset, a differential diagnosis of pseudo-multiple sclerosis, which may, in part explain the described symptoms, seems unlikely [13].

Even with evidence from functional MRIs, other areas (except thalamus) which exert master control over the sensory system are yet to be identified. Our review suggests that a nearly similar case with hemi-hyperhidrosis (not hemisensory loss) was described in 1995 [14]. Another case report suggests isolated thermo anesthesia after a mid-lateral medullary infarction [15].

Yet another report, suggests a putaminal haemorrhage that may have disrupted the thalamocortical connection in the secondary somatosensory cortex [16].

Such atypical presentations are the constant reminders that while we have mapped areas and functions of the brain with reasonable certainty, much of the human brain and its intricate architecture remains a mystery.

Patient's Perspective: The patient experienced complete resolution of dysphagia and was started on oral feeds. She had partial resolution of ptosis and vertigo. However, her thermal hemi sensory loss remained status quo. She was discharged and advised frequent follow up.

Consent-A comprehensive written consent was obtained from the patient for the purposes of this publication.

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