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AN ATYPICAL PRESENTATION OF HEMIMEDULLARY SYNDROME WITH CONTRALATERAL UPPER MOTOR NEURON TYPE FACIAL PALSY – A CASE REPORT

Dr S Arun Saravanan	Institute Of Neurology, Madras Medical College, Chennai
Prof Dr M Jawahar	Institute Of Neurology, Madras Medical College, Chennai
Dr V Nanda Kumar	Institute Of Neurology, Madras Medical College, Chennai
Prof Dr S Balasubramanian	Institute Of Neurology, Madras Medical College, Chennai

ABSTRACT The identity of hemimedullary stroke which has both the features of medial and lateral medullary syndrome is very rare because both medial and lateral medullary infarctions rarely occur simultaneously as the vascular supplies of the medial and lateral medulla usually differ. The association of medullary syndrome with UMN-type facial palsy is very rare and is attributed to the involvement of hypothetical supranuclear aberrant corticobulbar fibres of the facial nerve. Here, we have discussed a 72 year old male with clinical features of hemimedullary syndrome with contralateral UMN-type facial palsy and contralateral hemisensory loss of pain and temperature sensation of the face, trunk, and limbs. We reported this case to address the various atypical presentation of the medullary syndrome and their characteristics to be known and understood.

KEYWORDS :

INTRODUCTION:

The medulla oblongata has a very compact arrangement of structures. Even relatively small lesions usually give a constellation of symptoms that constitute syndromes. There are three unilateral syndromes of the medulla: The more common lateral medullary Wallenberg's syndrome,[1] the rarer medial medullary Dejerine's syndrome[2] and the very rare hemimedullary syndrome, in which medial and lateral medullary lesions occur simultaneously.

The lateral medullary syndrome is caused most commonly due to atherothrombotic vertebral artery occlusion, followed by posterior inferior cerebellar artery (PICA) and medullary arteries and the medial medullary syndrome is caused by infarction of paramedian branches of the anterior spinal artery, the vertebral artery or the basilar artery.[3]

The lateral medullary syndrome presents with loss of pain, temperature sensation on the ipsilateral half of the face, and on the contralateral trunk and extremities, ipsilateral Horner's syndrome, vertigo, nausea, vomiting, diplopia, hiccups, ipsilateral cerebellar symptoms and signs. The medial medullary syndrome presents with ipsilateral hypoglossal palsy, contralateral hemiplegia and contralateral impairment of vibration and position sense.

The identity of hemimedullary stroke which has both the features of medial and lateral medullary syndrome is extremely very rare because both medial and lateral medullary infarctions rarely occur simultaneously as the vascular supplies of the medial and lateral medulla usually differ as described above.[4]

Our Case:

A 72 year old man, previously not evaluated for any co-morbidities presents with acute onset giddiness, recurrent episodes of vomiting, difficulty in swallowing, inability to use left upper and lower limbs, slurring of speech and tremulousness of right upper and lower limbs. The patient also had had deviation of the angle of mouth to the right side and drooping of the right eyelid.

On examination, the patient was conscious, obeys verbal commands, had elevated blood pressure, right-sided partial ptosis, reduced pain, touch sensation over left side face, Left UMN type facial palsy, uvula deviated to the left side, reduced palatal movements and gag reflex on the right side, right-sided hypoglossal palsy, left-sided hemiplegia, left-sided extensor plantar response, reduced pain, touch and temperature sensation over the left side of the face, trunk and limbs, absent vibration over the left side of the body, absent joint position sense over left-sided limbs, truncal ataxia, gaze-evoked nystagmus and right-sided cerebellar signs. The patient also had the guttural type of dysarthria. The cranial nerve involvement of the patient is shown in the images 1,2 and 3.

Image 1 and Image 2: Patient having left sided UMN type facial palsy

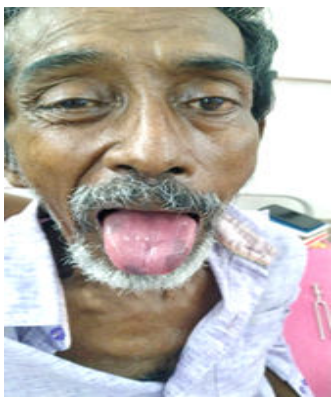

Image3: Patient having right-sided partial ptosis with tongue weakness on the right side.

MRI Brain revealed diffusion restriction in the right side medulla suggestive of acute infarct in the DWI sequence and was supported by the ADC sequence as shown in the images 4 and 5. ECG and Echocardiography findings were within normal limits.

