Brain Stem: Medial and Lateral Medullary Syndrome

Brain stem syndromes are characterized by infliction of precise structural entities with specific consequent clinical manifestations. Relevant appraisal of anatomy and blood supply of medulla is essential to understand the peculiar features of medullary syndromes. In this article, we emphasize on clinical characteristics of medial medullary syndrome and lateral medullary syndrome in perspective of the topographical organization of medulla. We conclude with therapy and prognosis.

Organization and Blood Supply of Medulla

*Medulla oblongata* is the terminal segment of the brain stem which continues as spinal cord at the foramen magnum. At the level of the pyramids and inferior olivary nucleus, the following structures can be discerned on the cross-section.

Constituting the medial medulla
Pyramids: decussating corticospinal tracts constitute the pyramids. It is located in between median fissure and ventrolateral sulcus.

Hypoglossal nuclei and nerve fibers- arises from the meeting pont of pons and medulla.

Medial longitudinal fasciculus: connects various cranial nerve nuclei such as III, VI and VIII with the spinal cord bringing about co-ordination in eye, body and neck movements.

Medial lemniscus: mediating position, vibration and fine touch sensations; the posterior column fibers from gracile and cuneatus nuclei cross as internal arcuate fibers and further continue in the contralateral medial lemniscus to the thalamus.

Collectively constituting the lateral medullary segment

Inferior cerebellar peduncle: mainly concerned with proprioception, integration and co-ordination of motor tasks, following tracts traverse the inferior cerebellar peduncle:

<table>
<thead>
<tr>
<th>Tract</th>
<th>Function</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dorsal spinocerebellar tract</td>
<td>Conveys unconscious proprioception from the spinal cord</td>
</tr>
<tr>
<td>Climbing fibers from inferior olivary nucleus to cerebellum</td>
<td>Major input to the cerebellum for motor co-ordination</td>
</tr>
<tr>
<td>Vestibulo-cerebellar tract</td>
<td>Vestibular information is conveyed to the flocculo-nodular lobe (vestibulocerebellum)</td>
</tr>
<tr>
<td>Cuneo-cerebellar tract</td>
<td>Originating in the ipsilateral accessory cuneate nucleus, it mediates unconscious proprioceptive information from neck and upper limb.</td>
</tr>
</tbody>
</table>

Vestibular nuclei: its location is on the dorsolateral regions of medulla and pons. It is comprised of 4 sub-nuclei, it gives rise to the vestibular component of
the vestibule-cochlear nerve (eighth cranial nerve). The latter is important for maintaining balance, orientation and proprioception of the head and body in the space. The sub-nuclei are:

- Inferior vestibular nucleus (Spinal or descending nucleus)
- Medial vestibular nucleus (Schwalbe nucleus)
- Lateral vestibular nucleus (Deiter nucleus)
- Superior vestibular nucleus (Bechterew nucleus)

**Spinal nucleus of trigeminal nerve:** gives rise to the trigemino-thalamic tract, which mediates pain and temperature sensations from the face.

**Nucleus ambiguus:** gives rise to the branchio-motor supply of IX, X and XI cranial nerves.

**Solitary nucleus:** the rostral part mediates taste sensation while the dorsal segment is responsible for visceral sensations and the carotid reflex.

**Descending sympathetic fibers:** proceed towards the ciliospinal center of Budge in the cervical spinal cord.

**Spinothalamic tract:** mediates pain and temperature sensations from the body to the thalamus.

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**Blood supply**

Medulla is supplied by the vertebro-basilar trunk and its branches. The two vertebral arteries join to form the basilar artery. Three types of end-arteries supply the medulla:

- Long circumferential vessels
- Short circumferential vessels
- Paramedian perforators.

The major long circumferential vessels and their vascular territories are as follows:
The vascular territories being exclusive with sharp demarcation, occlusion of specific branches lead to specific set of symptoms and manifestations depending on the territory involved.

We are now set to discuss the individual syndromes.

**Medial Medullary Syndrome**

First described in 1908 by Spiller, medial medullary syndrome is characterized by damage and dysfunction of the medial medullary structures.

Dejerine introduced the **triad of contralateral hemiplegia sparing the face, contralateral loss of deep sensation and ipsilateral hypoglossal paralysis** to this disease, now also known as Dejerine syndrome. It is also variably known as hypoglossal alternating hemiplegia, inferior alternating syndrome and lower alternating hemiplegia.

**Lateral Medullary Syndrome**

Damage of the lateral medulla leads to lateral medullary syndrome. Gaspard Viesseux first described it in 1808. It is also known as ‘Wallenberg syndrome.’ Of interest are the following variants:

**Opalski syndrome:** lateral medullary syndrome with ipsilateral hemiplegia

**Babinski-Nageotte syndrome:** lateral medullary syndrome with contralateral...
Lateral medullary syndrome may be complete or partial depending on the vessel and the subsequent vascular territory involved.

