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Hemorrhagic Wallenberg Syndrome Accompanied by Horner Syndrome at Young Age

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ABSTRACT

Introduction: Wallenberg syndrome is a rare syndrome involving the medulla oblongata, an uncommon hemorrhage site. It is also known as lateral medullary syndrome or posterior inferior cerebellar artery syndrome. Wallenberg syndrome has several symptoms that can be divided into vestibulo-cerebellar, bulbar, autonomic, sensory, and atypical. **Case:** A 27-year-old man came to the emergency unit complaining of dizziness and swinging, as well as a left eyelid that did not open completely, a lack of sweat on the left side of the face, a hoarse voice, swallowing disturbances, hiccups, decreased hearing in the left ear, and a lack of sensation on the left side of the face and the right side of the body. The patient tended to fall to the left and was unsteady when standing. Physical examination revealed Horner syndrome, bidirectional nystagmus, left dysdiadochokinesia, and left ataxia. A CT scan of the head showed a left-sided medulla oblongata hemorrhage. The patient was given conservative therapy, feeding through a nasogastric tube, and medical rehabilitation. A repeat CT scan of the head was done one week later, and it was discovered that the hemorrhage in the medulla oblongata had begun to absorb. **Conclusion:** Wallenberg syndrome, with a manifestation of hemorrhagic stroke in the medulla oblongata, can occur at a young age with various clinical symptoms. The diagnosis of Wallenberg syndrome is made based on clinical symptoms and imaging findings. Conservative therapy using medicines and medical rehabilitation can provide a speedy recovery. An incorrect diagnosis is a major hazard to patient safety and can have serious consequences, including permanent disability or death.

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INTRODUCTION

Wallenberg syndrome is a rare syndrome involving the medulla oblongata with clinical manifestations of loss of temperature and pain sensation in the contralateral body, ipsilateral ataxia, nystagmus, loss of swallowing reflex, loss of taste, and Horner syndrome. It is also known as lateral medullary syndrome or posterior inferior cerebellar artery syndrome. It is rarely accompanied by hiccups or the cerebral-cardiac syndrome. The medulla oblongata is a rare site for hemorrhage.

Most Wallenberg syndrome cases are caused by occlusion of the vertebral artery or posterior inferior cerebellar artery. Brainstem hemorrhages commonly occur in the pons. A patient with Wallenberg syndrome is typically an elderly person with vascular risk factors. In this case, we discuss Wallenberg syndrome, caused by hemorrhagic stroke in the medulla oblongata in a young male.[1,2,3,4]

CASE

A 27-year-old man came to the emergency unit complaining of dizziness and swinging, as well as a left eyelid that did not open completely, a lack of sweat on the left side of the face, a hoarse voice, swallowing disturbances, hiccups, decreased hearing in the left ear, and a lack of sensation on the left side of the face and the right side of the body. The patient tended to fall to the left and was unsteady when standing. There was no history of hypertension or use of anticoagulants, and he was in good health. Vital signs were within normal limits with a blood pressure of 120/80 mmHg, a pulse of 98 per minute, a respiration rate of 20 per minute, a temperature of 36.7 °C, and an NRS pain scale of 5. He was admitted with a GCS of 15. Physical examination revealed Horner syndrome (left pupillary miosis, left facial anhidrosis, and left ptosis), bidirectional nystagmus, left dysdiadochokinesia, and left ataxia.

A head CT scan showed left-sided hemorrhage in the medulla oblongata with an estimated bleeding volume of 3 cc. The patient was diagnosed with central vertigo, ataxia, Horner syndrome, left trigeminal nerve palsy, left vestibulocochlear nerve palsy, left glossopharyngeal nerve palsy, left vagus nerve palsy, left central hypoglossal nerve palsy, right hypoalgesia, bidirectional nystagmus, and intracerebral hemorrhage in the medulla oblongata (Figure 1). He was admitted with an NIHSS score of 2 (left extremity ataxia and sensory loss in the right extremity).

