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Review Article

Lateral medullary syndrome: an unwanted ischemia to identify early

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ABSTRACT

Lateral medullary syndrome also referred to as Wallenberg syndrome, posterior inferior cerebellar artery (PICA) syndrome, or vertebral artery syndrome is a cluster of neurological symptoms and signs brought on by obstructions in the blood vessels supplying the medulla, which causes ischemia or infarction of the brainstem. The vertebral artery or the posterior inferior cerebellar artery are the arteries most frequently affected with lateral medullary syndrome. The most prevalent symptom is transient ischemic attack (TIA) with dizziness or vertigo and atherosclerosis in the posterior cerebral circulation being the most common cause. Lateral Medullary Syndrome patients suffer from strokes or infarction and also present with vomiting, nausea, gait impairment, instability, hoarseness, and swallowing difficulties. Depending on the particular nuclei and fibers involved, different indications will appear. CT/MRI imaging is used to diagnose conditions. The majority of management is supportive, including risk factor reduction for additional ischemia events as well as speech and occupational therapy following an acute intervention. In this review article, we discuss the etiopathogenesis and factors leading to lateral medullary syndrome along with a comprehensive discussion on its clinical features, challenges in diagnosis, and treatment.

Keywords: Wallenburg, Lateral medulla, Stroke, Vertebral artery, Horner's syndrome

INTRODUCTION

The neurological disorder known as lateral medullary syndrome is brought on by ischemia in the brainstem's medulla oblongata, which causes a variety of symptoms. The vertebral artery or the posterior inferior cerebellar artery are the two most frequently blocked blood vessels that trigger ischemia.[1] Other names for lateral medullary syndrome include Wallenberg's syndrome, vertebral artery syndrome, or posterior inferior cerebellar artery (PICA) syndrome.[2] The lateral medullary syndrome is named after famous neurosurgeon Adolf Wallenberg for his clinical explanations of this syndrome. A 38-year-old man with symptoms of vertigo, numbness, loss of pain and temperature sensitivity, multiple site paralysis, ataxia, and other conditions served as Wallenberg's first patient in 1885. He was able to identify the patient's lesion in the lateral medulla and link it to a blockage of the ipsilateral posterior inferior cerebral artery. He was able to support his conclusions with postmortem evidence after his patient passed away in 1899.[3] He kept working with numerous individuals, and by 1922, he had documented the clinicopathological correlations for 15 patients. All these individuals had the same pathophysiology leading to Wallenburg's discovery of this condition. The posterior inferior cerebellar artery is compromised, causing infarction of the lateral medulla, which results in the lateral medullary (Wallenberg)

syndrome. Patients with the full syndrome exhibit ipsilateral Horner syndrome, ipsilateral cerebellar symptoms, and crossing hemisensory dysfunction (ipsilateral face, contralateral body). According to a historical report from 1961, the syndrome is thought to be responsible for 2.5% of ischemic strokes. However, considering the difficulties in making a diagnosis, this estimate is probably too low.[1] For the purpose of making a diagnosis and selecting the patients who need immediate neuroimaging and acute stroke treatments, it is essential to assess clinical signs and symptoms accurately. Since the lesion is caudal to the cochlear nerve entry zone and cochlear nucleus and does not affect the AICA, hearing loss is not noticed. Some patients will experience a noticeable motor disturbance that causes their body to slant toward the lesion side as if being pulled there by an external force. The oculomotor system is also impacted by this type of lateral pulsion, resulting in abnormally small saccades away from the lesion and excessively big saccades directed toward its side. Most Wallenberg syndrome patients continue to experience neurologic impairments months or even years after acute infarction.

Search strategy and selection criteria

References for this Review were identified by searches of PubMed, Scopus, and Cochrane Library between 1990 and August 2022, and references from relevant articles. The search terms “Wallenburg”, “lateral medullary syndrome”, “stroke”, “vertebral artery”, and “posterior inferior cerebellar artery” were used. There were no language restrictions. The final reference list was generated on the basis of relevance to the topics covered in this Review.

