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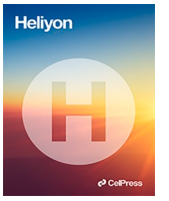

Opalski syndrome, a rare variant of wallenberg syndrome, the first case reported from Pakistan: A case report

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ABSTRACT

Background: Wallenberg syndrome, also known as a lateral medullary syndrome, is a rare neurological condition caused by an infarction in the brainstem's lateral medulla. There are subtypes of Wallenberg syndrome with distinctive and atypical symptoms, such as Opalski syndrome.

Case presentation: A 41-year-old hypertensive male arrived at the emergency department with abrupt onset of right-sided bodily weakness, vertigo, facial numbness, dysphagia, hoarseness of voice, and double vision. The neurological examination indicated right hemiparesis, right facial numbness, crossed sensory deficit, right limb ataxia, right uvulopalatal deviation, and vertical double vision. An MRI showed a lateral medullary infarct leading to the suspicion of the atypical lateral medullary syndrome. The patient was treated with physiotherapy and daily oral medications including aspirin, clopidogrel, atorvastatin, and Cap Risek. On follow-up 14 days later, the patient's condition had significantly improved.

Conclusion: This case study demonstrates the significance of recognizing atypical variants of Wallenberg syndrome, such as Opalski syndrome, in order to provide a correct diagnosis and the most effective treatment. Our patient's condition improved as a result of the therapy measures used, however, people with Opalski syndrome may have a poor prognosis and require continuous monitoring.

1. Introduction

Posterior circulation strokes make up 20 % of all ischemic strokes [1]. Vertebral artery disease involved in PICA is the most common cause of lateral medullary syndrome (67 %) [2]. Lateral medullary syndrome, also known as Wallenberg's syndrome, is a rare neurological disorder characterized by a set of symptoms caused by an infarction in the lateral medulla of the brainstem [3]. The symptoms typically include vertigo, diplopia, dysarthria, Horner's syndrome, and numbness on one side of the face and the opposite limb [4]. However, there are subtypes of lateral medullary syndromes such as Opalski syndrome which present with unique and atypical symptoms. In Opalski syndrome, the infarct extends caudally to involve the "corticospinal fibers" after the "pyramidal decussation", leading to ipsilateral hemiplegia [5].

Magnetic resonance imaging and pathological studies have shown that the lesions responsible for causing Opalski syndrome are

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located behind the lesions in Wallenberg syndrome and affect the corticospinal fibers below the pyramidal decussation [6]. According to reports, the most prevalent causes of the syndrome are dissection, stenosis, and occlusion of the vertebral artery [7]. Whether the hemiparesis observed in atypical Wallenberg syndrome is caused by an extension of a lateral medullary infarct into the pyramidal tract or by hypotonic dysregulation of motor loops is the subject of ongoing discussion. The MRI has not consistently revealed pyramidal tract involvement in Opalski syndrome patients [7].

Opalski syndrome is normally managed by a multidisciplinary team of stroke specialists, neurosurgeons, and radiologists [8]. The primary goal of treatment is to treat the underlying cause of the illness, which is usually a stroke caused by obstruction of the vertebral or posterior inferior cerebellar arteries [9]. Immediate management may involve supportive care such as airway, breathing, and circulatory support, as well as medication to prevent additional harm and enhance blood flow to the affected area [10]. Thrombolytic therapy, anticoagulation, and antiplatelet medicines, as well as blood pressure and glucose control, may be included. Rehabilitation is an important element of management as well [7]. Physical, occupational, and speech therapy may be used to assist patients in regaining function and independence. In some situations, surgery may be required to remove the clot or repair the damaged blood vessel [11].

This subtype of the lateral medullary syndrome can present diagnostic challenges and require a different management strategy. The present case study aims to describe the obstacles encountered in the diagnosis and management of a patient with Opalski syndrome and the strategies employed to overcome these obstacles.

The unique aspect of this case is that it is the first reported case from Pakistan. This adds to the medical literature by expanding the understanding of the prevalence and presentation of this rare condition in different regions of the world.

2. Case presentation

2.1. Patient history and timeline

On June 14, 2022, a 41-year-old nondiabetic nonsmoking male with a history of hypertension reported to our emergency department with abrupt onset of right-sided bodily weakness, vertigo, facial numbness, dysphagia, hoarseness of voice, and double vision. The patient was initially admitted to a different hospital, where it was stated that he had been experiencing right-sided bodily weakness for 15 days, as well as dysphagia and dysarthria for 4 days. Additionally, there he faced difficulties swallowing both liquid and solid food for four days. The patient did also experience two to three episodes of mild to moderate occipital headache. At first, the patient's condition improved for four to five days, but then he abruptly developed right-sided body weakness and vomiting. Following treatment at the previous hospital without any improvement, the patient was subsequently transferred to our medical facility. While the patient was under our care at the hospital, the weakness persisted for an additional two days. However, subsequently, the patient began to show signs of improvement.There was no history of allergies, hyperlipidemia, or arrhythmias, neither patient had any history of posterior circulation vessel stenosis, arteriovenous malformation or dissection. Patient initial National institute of stroke severity scale was 9.

