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Atypical Presentation of Medullary Syndrome in a Young Woman

Geetanjali Sharma[1*]

[1]Department of Physiology, Pandit Bhagwat Dayal Sharma Post Graduate Institute of Medical Sciences, University of Health Sciences, Rohtak, Haryana, India.

Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

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Case Report

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ABSTRACT

Wallenburg syndrome occurs due to damage to lateral segment of the medulla. Medial medullary syndrome occurs due to damage to upper portion of the medulla. I report a case of a 30 years old woman diagnosed with medullary syndrome [both medial & lateral features] in absence of CT scan findings sent to the department of Physiology for electrophysiological tests like nerve conduction studies, blink reflex, brainstem auditory evoked potential (BERA) & visual evoked potential (VEP). She had loss of sensations on the ipsilateral half of face (right), hemisensory loss on contralateral trunk & extremities, headache, contralateral hemiparesis (left), ipsilateral lingual paresis with atrophy, fibrillations with contralateral positive Babinski's sign. The electrophysiological tests showed decrease in conduction velocity of right facial nerve, left tibial & peroneal nerves with decreased amplitude. The nerve conduction studies of median nerve (both motor & sensory) were normal. In blink reflex, latency of R2 ipsilateral & R2 contralateral of supraorbital nerves were increased on right side. There were increased latencies of waves II, III, IV & V of BERA & increased P100 latencies in VEP.

The results of the electrophysiological tests of the patient showed that she had features of both medial & lateral medullary syndrome suggesting a lesion of both upper & middle medulla. The CT scan in this case was normal but conduction of MRI & CT guided angiography of posterior cerebral & vertebral arteries could have further localized the lesion causing this mixed symptomology.

**Corresponding author: E-mail: drgeeta1212@yahoo.com;*

Keywords: Medullary syndrome; electrophysiological tests; BERA; VEP; blink reflex.

1. INTRODUCTION

Wallenburgs syndrome or lateral medullary syndrome also called as posterior inferior cerebellar artery syndrome occurs due to damage to the lateral segment of the medulla posterior to the inferior olivary nucleus. It is the most typical posterior circulation ischemic stroke syndrome [1,2].

It is caused most commonly by atherothrombotic occlusion of the vertebral artery followed by posterior inferior cerebellar artery and then the medullary arteries. The risk factors are hypertension, smoking, diabetes, vertebral artery dissection, neck manipulation and injury (in young patients), Marfan syndrome, Ehlers Danlos syndrome & fibromuscular dysplasia [3,4].

Following deficits may be found in lateral medullary syndrome:

On the side of lesion- vertigo with nystagmus (due to involvement of nuclei of IX & X cranial nerves); Horner Syndrome (due to involvement of sympathetic fibres); ipsilateral ataxia (involvement of inferior cerebellar hemisphere, spinicerebellar fibres & inferior cerebellar peduncles); pain & numbness with impaired facisl sensation (involvement of descending trigeminal tract) and impaired taste sensation (involvement of nucleus tractus solitarius).

On the contra lateral side: impaired pain & temperature sensation in arms & legs (involvement of spinothalmic tract). There is none or minimal weakness of the contralateral side (corticospinal fibres are ventral in location).

More rostral lesions tend to be more ventrally located & patients present with marked dysphagia & dysphonia due to involvement of nucleus ambiguous. More caudal lesions involve more dorsolateral structures & these patients present with vertigo, ataxia, nausea, vomiting & Horner syndrome [5,6,7].

In Babinski Nageotte syndrome, lateral medullary syndrome is associated with hemiplegia or hemiparesis due to involvement of pyramidal tract which is affected before decussation [8].

Dejerine proposed a triad of symptoms of medial medullary syndrome which includes contralateral hemiplegia sparing the face, contralateral loss of deep sensation & ipsilateral hypoglossal paralysis [9].

Pathological examination first conducted in 1937 demonstrated thrombotic occlusion of the anterior spinal artery and adjacent vertebral artery [10].

Here I report a case of a young female diagnosed clinically as medullary syndrome with features of both medial & lateral medullary syndrome which was referred from the medicine OPD to the department of Physiology for electrophysiological tests like nerve conduction studies, blink reflex, brainstem auditory evoked potentials and visual evoked potentials.

2. CASE REPORT

The patient was a 30 years old female with complaints of difficulty in chewing food and weakness in limbs for the last three years. The patient was apparently healthy before that. The initial symptoms were persistent headache for 3 months followed by sudden weakness of right side of face and left leg, improper movements of the tongue & face, difficulty in chewing of food, sensation loss on right side of mouth, difficulty in speech & numbness on right side of face. All the complaints were progressive in nature.