All brain stem syndromes are ‘crossed syndromes’ with involvement of ipsilateral cranial nerve and contralateral motor weakness.

Epidemiology

There are very few clinically and pathologically proven patients of medial medullary syndrome. Lateral medullary syndrome is relatively more common. According to a study, it is highest incidence is seen in middle aged males at 50-60 years. Hypertension, diabetes, smoking, and atherosclerosis are its major risk factors.

Pathogenesis of Medullary Syndrome

Medial medullary syndrome is usually caused by infarction following vascular insult to the medial medulla. Occlusion of the ASA or damage to the paramedian perforators culminates in medial medullary syndrome.

Infarction of the lateral medulla following insult to the PICA or the vertebra-basilar trunk or brain stem perforators gives rise to lateral medullary syndrome.

The etiogenesis is similar in both the syndromes.

Infarction and vascular insult:

- **Thrombosis**: secondary to atherosclerosis and hypertension (most common); less likely due to hypercoagulable states
- **Embolic occlusion**: seen secondary in cocaine abuse, medullary neoplasms, radionecrosis, hematoma, neck manipulation, and trauma and bullet injury to the vertebral artery
- Spontaneous or post-traumatic or iatrogenic vertebral artery dissection
- Post-traumatic or iatrogenic vertebra-basilar trunk occlusion/injury
- **Vasculitis**
- Post-infective vasculitis or occlusion
- Post-endovascular intervention
- Post-surgical intervention for vascular anomalies like AVM
- Post-surgical intervention of the spine
- Other rare causes are:
  - Structural lesions such as brain tumors
  - Hemorrhage
  - Vascular anomalies: leading to “steal phenomenon” causing secondary hypo-perfusion of the medulla
  - Idiopathic.

Classification of Medullary Syndrome

Based on the location of the infarct on imaging, there are three types of medial medullary syndrome:

- **Ventral**: contains the pyramid (most common)
- **Middle**: includes the medial lemniscus
Dorsal: includes the medial longitudinal fasciculus.

Symptoms and Diagnosis of Medullary Syndrome

Frequently being acute ischemic events, **brain stem strokes** are characterized by vomiting, vertigo, diplopia, headache, ataxia, hiccups and dysarthria. Facial paresis, though rare, can be seen due to involvement of few aberrant descending fibers. It can causes ailments of many systems like vestibule-cerebellar, bulbar, sensory, respiratory or autonomic systems.

More often due to a systemic cause such as **hypertension** and **atherosclerosis**, medial medullary syndrome is often **bilateral** with worse prognosis.

The tracts involved with resultant specific symptoms in medial medullary syndrome can be summarized in the following manner:

<table>
<thead>
<tr>
<th>Tract involved in medial medullary syndrome</th>
<th>Manifestation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoglossal nerve fibers</td>
<td>Ipsilateral tongue weakness, deviation of tongue to ipsilateral side</td>
</tr>
<tr>
<td>Corticospinal nerve fibers in the “pyramid”</td>
<td>Contralateral motor weakness</td>
</tr>
<tr>
<td>Medial lemniscus</td>
<td>Contralateral posterior column sensations affection-proprioception, vibration and fine touch with relative sparing of pain and temperature</td>
</tr>
<tr>
<td>Medial longitudinal fasciculus (MLF)</td>
<td>Disruption of MLF leads to “internuclear ophthalmoplegia” characterized by failure to adduct the contralateral eye</td>
</tr>
</tbody>
</table>

Clinical recognition of the lateral medullary syndrome depends on identification of the **triad of ipsilateral hyperalgesia of the face, Horner’s syndrome and ipsilateral ataxia.**

<table>
<thead>
<tr>
<th>Tract involved in lateral medullary syndrome</th>
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</tr>
</thead>
<tbody>
<tr>
<td>Inferior cerebellar peduncle</td>
<td>Ipsilateral ataxia, ipsilateral appendiceal cerebellar signs as dysmetria, intentional tremors</td>
</tr>
<tr>
<td>Vestibular nuclei</td>
<td>Vomiting, vertigo, nystagmus, diplopia.</td>
</tr>
<tr>
<td>Trigemino-thalamic tract from spinal nucleus of trigeminal nerve</td>
<td>Loss of pain and temperature sensation from ipsilateral half of the face</td>
</tr>
<tr>
<td>Nucleus ambiguus and solitary nucleus</td>
<td>Lower cranial nerve dysfunction-hoarse voice, nasal twang and dysphonia, dysphagia, decreased gustation weak gag, regurgitation, aspiration and increased risk of aspiration pneumonia</td>
</tr>
<tr>
<td>Lateral spinothalamic tract</td>
<td>Loss of pain and temperature sensation from contralateral half of the body</td>
</tr>
<tr>
<td>Descending sympathetic fibers</td>
<td>Ipsilateral Horner’s syndrome (miosis, ptosis, anhydrosis and loss of ciliospinal reflex)</td>
</tr>
<tr>
<td>Central tegmental tract</td>
<td>Palatal myoclonus</td>
</tr>
</tbody>
</table>