2.1.1. Physical examination and neurological findings

The general physical examination of the patient indicated that his height and weight are average. At presentation, the patient's vital signs consisted of a blood pressure of 150/100 mmHg, a heart rate of 80 beats per minute, a respiratory rate of 20 breaths per minute, and a temperature of afebrile.

On neurological evaluation, it was determined that the patient had right hemiparesis with a right-side power of 4/5. In addition, he exhibited right facial numbness, crossed sensory deficit with right face sensory loss and left body below the neck, right limb ataxia, and right uvulopalatal deviation. Furthermore, the patient had double vision in the vertical gaze. But his score on the Glasgow coma scale was 15/15. Moreover, an assessment of the cranial nerves revealed that the ninth glossopharyngeal and tenth vagus nerves were damaged. There were no issues with the functioning of the cerebellum.

On the assessment of the extremities, the proximal and distal muscles of both the right upper limb and right lower limb had a power rating of 4/5. While the power of the distal and proximal muscles of the left lower and upper extremities was normal. His deep tendon reflexes were discovered to be intact, but his planters were deteriorating.

2.1.2. Laboratory investigations

Laboratory investigations were performed on June 14, 2022. All test results were determined to be within the normal range. However, there were no liver function tests performed.

Laboratory Investigations	Values
HB	14.8
Blood Glucose	120
Urea	36
Creatinine	0.7
Sodium	139
Potassium	4.2
Cholesterol	151

(*continued on next page*)

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Laboratory Investigations	Values
Triglycerides	67
Low-Density Lipoprotein	77
High-Density Lipoprotein	35
Calcium	9.9
Phosphate	4.3
Alkaline phosphate	4.3
Albumin	4
Prothrombin	15.8
International normalized ration	1.19
Partial Thromboplastin time	31

2.1.3. Imaging and diagnosis

In contrast, magnetic resonance imaging conducted on June 16, 2022, revealed lateral medullary infarction. However, the results of the computed tomography scan and chest X-ray were unremarkable. The chest X-ray performed on 15 June did not show any abnormalities or signs of pathology in the chest area, including the lungs, heart, and surrounding structures. Similarly, the CT scan of the brain performed on 15 June did not reveal any notable findings or abnormalities in the brain parenchyma, blood vessels, or other structures within the imaged area. On the basis of the patient's neurological symptoms such as right sided body weakness, crossed sensory deficit with right face sensory loss and left body below the neck, right limb ataxia, and right uvulopalatal deviation. and MRI findings (infarction in the focal area of diffusion restriction in the dorsolateral medulla on the right side), Opalski syndrome, a rare version of the atypical lateral medullary syndrome (Wallenberg syndrome), was suggested. Prior to the MRI, the differential diagnosis included lateral pontine syndrome and right cerebral infarction. These diagnoses were not determined solely based on their clinical presentations. Lateral pontine syndrome typically presents with ipsilateral loss of lacrimation, salivation, and paralysis of the lower and upper face, whereas right cerebral infarction is associated with symptoms like dysarthria, vomiting, and headache. However, in this case, the clinical presentation, along with the MRI findings, pointed toward the diagnosis of Opalski Syndrome Fig. A.

It is important to note that vascular imaging was not performed in this case to investigate the underlying cause of the patient's symptoms. The decision not to pursue vascular imaging was based on the clinical judgment of the doctors involved in the patient's care.

The initial treatment plan included Lopron 75mg once a day, Lowplat 75mg once a day, Injection of Citron 1g twice a day, Tab Lochol 40mg, Injection of Tanzo 2.25g Intravenously thrice a day, Nebulizer Ventolin 8 hourly, Tab Stomitil thrice a day, tab Moxelom Three times daily and intravenous Flagyl 500 mg injection three times daily. The patient was also prescribed daily oral aspirin 75 mg, clopidogrel 75 mg, atorvastatin 40 mg, and Cap Risek 40 mg, as well as chest physiotherapy. Following the treatment initiation, the patient showed significant improvement in his condition. At a follow-up 14 days after discharge, the patient's symptoms had notably improved. The right-sided bodily weakness, vertigo, facial numbness, dysphagia, hoarseness of voice, double vision, and occipital headache had either resolved or significantly decreased. The patient's power in the right limb had improved, and there were no further