The patient was given conservative therapy, feeding through a nasogastric tube, and medical rehabilitation. After one week of treatment, another head CT scan and cerebral CT angiogram were done, and it appears that the hemorrhage in the medulla oblongata has begun to be absorbed (Figure 2 and 3). The patient was discharged from the hospital and further treated as an outpatient, with considerable clinical improvements such as reduced dizziness, re-widening eyelids, a less hoarse voice, no longer having hiccups, and right-sided sensory loss that began to decrease. He returned home with the nasogastric tube still in place, and his NIHSS score was 2, but the sensory impairment in the right extremity began to improve. After three months, the patient had an mRS (Modified Rankin Scale) score of 2 (mild disability).

DISCUSSION

German internist Adolf Wallenberg described the clinical manifestation of Wallenberg syndrome in 1895. Brain stem hemorrhages often occur in the pons, whereas medullary hemorrhages (Wallenberg syndrome or lateral medullary syndrome) are rare. The frequency of hemorrhagic medulla oblongata ranges from 0.1 to 0.5% of the total intraparenchymal hemorrhage. The reason for the low occurrence of medulla oblongata hemorrhage is not clearly understood, but it is believed to be related to the regional anatomy or hemodynamic characteristics of the perforating arteries in the medulla oblongata.

Most perforating arteries run horizontally after leaving the main arteries. The perforating vessels originate from the main cerebral arteries as small branches at the base of the brain. The vertebral artery perforating branches originate from the parent artery or its anterolateral or lateral pial branches. The vertebral arteries are 1.2 in number and 314 μm in diameter on average. Below the vertebral artery–basilar artery junction, the perforator artery enters the anterior median sulcus and supplies the paramedian part of the medulla oblongata.[3,5,6,7] The perforator arteries branch into the anterolateral branches (50%) that cross the pyramid of the medulla. When the vessels are absent on one side, they are supplemented by the perforator arteries of the adjacent arteries. The location and symptoms of the ischaemic area are determined not only by the luminal diameter but also by the branching pattern and position of the perforator artery. The medulla oblongata consists of a mass of neurons responsible for cardiac, respiratory, and vasomotor autonomic functions.[3,5,6,7]

In general, the clinical features of stroke in the anterior and posterior circulations have many similarities. Still, there are anatomical differences between the carotid and vertebrobasilar vascular anatomy that affect some of the differences in how a stroke in the posterior circulation is conceptualized. The posterior circulation consists of the vertebral arteries originating from the subclavian arteries, the

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basilar artery, three paired cerebellar arteries, and the posterior cerebral arteries. The basilar artery is formed by the union of two bilateral vertebral arteries. The basilar artery supplies the brainstem, occipital lobes, and thalamus.

The paramedian and lateral bulbar arteries supply vascular flow to the medulla oblongata. The paramedian bulbar arteries arise from the vertebral arteries and supply the medial part of the medulla oblongata. The lateral bulbar branches originate from the vertebral artery or the posterior inferior cerebellar artery and supply the medulla's lateral aspect (Figure 4). The complex and functional anatomy of the brainstem structure may make it difficult to localize clinical signs and identify the site in the posterior circulation.[8,9,10]

The most prevalent risk factors for Wallenberg syndrome are high blood pressure, diabetes, and smoking. Wallenberg syndrome can also be caused by Marfan syndrome, Ehlers-Danlos syndrome, fibromuscular dysplasia, and vertebral artery dissection.[11]

Hemorrhagic stroke in the medulla oblongata has several main symptoms. Dysarthria and dysphagia are bulbar symptoms, whereas nausea or vomiting, dizziness, swaying, and diplopia are vestibulo-cerebellar symptoms. Autonomic symptoms include Horner syndrome and hiccups, while sensory symptoms include pain and temperature loss in the contralateral body and ipsilateral face. However, hemiparesis and facial nerve palsy are uncommon. Contralateral hemiparesis could result from a caudal extension of the pyramidal tract before the decussation in the medulla oblongata.[11,12,13]

A study by Zhang *et al.* reported a healthy 19-year-old woman who presented with nausea, vomiting, dizziness, and vertigo. It progressively worsened with hoarseness, swallowing disturbance, imbalance, and numbness of the right face and left side limbs and trunk with an acute onset. The diagnosis of Wallenberg syndrome was established by clinical manifestations and magnetic resonance imaging (MRI) with computed tomography (CT) investigation. Supportive treatment was given to her, and the patient's neurologic status improved after 40 days of treatment.[1]