Etiopathogenesis

Wallenberg syndrome is most frequently brought on by an ischemic stroke of the brain stem, which frequently results from a thrombus or an embolism. Other, less frequent causes include: mechanical injury to the vertebral artery in the neck, arteriovenous malformations (AVMs), vertebral arteritis (inflammation of the artery wall), aneurysm of the vertebral artery, head injury, multiple sclerosis (MS). The three most prevalent risk factors for Wallenberg syndrome are diabetes, cigarette smoking, and hypertension (high blood pressure). Connective tissue abnormalities such as Marfans syndrome, Ehlers-Danlos syndrome, and fibromuscular dysplasia are additional, less frequent risk factors. Vertebral artery dissection is the most typical cause of Wallenberg syndrome in younger persons.

Clinical features

A wedge of the dorsolateral medulla just posterior to the olive makes up the zone of infarction that causes the lateral medullary syndrome (Wallenberg syndrome). Rarely does occlusion of the PICA cause the condition; instead, it typically comes from obstruction of the ipsilateral vertebral artery. Vertigo, diplopia, dysarthria, Horner's syndrome, and numbness (ipsilateral face and contralateral limb) are the hallmarks of lateral medullary (or Wallenberg's) syndrome, which generally lacks any limb weakening. Due to the distinctive appearance, restricted blood supply, and very tiny area of involvement, localization in this illness is simple. However, lateral medullary syndrome is linked to hemiplegia in Opalski syndrome and Babinski-Nageotte syndrome.[4] These two are variants of the lateral medullary syndrome. Due to the infarct's caudal extension to engage the corticospinal fibers after the pyramidal decussation, Opalski syndrome causes ipsilateral hemiplegia. Because the pyramidal tract is impacted before decussation, Babinski-Nageotte syndrome results in contralateral hemiparesis.[5]

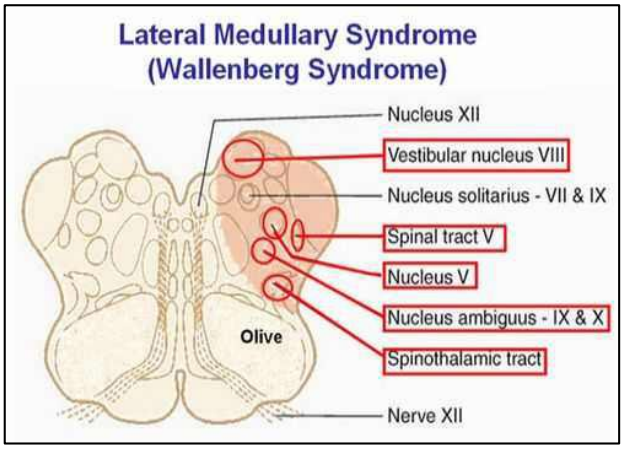

Figure 1: Lateral medullary syndrome affected structures.

The brain's lateral medulla is damaged in the lateral medullary syndrome, which results in a constellation of neurologic symptoms.[6] On the opposite side of the infarction, sensory impairments may impact the torso and the extremities. On the contralateral side of the body, pain and temperature perception may be lost.[7,8] In most cases, there are additional sensory abnormalities that affect the facial and cranial nerves on the same side as the infarct.[9,10] On the ipsilateral (same) side of the face, pain and warmth sensitivity have been lost. Swallowing problems, slurred speech, face pain, vertigo, Horner syndrome, and perhaps palatal myoclonus are all examples of clinical symptoms. Due to ipsilateral vocal fold paralysis, palatal and nasal regurgitation, dysphonia, dysphagia, and dysphonia are otolaryngological characteristics.[11] Dysphonia, dysphagia, and nasal regurgitation due to ipsilateral vocal fold paralysis, palatal, and pharyngeal paresis are otolaryngological characteristics. It might also be linked to airway blockage and obstructive sleep apnea.[5] The predominant clinical sign of a brainstem injury in this syndrome is "crossed hemiparesis or hemianesthesia," and complete Wallenberg syndrome is unusual. Gait instability is the most common sequelae and the majority of patients are