Simultaneously the patient suffered from weakness in left leg which was gradually progressive, limping during walking, unable to bear body weight, sudden falls during walking with decreased sensations in the left leg and right side of face.

There was history of fall in blood pressure, no history of trauma, palpitations, Myocardial insufficiency, breathlessness. There was no history of oral contraceptive drug intake or any major surgery. There was no history of double vision, hoarseness of voice, hypertension, fluctuating blood sugar and symptoms of chronic obstructive pulmonary disease.

Family history: The patient was a widow, she had two children 16 & 13 years of age.

Personal history: She was a non alcoholic, non smoker and was a vegetarian.

On clinical examination: She was conscious, cooperative, well oriented in place, time and person.

She has dysarthria, she was apprehensive, afebrile. The blood pressure was 130/80 mm of Hg, pulse was 78/minute, no cyanosis, jugular venous pressure was normal.

Cardiovascular system: Both heart sounds were normal, no murmur was present. The peripheral pulsations were normal. There was no carotid bruit.

Respiratory system: Chest sounds were normal.

Per abdomen: The abdomen was soft on palpation.

Central nervous system: Conscious, well oriented, normal behavior.

Cranial nerve examination:

I: within normal limits
II: within normal limits
III, IV and VI: within normal limits
V & VII: wasting bilateral temporalis, masseter, pupillary reflex were normal. Jaw jerk was normal.

Sensory loss:

Touch & pain:

Right	Left
a. Ophthalmic: Decreased sensation	Normal
b. Maxillary: Decreased sensation	Normal
c. Mandibular: Decreased sensation	Normal

VIII Nerve: within normal limits
IX, X and XI: within normal limits
XII: Tongue movement: Left was normal, right was decreased. The patient wasn't able to protrude the tongue; wasting of tongue was present, fasciculation was present.

No abnormality was detected in the autonomic system functions.

Investigations: Routine haemogram, lipid profile, Kidney Function tests & Liver Function tests were all normal.

CT Scan Head: Cerebellar hemisphere were normal. Basal Ganglia, both ventricles, basal cisterns and calvarium were normal.

Table 1. Showing detailed CNS examination

S. no			Right		Left	
			Upper limb	Lower limb cm	Upper limb	Lower limb
1.	Bulk	Proximal	Normal	Normal 37 cm	Normal	Decreased 31 cm
		Distal	Normal	Normal 20	Normal	Decreased 18
2	Tone		Normal	Normal	Normal	Decreased
3.	Power		5/5	5/5	5/5	Proximal 4/5 Distal 2/5
4.	Reflexes		2+	1+	2+	1+
	Bilateral Plantar reflex silent					

Table 2. Showing detailed sensory examination

Sr. no	Examination	Right	Left
1.	Touch	Decreased on right face	Decreased in left lower limb
2.	Pain	Decreased on right face	Decreased in left lower limb
3.	Vibrations	Normal	Decreased on left side
4.	Position	Normal	Impaired on left side

Table 3. Cerebellar signs present in the patient

S. no	Cerebellar signs	Status on examination
1.	Nystagmus	Absent
2.	Finger nose test	Normal
3.	Dysdiadokinesia	Absent
4.	Dysarthria	Present
5.	Dysmetria	Absent
6.	Romberg's sign	Positive; Babinski's sign positive
7.	Gait	Walking with limp

Table 4. Conduction velocity in upper & lower limbs

S. no	Nerve	Conduction velocity (m/s)	Latency (m/s)	Amplitude (milli volts)
	Motor			
1.	Lt. Median	58.6	6.77	8.1
2.	Rt. Median	58.08	5.73	11.8
3.	Rt. Facial	27	2.92	2.1
4.	Lt. Facial	32	2.9	2.4
5.	Rt. Peroneal	40.54	9.38	2.6
6.	Lt. Peroneal	19.72	20.94	65.7 μ volt
7.	Rt. Tibial	41.72	11.35	1.7mv
8.	Lt. Tibial	30.2	12.29	1mv
	Sensory			
1.	Lt. Median	50.69	2.17	58.4 μ volt
2.	Rt. Medial	47.87	1.88	47.87 μ volt

Table 5. Blink reflex

Side	Latencies		
	R1 (ipsilateral)	R2 (ipsilateral)	R2c (contralateral)
Right	9.8	42.4	42.2
Left	9	37	38