A similar study by Ueda *et al*. reported a 48-year-old normotensive woman with dizziness, right-sided ataxia, and facial numbness. On a T2-weighted image, brain magnetic resonance imaging (MRI) showed a right medullary hemorrhage and multiple cavernous malformations. Wallenberg syndrome was diagnosed using clinical manifestations and brain magnetic resonance imaging (MRI).[2]

In Wallenberg syndrome, damage to the cerebellum or inferior cerebellar peduncle results in ataxia. Damage to the hypothalamospinal fibers (sympathetic tract or oculosympathetic pathway) results in Horner syndrome. The sympathetic tract innervates sweat glands on the ipsilateral body and face, dilator muscles in the ipsilateral eye, and retractor muscles in the ipsilateral upper and lower eyelids (Figure 5).[14]

Some ocular movement abnormalities are associated with Wallenberg syndromes, such as horizontal, vertical, and torsional nystagmus, gaze-evoked nystagmus, and ipsipulsion. The ipsipulsion movement specified in the Wallenberg syndrome is characterized by a tonic deviation of the eyes in the direction of the lesion location. Ocular contrapulsion followed by ocular ipsipulsion after seven days in Wallenberg syndrome has been reported.[15]

The involvement of the inferior vestibular nucleus causes nystagmus and vertigo, while the ambiguous nucleus causes hoarseness and an inability to swallow. Lack of sensation in the contralateral body happens due to the damage of the lateral spinothalamic tract. Whereas hearing loss is caused by damage to the cochlear nerve nucleus, and hiccups entail the reticular formation.[9,16,17]

The diagnostic approach in a patient with suspected Wallenberg syndrome requires rapid assessment, a precise time of symptom onset, and a proper neurologic examination. Imaging tests, such as a head CT scan or MRI, can detect hemorrhage in the medulla oblongata. Previous studies have concluded that worsening clinical symptoms after brainstem hemorrhage are associated with increased mortality and disability. Various factors can contribute to the worsening of clinical symptoms following a hemorrhage of the medulla oblongata, including active extravasation, the development of brain edema, and the mass effect surrounding the hemorrhage. There are no clear criteria for determining the hemorrhage outcome in the medulla oblongata using clinical and imaging parameters.[3,18]

The clinical signs and imaging results of the patient are used to make the diagnosis of Wallenberg syndrome. The resulting neurological deficit is related to damage to the lateral medulla oblongata, inferior cerebellar peduncle, trigeminal nerve nucleus, nucleus and fibers of the vagus, glossopharyngeal nerves, descending sympathetic tract, spinothalamic tract, and/or vestibular nucleus (Figure 6). The onset of the disease is acute. The most frequent complaints were dizziness, swinging, vertigo, nystagmus, hoarseness, and dysphagia.[3,9,19]

Kumral *et al*. stated that an increase in NIHSS at admission and the amount of bleeding can be an outcome predictor in patients with hemorrhage in the medulla oblongata. Increased NIHSS can predict the worsening of neurological deficits. However, to determine the size of the bleeding that can affect the outcome, it is necessary to measure the bleeding in the

horizontal (ventrodorsal) and vertical (rostrocaudal) directions. Where bleeding is in the rostrocaudal direction, it can affect more of the corticospinal tract and other anatomical structures, resulting in worse clinical symptoms.[3]

Hemorrhagic mechanisms in the medulla oblongata include ruptured vascular malformations, hypertension, and the use of anticoagulants. Hypertension is significantly more common in patients with poor outcomes from a variety of causes of bleeding. In medulla oblongata hemorrhages due to vascular malformations such as arteriovenous malformations (AVM), cavernous malformations usually cannot predict neurologic deficits and outcomes. The secondary hemorrhagic characteristics of vascular malformations differ from the mechanisms that occur due to hypertension. After three months, the general outcome of patients with cavernous malformations is better, and more cases can perform their daily activities independently.[3,20]

CONCLUSION

Brainstem hemorrhagic strokes happen less often but kill more people than other hemorrhagic strokes. Two important predictors are the level of consciousness and the size of the bleeding. To date, conservative therapy still plays an essential role in managing brainstem hemorrhagic stroke, and surgery is not recommended.[21]

