Case Report

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Lateral Medullary Syndrome Presenting as Persistent Singultus and Upper Motor Neuron Facial Palsy in a young Adult: A Rare Case

Internal Medicine Section

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ABSTRACT

Lateral Medullary Syndrome (LMS) is a collection of different neurologic symptoms after cerebral haemorrhage/infarction. The areas most frequently afflicted are the cerebellum and the posterolateral portion of the brain stem's medulla oblongata; the Posterior Inferior Cerebellar Artery (PICA) supplies blood. Dizziness, vertigo, nystagmus, nausea, ataxia, dysphagia, vomiting, hiccups, hoarseness, sensation impaired on half of the face, impaired thermal sensation and pain on the contralateral side of the limbs, trunk, ipsilateral face, along with Horner's syndrome are amongst the typical symptoms of lateral medullary infarction. Hiccups are among the signs that are simple to ignore, although they can also result in oesophagitis, and respiratory depression, as Wallenberg's syndrome is the most frequently reported. The authors discuss the case of a 30-year-old male who complained of recurrent, uncontrollable hiccups as an LMS-presenting symptom and was brought to our hospital. He arrived at our emergency room complaining of persistent, intractable hiccups and cerebellar symptoms on the left-side. In addition to being hypertensive, he had a long history of tobacco (chewing) and alcohol addictions dating back six years. He had 7, 9 and 10th cranial nerve involvement. The authors are reporting this case because the patient had Upper Motor Neuron (UMN)-type facial palsy, a rare presentation in LMS.

Keywords: Hiccups, Posterior inferior cerebellar artery, Stroke

CASE REPORT

A 30-year-old male patient came in complaining of vomiting 5-6 times, uncontrollable hiccups for 10 days slurring of speech, and left-sided facial weakness for one day. The patient has just received antacids (inj. Pantoprazole 40 mg), but there was no significant improvement in the symptoms. The patient has had a history of hypertension for the last 10 years for which he was taking regular medication (Tab Telmisartan 40 mg once a day). The patient has been an alcoholic for six years and used to consume around 40-60 mL twice a week and tobacco chewer for eight years. On general examination, his pulse was 80 per minute and his blood pressure was 170/100 mmHg in his right arm, supine position. His respiratory, abdominal and cardiovascular system examination was within normal limits.

On Central Nervous System (CNS) examination his higher mental function was unimpaired. He had a deviation of the angle of mouth to the right, suggestive of Upper Motor Neuron (UMN) 7th cranial nerve palsy [Table/Fig-1]. He was also having voice hoarseness as well as nasal regurgitation due to a lack of gag reflex, suggestive of 9th and 10th cranial nerve involvement. On motor system examination his tone was normal with 5/5 power and all deep tendon reflexes were intact. The left finger-nose test, left heel-to-knee test, and left dysdiadochokinesia test were impaired and torsional nystagmus was present, suggestive of the involvement of the cerebellum. Complete Blood Count (CBC), Liver Function Test (LFT), Kidney Function Test (KFT), serum electrolytes, and sugars were all within normal limits [Table/Fig-2].

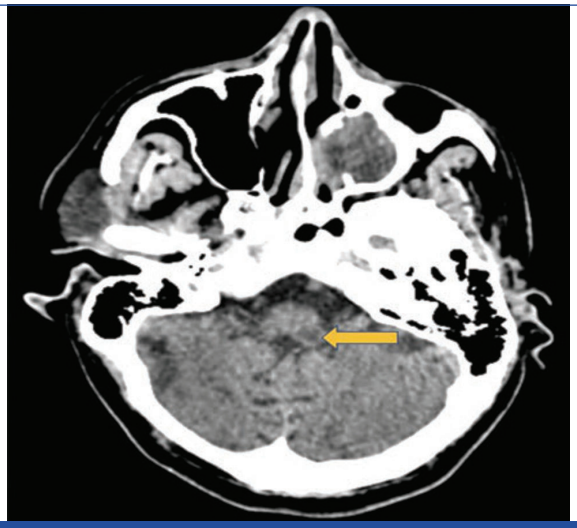
Computed Tomography (CT) of brain showed an acute infarct in the medulla on the left-side [Table/Fig-3] (Yellow arrow). Magnetic Resonance Imaging (MRI) of brain showed [Table/Fig-4] (Red arrow) a Diffusion-weighted image showing a high signal in the lateral part of the left medulla oblongata. The patient was started on dual antiplatelet (Tab Ecosprin 75 mg od, Tab Clopidogrel 75 mg od), statins, Tab Baclofen 10 mg once a day and increased to 20 mg three times a day and Injectable prokinetic agents (Metoclopramide 10 mg three times a day), and later was given four times a day; there

[Table/Fig-1]: Deviation of the angle of mouth to the right, suggestive of Upper Motor Neuron (UMN) 7th cranial nerve palsy.