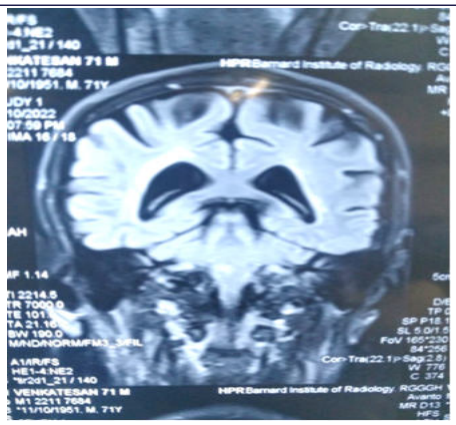

Image 4: T2 FLAIR image of MRI Brain showing infarct of right side medulla

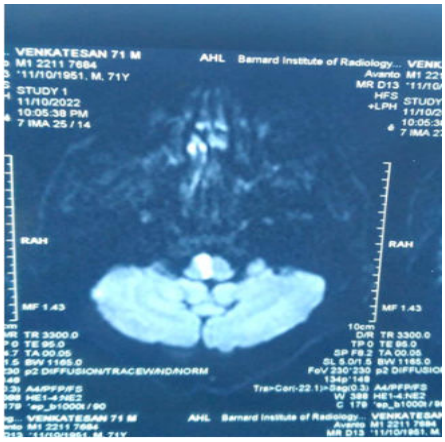

Image 5: MRI Brain showing diffusion restriction suggestive of acute infarction in the right side medulla

He was treated with supportive therapy, antiplatelet medications, statins, antihypertensive medications, physiotherapy and improved gradually.

DISCUSSION:

The clinical manifestations of the lateral medullary syndrome include ipsilateral paralysis of the soft palate, pharynx, and larynx with dysphagia, dysarthria, dysphonia, ipsilateral anesthesia of the face for pain and temperature sensation, ipsilateral Horner's syndrome, and ipsilateral cerebellar hemiataxia. Contralateral loss of pain and temperature sensation on the limbs and trunk are present. Structures affected include the nucleus ambiguus and the fibres of the vagus and glossopharyngeal nerves, the descending tract and nucleus tractus spinalis of the trigeminal nerve, the descending sympathetic pathways, posterior spinocerebellar tract and the lateral spinothalamic tract.[1] The clinical manifestations of the medial medullary syndrome include Ipsilateral paralysis and atrophy of one half of the tongue, contralateral impaired tactile and proprioception sense over the body sparing the face and contralateral hemiparesis of the body.[2] Structures affected are hypoglossal nerve, medial lemniscus and pyramidal tract.[5] Here we present our case with features of both medial and lateral medullary syndrome with ipsilateral hypoglossal palsy and contralateral facial palsy.

Atypical features in our patient concerning hemimedullary syndrome are:

I) Contralateral UMN facial palsy
II) Contralateral loss of pain and touch sensation over the face

Hemimedullary syndrome:

Since the publication of Nakane et al in 1991, patients with a combined medial and lateral medullary syndrome are considered to have Babinski-Nageotte's syndrome and it was assumed that Babinski-Nageotte's syndrome is the classical brainstem syndrome corresponding to hemimedullary lesion.[6] However, prominent hypoglossus palsy has not been included in Babinski-Nageotte's syndrome according to the original description.[7] Another variant of a lateral medullary syndrome associated with hemiparesis is Opalski syndrome where hemiparesis is ipsilateral and hypoglossal palsy was not included. The peripheral hypoglossal paralysis is an obligate symptom of the Dejerine's medial medullary syndrome[2] as well as of hemimedullary infarction, but it is not a symptom of Babinski-Nageotte's syndrome or Opalski syndrome.

Thus, Babinski- Nageotte's syndrome occurs by the caudal extension of infarct to involve the pyramidal tracts before decussation at the medulla[8] and Opalski syndrome occurs by the caudal extension of infarct to involve the pyramidal tracts after decussation at the medulla.[9] The Cestan-Chenais's syndrome includes all the symptoms of Babinski-Nageotte's syndrome except ipsilateral cerebellar hemiataxia.[10]

The clinical features of the hemimedullary syndrome, described in 1894 by Reinhold are different from other classical brainstem syndromes which included the clinical features of both lateral and medial medullary syndrome with ipsilateral hypoglossal palsy.[11] The hemimedullary syndrome is characterized by a simultaneous infarction of median, paramedian lateral, and dorsal areas of the medulla oblongata. Our patient had manifested the clinical features of the complete lateral medullary syndrome as well as of the complete medial medullary syndrome with ipsilateral tongue weakness consistent with features of the Reinhold syndrome.