Wallenberg syndrome with manifestations of hemorrhagic stroke Medulla oblongata can occur at a young age with various clinical symptoms. Conservative medical therapy and rehabilitation can provide a speedy recovery. Wallenberg syndrome has the potential to put patients in poor condition. Early detection, management, and rehabilitation are essential for post-stroke recovery. Rehabilitation therapy, which consists of physical, occupational, and speech therapy, will enhance the speed of recovery.[22]

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Author contributions
Author: examined the patient, contributed to data collection, manuscript description, and wrote the manuscript.
Co-author: examined the patient together with the author, contributed to data collection, and reviewed the manuscript.
All authors contributed to the article and agreed on the submitted version.

Conflict of Interest
The authors have no conflicts of interest regarding the publication of this article

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TABLES AND FIGURES

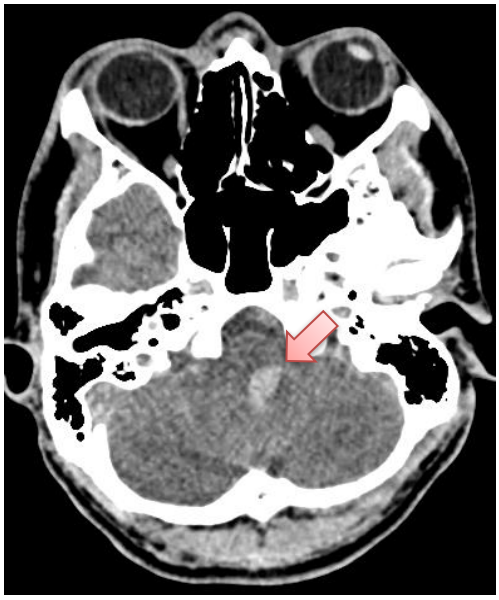

Figure 1. CT scan of the head showing hemorrhagic medulla oblongata

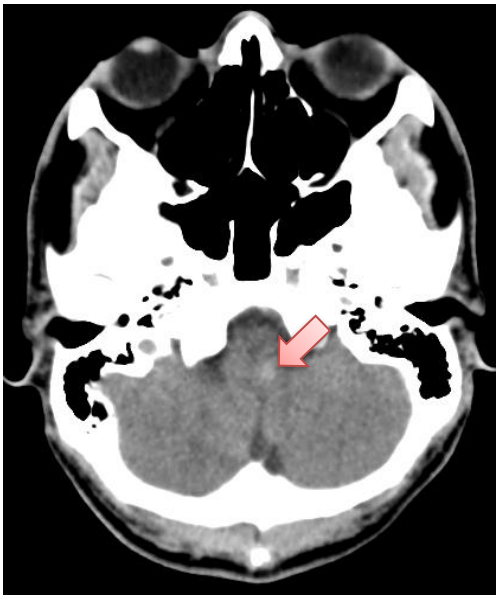

Figure 2. CT scan of the head after 7 days of treatment

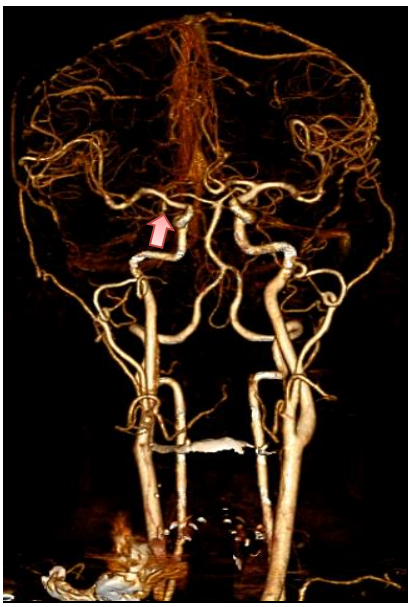

Figure 3. Cerebral CTA (1 week after onset). It appears that hemorrhagic has begun to be absorbed