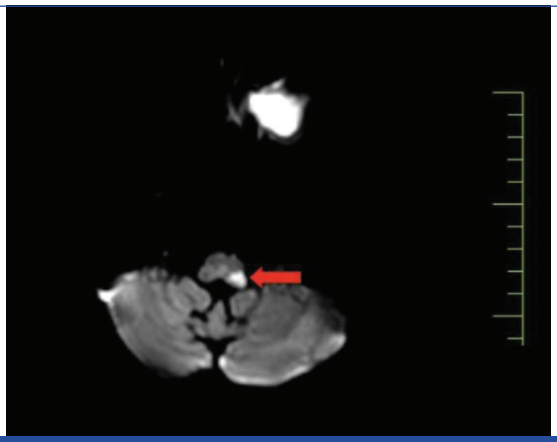
Laboratory parameters	Observed value	Normal range
Haemoglobin	16 g%	13-17 g%
Mean corpuscular volume	80.2 fL	83-101 fL
Total leucocyte count	4440 cells/cu mm	4000-10000 cells/cu mm
Platelets	3.57 lakhs/cu mm	1.5-4.1 lakhs/cu mm
Blood urea	54 mg/dL	19-43 mg/dL
Serum creatinine	0.9 mg/dL	0.66-1.25 mg/dL
Serum sodium	132 mmol/L	137-145 mmol/L
Serum potassium	4.1 mmol/L	3.5-5.1 mmol/L
Serum calcium	9.2 mg/dL	8.4-10.2 mg/dL
Serum magnesium	2.0 mg/dL	1.6-2.3 mg/dL
Serum phosphorous	2.9 mg/dL	2.5-4.5 mg/dL
Serum uric acid	4.2 mg/dL	3.5-8.5 mg/dL
Serum alkaline phosphatase	130 U/L	38-126 U/L
Serum alanine amino transferase	22 U/L	<50 U/L

Serum aspartate amino transferase	48 U/L	17-59 U/L
Serum albumin	4.4 g/dL	3.5-5 g/dL
Serum total bilirubin	0.9 mg/dL	0.2-1.3 mg/dL
Serum conjugated bilirubin	0.3 mg/dL	0.0-0.3 mg/dL
Serum unconjugated bilirubin	0.6 mg/dL	0.0-1.1 mg/dL
Serum homocysteine	78 micro mol/L	6.6-14.8 micro mol/L
Random blood sugar	134 g/dL	90-140 g/dL
Serum vitamin B12	800 pg/mL	239-931 pg/mL
Serum vitamin D	8.27 ng/mL	<20 ng/mL
Serum thyroid stimulating hormone	0.522 mIU/mL	0.465-4.68 mIU/mL

[Table/Fig-2]: All laboratory investigations.



[Table/Fig-3]: Computed Tomography (CT) of brain showing hypodense area in lateral part of left medulla oblongata (yellow arrow).



[Table/Fig-4]: Magnetic Resonance Imaging (MRI) of brain on the diffusion-weighted image showing high DWI signal in the lateral part of the left medulla oblongata (Red arrow).

was no improvement in the frequency and intensity of hiccups. Suctioning was done frequently. Due to an absent Gag reflex, he was intubated to protect his airway from the pent-up secretions. As we could not wean off the mechanical ventilation, a tracheostomy was done. He was discharged after one month and followed-up every month. The patient improved significantly, and the hiccups settled after 2.5 months. The tracheostomy tube was decannulated after three months. On regular follow-up, the patient is doing well.

DISCUSSION

Repeated involuntary, spasmodic, and momentary contractions of the diaphragm cause hiccups characterised by rapid closure of the glottis and a distinctive "hic" sound [1]. Most people will sometimes suffer hiccups, also known as singultus, which are often short-lived and self-limiting [1,2]. Rarely do hiccups persist for more than 48 hours, necessitating immediate medical attention and thorough medical examinations to identify any underlying pathology [3].

Although the precise location of the hiccups' neuroanatomical center is unknown, it is thought to be a region of the brain stem that interacts with the brain stem's respiratory centers, hypothalamus, medullary reticular formation, and phrenic nerve nuclei. The dorsal sympathetic fibres and sensory branches of the vagus and phrenic nerves make up the afferent pathway. In contrast, the motor fibres of the phrenic nerve make up the efferent pathway. Both internal and external factors can produce hiccups [3]. Lesions in the pathway between the CNS and the phrenic nerve, most commonly in the brain stem, illnesses such as a dolichoectatic basilar artery, ischaemic stroke, encephalitis, tumours and multiple sclerosis, cause major hiccups. Phrenic nerve disorders can result in peripheral hiccups like gastric distension [4].