Table 6. Brainstem evoked potential (BERA)

S. no	Wave	Left latency	Right latency
1.	I	1.96	1.98
2.	II	3.17	2.54
3.	III	4.31	3.81
4.	IV	6.02	4.50
5.	V	7.29	5.77
6.	Inter peak latency: I-III	2.35	1.83
7.	I-V	5.33	3.79
8.	III-V	2.98	1.96

Table 7. Visual evoked potential (VEP)

Left			Right		
N75 (m/s)	P100 (m/s)	N145 (m/s)	N75 (m/s)	P100 (m/s)	N145 (m/s)
76.3	111.9	156.3	77.5	116.3	150.6

3. RESULTS

- Clinical features of both medial & lateral medullary syndrome were present in this patient.
- Motor conduction velocities as well as amplitude of right facial, left peroneal & left tibial nerves were reduced.
- Sensory conduction velocities of bilateral median nerve were normal.
- In Blink reflex of right orbital nerve, latency of R2 (ipsilateral) & R2 (contralateral) orbital nerves were increased. They were normal on left side.
- BERA showed increased latency of waves 2, 3, 4 & 5.
- VEP showed increased P100 latency.

4. DISCUSSION

This patient showed both the features of lateral & medial medullary syndromes. The features of lateral medullary syndrome were loss of sensation over ipsilateral half of face, hemisensory loss on contralateral trunk & extremities, ipsilateral cerebellar signs & symptoms. These findings were similar to those of Kim H [11], Battel [12], Kim JS [13] and Cidad [14].

The features of medial medullary syndrome in the patient were headache, contralateral hemiparesis, ipsilateral lingual paresis with atrophy and fibrillations with contralateral Babinski sign. These features were similar to the findings of Kim JS [15].

This study suggests that both upper & middle medulla may be affected in this patient which was similar to the study as reported by Kameda et al. [16].

The hypoglossal nerve was also affected in this patient. The proximal cisternal portion of the 12th cranial nerve is mainly supplied by branches of the vertebral artery. The vertebral artery's dissection is an important cause of posterior circulatory ischemia particularly in young & middle aged patients [17] as was the case with this patient who was young.

The CT scan of the brain of the patient didn't show any abnormality. The cause of this may be the fact that CT scan gives suboptimal visualization of the posterior fossa structures due to obscuration by artefacts (bony structures) and early ischemic changes may not be visible [18].

MRI and CTA couldn't be done on the patient to know exact site of the lesion.

This study differs from previous ones as electrophysiological tests like nerve conduction have also been conducted on a patient of medullary syndrome. These tests showed decrease in conduction velocity of right facial nerve, left peroneal & tibial nerve with decreased amplitude. The conduction velocity of bilateral median nerves was normal. Sensory conduction velocity of bilateral median nerves was normal.

The blink reflex was conducted by stimulation of the supraorbital nerve [19].

The latency of R2 ipsilateral & R2 contralateral orbital nerves were inceeased indicating a lesion covering the entire trigeminal spinal tract and nucleus (TSTN). Similar results were reported by Fitzek et al. [20].

In this patient, absence of Horners syndrome, palatal pharyngeal or laryngeal palsy can be explained by the dorso lateral involvement of the medulla. Similar results were also reported by Rohan et al. [21].

Dorso lateral lesions tend to be more superficial and hence don't involve nucleus ambiguous which is situated more deeply [22].

Brainstem auditory evoked potentials showed increased latency of wave II, III, IV and V. Similar results were reported by Elwany [23].

BERA may be of prognostic value in the early evaluation of patients with brainstem ischemic stroke [24].

Visual evoked potentials showed increased P100 latency which indicates subclinical involvement of visual pathway in medullary syndrome in this patient.

5. CONCLUSION

The present case study revealed that this young female patient had features of both lateral & medial medullary syndrome suggesting a lesion of both upper & middle medulla. This is a less reported condition in literature and more work should be done on this condition with more emphasis on early diagnosis so that the appropriate treatment protocol may be started to alleviate the symptoms of the patients. In this regard, electrophysiology can play a vital role.

CONSENT

As per the international guidelines an informed and written participant consent explaining all the details of the study has been collected and preserved by the author.

ETHICAL APPROVAL

As per international standard written ethical permission has been collected and preserved by the author(s).

COMPETING INTERESTS

Author has declared that no competing interests exist.

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