able to resume their normal activities with satisfaction in the projected 60,000 new cases of the Wallenburg syndrome that usually occur every year in the United States. Patients frequently experience ataxia, trouble walking or balancing, or a variation in an object's temperature depending on which side of the body it is touching. Some patients may walk slantingly, exhibit skew deviation, or have the impression that the space is tilting.[7] Nystagmus is frequently linked to dizziness attacks. Because the Deiters' nucleus region is involved, these vertigo spells can cause falling.[8] Ataxia usually results due to injury to the inferior cerebellar peduncle or the cerebellum itself. Miosis, anhidrosis, and partial ptosis are signs of hypothalamospinal fiber damage that affects the sympathetic nervous system relay and is related to Horner's syndrome. Other signs and symptoms include hoarseness, nausea, vomiting, a reduction in sweating, issues with feeling hot or cold, dizziness, difficulties walking, and problems with balance. Additionally, bradycardia, a slow heartbeat, and changes in the patient's average blood pressure might be brought on by lateral medullary syndrome.[6] Based on anatomical location: involvement of the nucleus ambiguous causes dysphagia, dysphonia, and dysarthria, laryngeal, palatal, and pharyngeal paralysis, involvement of the trigeminal nucleus causes ipsilateral facial and corneal anaesthesia, involvement of the spinothalamic tract causes loss of pain and temperature sensation on the opposite side of the body, involvement of the cerebellum causes ataxia, involvement of hypothalamic fibres causes Horners syndrome and aberrant sympathetic nervous system, nystagmus and vertigo are caused due to involvement of the Deiters' nucleus and other vestibular nuclei and palatal myoclonus due to involvement of the central tegmental tract.

Diagnosis

A quick assessment, a precise determination of when symptoms first appeared, and the conduct of a neurologic examination with an emphasis on differentiating infarction from mimic diseases are all necessary when treating a patient with the suspected lateral medullary syndrome.[12] A straightforward three-step oculomotor examination added to a prospective cross-sectional study of 101 patients presenting with "acute vestibular syndrome" identified patients with stroke with 100% sensitivity and 96% specificity, as well as clinically identified all 17 patients with the lateral medullary syndrome.[13] The acronym HINTS (head-impulse-nystagmus-test of-skew) sums up these bedside maneuvers, which should be carried out on all patients exhibiting modest symptoms suggestive of posterior fossa infarction, such as solitary vertigo, nausea and vomiting, and gait intolerance. In individuals who have any of the following, a central cause should be assumed, and tests and treatment for an acute stroke should be considered: a typical horizontal head impulse test (gaze is maintained with a passive horizontal head thrust); direction-changing nystagmus on eccentric gaze; skew deviation (vertical ocular misalignment demonstrated with cover uncover testing of each eye).[13] In order to rule out alternative diagnoses and check for potential stroke therapy contraindications such as intracerebral hemorrhage, focal compression, or herniation, all patients with suspected acute stroke should have immediate neuroimaging.[14] Unenhanced CT is an excellent tool for this job and can be combined with clinical evaluation to identify patients who should receive intravenous thrombolytic therapy. Additional neurovascular imaging should be done when necessary to check for vertebral artery dissection. Computed tomographic angiography (CTA) is the modality of choice in the majority of clinics since it is widely accessible, quick to acquire images, and has few drawbacks for patients without renal impairment.[15] It is crucial to keep in mind that neither CT nor CTA are particularly sensitive for diagnosing acute posterior fossa ischemic stroke; according to one study, only 33% of acute brainstem infarctions verified by MRI were picked up by CT.[16] With an overall sensitivity of 83% and a specificity of 96%, MRI (diffusion-weighted sequences MRA) is the gold standard diagnostic for the diagnosis of acute stroke.[17] However, even MRI may not be accurate in cases of acute lateral medullary infarction. The sensitivity of MRI after 48 hours of symptom onset drops to 72% in the subset of patients with brainstem infarction, highlighting the value of clinical judgment in assessing this patient population. The goal of emergency department management, according to the Canadian best practice recommendations for stroke care, is to quickly assess patients with suspected acute strokes in order to identify those who would benefit from intravenous tissue plasminogen activator treatment within 60 minutes of presentation. According to current guidelines, all patients who report within 4.5 hours of the onset of symptoms should be given the option of receiving intravenous tissue plasminogen activator therapy after consulting with specialty services that handle acute stroke care (and via direct consultation or TeleStroke networks where available).[18] Management choices in these situations should be determined on an individual basis until more data are available, taking into account the clinical presentation, the level of vascular damage, and the accessibility of other endovascular therapies. Identifying individuals with infarction in those who are not candidates for thrombolytic therapy is still crucial because these patients should be brought to the hospital for further evaluation and treatment aimed at preventing further strokes.