Contralateral UMN VII cranial nerve palsy:

UMN-type facial palsy in medullary syndrome may be attributed to the interruption of hypothetical looping supranuclear facial fibres of the corticobulbar tract which descend in the ventro medial medulla, decussate at the level of the upper medulla and then ascend up in the dorso lateral medulla to reach the facial nerve nucleus. [12]

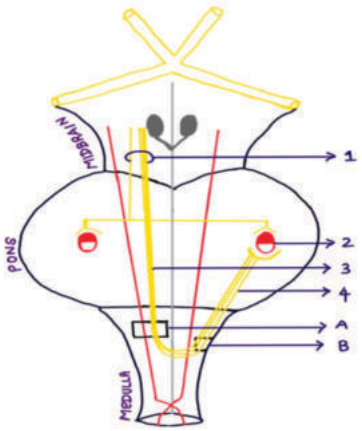

Image 6: Schematic diagram of brainstem showing the aberrant pathway of corticobulbar fibres of facial nerves and the site of the lesion.

1 - Supra nuclear facial corticobulbar fibres.
2 - Facial nucleus.
3 – Descending aberrant facial corticobulbar fibres in ventromedial medulla
4 - Ascending aberrant facial corticobulbar fibres in dorsolateral medulla
A - Interruption of the descending facial corticobulbar fibres in medial medulla at a pre-decussation level can result in contralateral UMN facial palsy
B - Interruption of the ascending facial corticobulbar fibres in lateral medulla after decussation level can result in ipsilateral UMN facial palsy

In our patient, the lesion in the medial medulla at the level of A as in picture 1 can explain the contralateral UMN type of facial palsy.

Contralateral loss of pain and touch sensation over the face (Pseudothalamic pattern of sensory loss in lateral medullary syndrome):

In the lateral medullary syndrome, classical sensory involvement is ipsilateral loss of pain and temperature over the face and contralateral loss of pain and temperature over the trunk and body. However, variant sensory involvement can occur as described by the Stopford type of sensory involvement in lateral medullary syndrome.[13] It is due to the

differential involvement of the following three structures:

1. The descending tract of the trigeminal nucleus which carries the pain and temperature sensation of the ipsilateral face
2. The ascending quintothalamic tract which carries the pain and temperature sensation of contralateral face
3. The laminated spinothalamic tract carries the pain and temperature sensation of contralateral trunk and body.

Vaudens et al have first described the pseudothalamic pattern of sensory loss in lateral medullary syndrome as Stopford Type 4 Ventral-tegmental lateral medullary infarction.[14]

In this, the descending tract of the trigeminal nucleus is spared and the ascending quintothalamic tract is involved along with the spinothalamic tract as shown in the image 7. It explains the loss of pain and temperature sensation of the contralateral face, trunk and body and sparing of pain and temperature sensation of the ipsilateral face as seen in our patient.

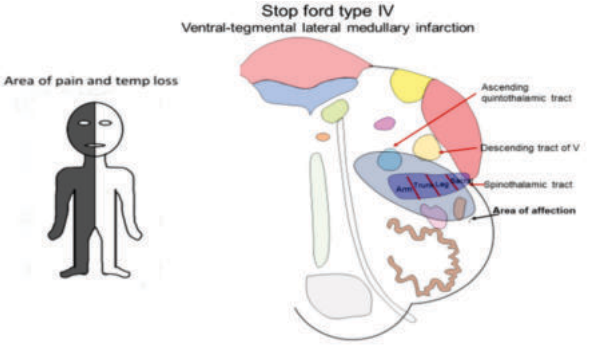

Image 7: structures involved and clinical pattern of sensory loss in Stop ford type IV lateral medullary infarction.[15]

CONCLUSION:

Our patient had manifested the clinical features of both lateral medullary syndrome as well as of the medial medullary syndrome with ipsilateral tongue weakness consistent with features of Reinhold syndrome. Our case also emphasizes that the medullary syndrome can present with atypical sensory involvement and UMN facial palsy. The stopford type of sensory involvement in lateral medullary syndrome and the involvement of hypothetical supranuclear aberrant corticobulbar fibres of the facial nerve can explain the atypical sensory findings and UMN type facial palsy respectively in our patient. Hence, the clinicians should be aware of the atypical presentation of brain stem strokes and understanding their neuroanatomical correlations will help not to miss the early clinical diagnosis and optimal management.

Declaration of patient consent:

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity.

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Nil.

Conflicts of interest:

There are no conflicts of interest.

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