With various clinical manifestations of Cerebrovascular Event (CVE), Lateral Medullary Syndrome (LMS) remains an intriguing clinical entity [5]. The posterolateral section of the medulla oblongata, which is the component getting arterial blood flow from the Posterior Inferior Cerebellar Artery (PICA) is the region of the brain stem involved in LMS [6]. Dizziness, vertigo, nystagmus, nausea, ataxia, dysphagia, vomiting, hiccups, hoarseness, sensation impaired on half of the face, impaired thermal sensation and pain on the contralateral side of the limbs, trunk, ipsilateral face, along with Horner's syndrome are amongst the typical symptoms of lateral medullary infarction. Hiccups are among the signs that are simple to ignore, although they can also result in oesophagitis, and respiratory depression, as Wallenberg's syndrome is the most frequently reported [7].

Though a few theories have been put up, how hiccups in people with LMS occur is still being determined. It involves the dorsolateral medulla region, which contains the nucleus ambiguous and the motor neurons of the vagus that project to the premotor neurons and larynx that govern the diaphragm. The motor neurons of the larynx, which control the glottis, and the premotor neurons, which control the inspiratory muscles, are affected, causing hiccups. Constant hiccups frequently point to a severe underlying illness requiring thorough examination. Most of the more than 100 causes of hiccups are found in the digestive system. Recently, there has been a notable focus on brain stem lesions, with particular emphasis on identifying significant contributing factors. Numerous different diseases of the medullary area can bring on hiccups. Clinical studies [3,6,8,9] have shown a correlation between the 9th and 10th cranial nerve symptoms and hiccups, which appeared to be associated with a complete lesion in the middle of the lateral medullary area [9]. A small region of the medullary reticular formation, the Hiccup-evoking Site (HES), can be electrically stimulated to cause a response resembling a hiccup, and hiccups are quickly stopped by microinjecting the Gamma Aminobutyric Acid (GABA) analogue baclofen into the HES [8].

Patients with LMS frequently had hiccups when the lesions were in the central medulla's dorsolateral region. Hiccups and signs of cerebellar, vestibular, and 5th, 9th, and 10th cranial nerve involvement were closely correlated [9]. It has been documented that gabapentin treats chronic hiccups in LMS patients. The effects of gabapentin may include enhancing GABA-mediated inhibition or specifically reducing calcium influx by inhibiting voltage-operated calcium channels [10]. This may stop the diaphragm's intermittent myoclonus, which results from the isolated nucleus's recurrent activity.

One case report revealed facial palsy in LMS with the possible hypothesis that there is abnormality in the Dejerine's pyramidal tract. The hypothetical supranuclear corticobulbar fibre loop in the medulla descends ventromedially, decussates at the superior medulla, and then ascends dorsolaterally to the facial nerve nucleus. Chakraborty U et al., opines that LMS exhibited unusual features such as hemiparesis, UMN facial palsy, and the absence of typical bulbar symptoms and classical Horner's syndrome,

which is similar to the present study. These variations stem from differences in the affected neuroanatomical structures [11]. When considering the neurological correlation, these manifestations align with a potential diagnosis resembling Babinski-Nageotte syndrome, specifically characterised by UMN-type facial palsy. Ibrahim M et al., [12] wrote in their case report that a patient presented with horizontal nystagmus, indicative of true bulbar palsy affecting the cranial nerves responsible for the bulbar function, which was not observed in the present case. The patient exhibited dysphagia, primarily for fluids, along with nasal phonation of the voice. An MRI of the brain was requested and revealed acute lacunar infarctions in the lateral and posterior aspects of the right side of the medulla, as well as the posterior inferior aspect of the right cerebellar hemisphere within the PICA territory. To manage the dysphagia, a nasogastric tube was inserted. The patient's treatment plan included lioresal 25 mg three times a day and phenergan (an antihistaminic) twice a day to control hiccups. Additionally, the patient was prescribed clopidogrel 75 mg/day and Angiotensin-converting Enzyme Inhibitors (ACEI) to regulate blood pressure. Upon discharge, the patient retained the nasogastric tube and was scheduled for follow-up at the neurology clinic [12].

CONCLUSION(S)

The LMS presenting with involvement of the facial nerve is very rare. Hence, the authors are presenting this case suggesting not overlooking the LMS in the presence of these rare involvements, instead recognising the unusual presentation at the earliest and managing appropriately for a better prognosis. Early recognition and diagnosis are imperative in optimising patient outcomes. Prompt intervention, which may include targeted therapies, rehabilitation, and supportive care, can contribute to better recovery and mitigate potential complications associated with LMS. The present case report emphasises the value of taking a cautious approach to seemingly unimportant hiccups, especially when they are present and exhibit symptoms resistant to initial treatments.

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