Treatment and prognosis

How soon lateral medullary syndrome is discovered will determine how it is treated. Focusing on symptom alleviation and active rehabilitation to assist patients in getting back to their normal activities are key components of the treatment for the lateral medullary syndrome. Speech therapy is administered to a lot of patients. Patients may exhibit depressed mood and social disengagement after the initial flurry of symptoms. In

more extreme situations, a gastrostomy or the placement of a feeding tube in the mouth may be required if swallowing is compromised. In some circumstances, medication may be used to lessen or get rid of lingering discomfort.[11] Anti-epileptic drugs like gabapentin have been successful in some studies in reducing the chronic neuropathic pain linked to the illness.

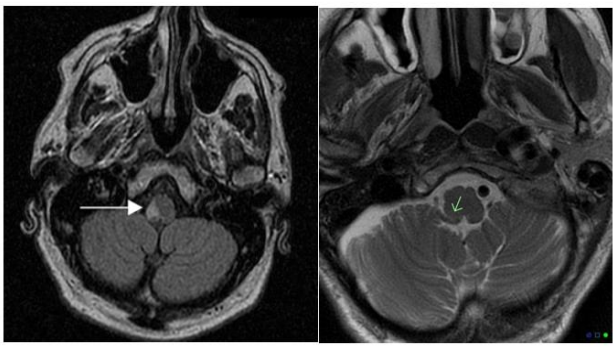

Figure 2: Axial FLAIR picture demonstrating a high signal focus consistent with an infarct in the dorsolateral part of the right side of the medulla oblongata (arrow).[19]

To reduce the chance of having another stroke, long-term treatment typically entails taking statins for the rest of their lives along with antiplatelet medications like aspirin or clopidogrel. If atrial fibrillation is present, warfarin is prescribed. In order to control high blood pressure and stroke risk factors, additional drugs could be required. In order to break up the infarction, restore the blood flow, and avoid further infarctions, a patient may be given a prescription for a blood thinner. Repetitive transcranial magnetic stimulation (RTMS) has been demonstrated to aid in the treatment of dysphagia symptoms. Because of the degree of the blockage and the location of the infarction, some people will always experience residual symptoms, making treatment for this illness unsettling.[11] Following a stroke, two people may have the identical initial symptoms, but one patient may recover completely while the other remains severely incapacitated. This discrepancy in results could be caused by a number of factors, including the size, location, and extent of the infarction. The prognosis for recovery from lateral medullary infarction remains positive with adequate medications, clinical monitoring, and post-stroke care. Over 85% of patients achieve functional independence with ambulation within a year, and the majority of patients have very minor limitations at six months.[18]

CONCLUSION

The Wallenburg syndrome typically results from obstruction of the ipsilateral vertebral artery. The classic presentation consists of sensory impairments affecting the face and cranial nerves ipsilateral to the infarct as well as sensory and motor deficits affecting the trunk and extremities on the opposite side of the infarct. It's crucial for patient care that the signs and symptoms of the lateral medullary syndrome are recognized. In order to rule out different diagnoses and treatments for acute stroke that are contraindicated, affected individuals should obtain immediate neuroimaging. Lateral medullary syndrome stands out from other posterior circulation strokes due to its positive prognosis.

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