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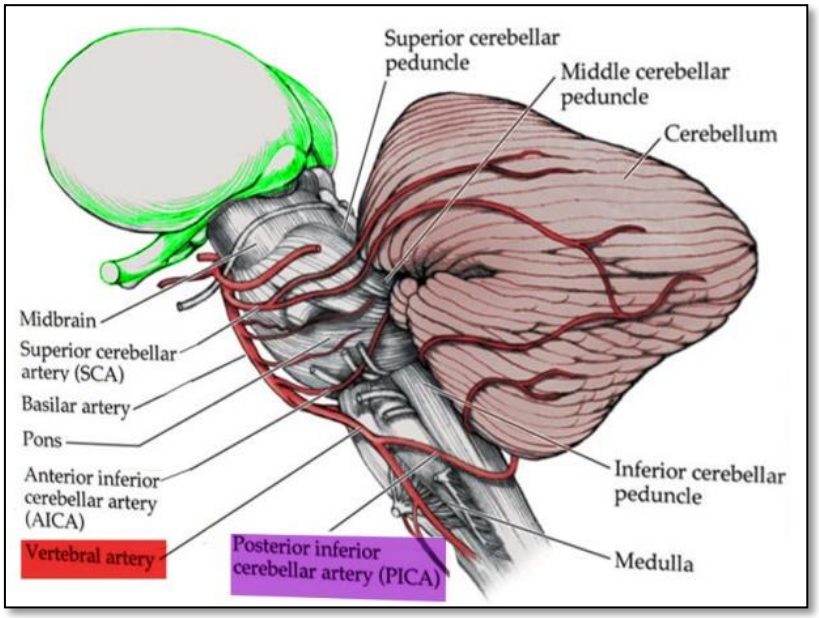

Figure 4. Medulla oblongata vasculature[10]

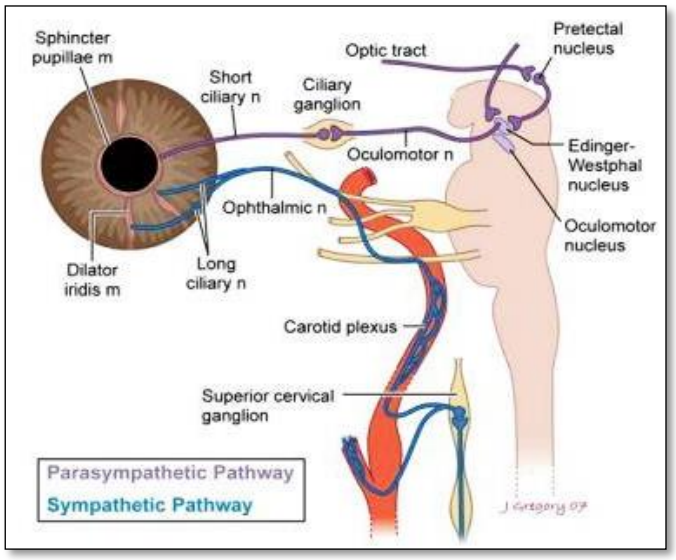

Figure 5. Structures affected in Horner syndrome[14]

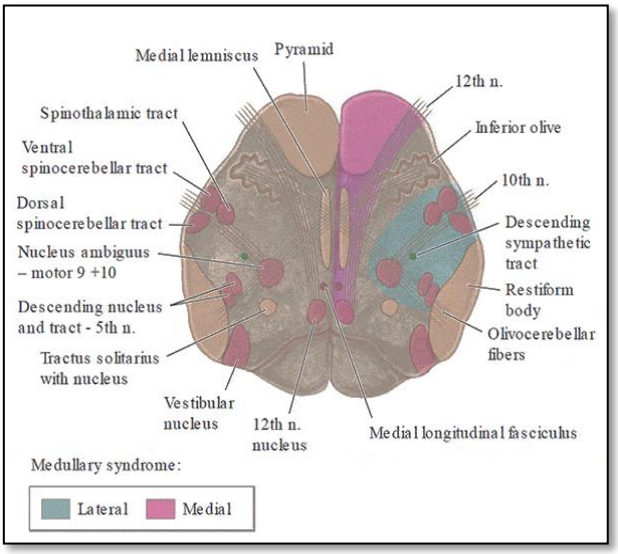

Figure 6. The affected structures of transverse section in Wallenberg's syndrome[13]