**Symptoms of lateral medullary syndrome**

- Vertigo
- Nausea and vomiting,
- Blurred vision
- Disbalanced sitting posture
- Horizontal or rotational nystagmus
- Loss of sensation of pain and temperature of half of the face
- Loss of sensation of pain and temperature of contralateral half of the trunk
- Hoarseness,
- Diplopia
- Dysphagia
- Hiccups
- Dysphonia
- Horner’s syndrome
- Poor gag reflex
- Ataxia of both limb and gait
- Bradycardia

**Diagnosis**

After the assessment of present symptoms, a neurological examination is performed to differentiate between true stroke and mimicking stroke by this syndrome. **HINTS (head-impulsive-nystagmus-test of skew)** is a three-step oculomotor examination that can determine the presence of infarction in lateral medulla.

**Imaging studies** are often the preliminary investigations performed. **CT brain (plain)** is the initial investigation, mainly to rule out hemorrhage. **MRI diffusion** detects infarcts within a few minutes. CT scan and MRI scan determine the exact location of the infarction. The etiology can be detected by performing studies like **CT angiography, MRI angiography** and **Arterial neck Doppler**. For large vessel disease, **digital subtraction angiography** is the gold standard.

**Therapy and Prognosis of Medullary Syndrome**

Management is usually conservative. **Blood pressure regulation** with cerebral perfusion maintenance is immediately sought. In case of ischemic stroke, blood thinners like heparin or warfarin are prescribed to reduce the blockage in the arteries that supply lateral side of medulla. The long-term outlook depends on active rehabilitation and symptomatic treatment.

Transcranial magnetic stimulation is given in repetition to rehabilitate patients having dysphagia due to Wallenberg syndrome.

Patients with lower cranial nerve weakness in lateral medullary syndrome are prone to **aspiration pneumonia**. They might benefit from **Ryle’s tube feeds** or diverted feeding techniques like **gastrostomy**. Patients who require **tracheostomy** usually have guarded prognosis.

Role of surgery, though minimal, can be summarized as:

- For structural lesions
- For vascular lesions like AVM
- For vertebro-basilar stenosis: endovascular intervention can also be performed.

Medullary syndromes are **lethal in many patients**. Those who survive are usually dependent and afflicted by **central poststroke pain**.
Summary

Brain stem syndromes are ‘crossed syndromes,’ usually secondary to ischemic strokes and infarcts.

Medial medullary syndrome is frequently caused by occlusion of ASA or vertebral artery. It is identified by ipsilateral tongue weakness, ipsilateral tongue deviation and contralateral motor weakness.

Lateral medullary syndrome is commonly due to PICA or vertebral artery occlusion. It manifests as triad of ipsilateral ataxia, ipsilateral Horner’s syndrome and ipsilateral hyperalgesia of the face. Loss of pain and temperature sensation in contralateral half of body is seen.

These medullary syndromes are diagnosed based on clinico-radiological correlation. Management is usually conservative with high mortality and morbidity.

Review Questions

The correct answers can be found below the references.

1. A 55-year-old male developed sudden onset vertigo and vomiting. On examination, he was found to have deviation of tongue to the left side with right-sided limb weakness. What is he suffering from?
   A. Medial medullary syndrome: right side
   B. Medial medullary syndrome: left side
   C. Lateral medullary syndrome: left side
   D. Lateral medullary syndrome: right side

2. The most common cause of lateral medullary syndrome is:
   A. Middle cerebral artery occlusion
   B. Carotid artery atherosclerosis
   C. Anterior spinal artery occlusion
   D. Posterior inferior cerebellar artery occlusion

3. Lateral medullary syndrome is characterized by the following triad:
   A. Ipsilateral hyperalgesia of the body, ipsilateral Horner’s syndrome and ipsilateral ataxia
   B. Ipsilateral ataxia, contralateral tongue weakness and ipsilateral Horner’s syndrome
   C. Ipsilateral Horner’s syndrome, ipsilateral ataxia and ipsilateral hyperalgesia of the face
   D. Ipsilateral ataxia, contralateral Horner’s syndrome and ipsilateral hyperalgesia of the body.

References


DeJong’s Textbook of Neurology.
Youman's Textbook of Neurological Surgery.


Correct answers: 1B, 2D, 3C

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