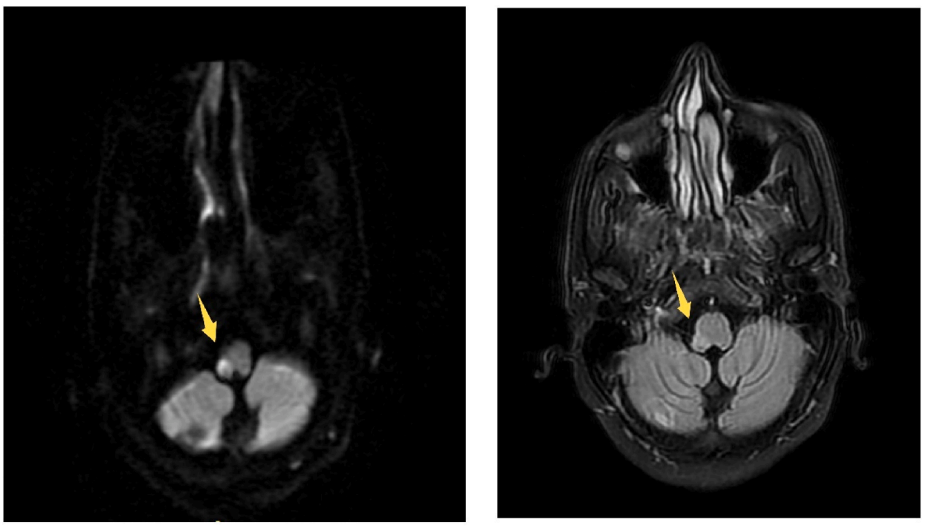

Fig A. Axial diffusion-weighted MRI image showing the focal area of diffusion restriction in the dorsolateral medulla on the right side confirms acute infarction. **Fig B:** Axial T2-weighted and FLAIR MRI images showing the small focal area of hyperintensity involving the dorsolateral medulla on the right side suggestive of the infarct.

episodes of vomiting reported.

The patient was discharged from the hospital on June 20, 2022, indicating that his condition had stabilized enough to continue recovery at home. A follow-up appointment was scheduled for the patient after a 14-day period. During the follow-up visit, a significant improvement was observed in the patient's condition, his NIHSS score was 0. Based on the presented signs, symptoms, and clinical examination, it was determined that the patient's condition was indicative of a vascular disease rather than a demyelinating disorder. Additionally, after the successful recovery, he did not come for further visits. **Discussion**.

This case report describes a case of a 41-year-old male with a history of hypertension who presented to the emergency room with abrupt onset of right-sided bodily weakness, vertigo, facial numbness, dysphagia, hoarseness of voice, and double vision. The patient also had mild to moderate occipital headaches and episodes of vomiting. These classical symptoms of opalski syndrome have also been reported in previous studies. According to the reports the traditional presentation consists of ipsilateral face numbness with crossover sensory abnormalities, notably loss of temperature and pain sensation affecting the trunk and limbs contralateral to the infarct [12]. The other symptoms include Vertigo, ipsilateral cerebellar abnormalities, nystagmus, dysphagia, Horner syndrome, and hoarseness [13]. The only difference between the presentation of our patient and the standard description of opalski syndrome was no involvement of Horner Syndrome, despite having classic crossed sensory involvement.

In terms of power, our patient had more power in extremities compared to the other case reports in which the patients had a power grade of 2/5 in the affected extremities [14]. Additionally, our patient also presented with PICA with involvement of the Vagus nerve and glossopharyngeal nerve as he had problems with speaking and swallowing. This involvement of cranial nerves has been supported by previous studies. Researchers describe that lateral medullary syndrome typically affects the "posterior inferior cerebellar artery" or the vertebral artery [1]. It can cause a variety of neurological symptoms, including damage to the "inferior cerebellar peduncle" and dorsolateral medulla, as well as to the descending spinal tract, the nucleus of the trigeminal nerve, fibers, and nuclei of the Glossopharyngeal and Vagus nerve, vestibular nuclei, spinothalamic tract and descending sympathetic tract fibers [2,15]. Similarly, in our case report, infarction of the dorsolateral medulla could be seen in the MRI scan.

In our case, laboratory investigations were within normal range, but an MRI revealed lateral medullary infarction. The diagnosis suggested was Opalski syndrome, a rare version of the atypical lateral medullary syndrome (Wallenberg syndrome). This has also been supported by studies in which MRI has been found to be the only reliable choice to diagnose this syndrome. As its clinical presentation is similar to the pontine syndrome and Wallenberg syndrome, it is hard to differentiate [16].

Additionally, it is unclear what causes weakness in lateral medullary infarcts [17]. Initially, Opalski hypothesized that it was produced by the spread of ischemia from the lateral medulla to the upper cervical cord, involving corticospinal fibers that are posterior to the pyramidal decussation [18]. He also suggested that the inclusion of the posterior spinal artery could be to blame [16]. Other hypotheses, such as the impaired medullary penetrating arteries originating from the anterior spinal artery or distal vertebral artery, which supplies the "pyramidal fibers" below the decussation, have been proposed [19]. Any damage to these structures might be responsible for weakness observed in Opalski syndrome. In our case, patient also observed unilateral weakness. Dhamoon et al. hypothesized that the involvement of pyramidal fibers may be due to regional perfusion failure of the border zone area in the spinal cord, which lies between the anterior and posterior spinal arteries and may be involved as a result of hemodynamic change caused by vertebral artery stenosis or occlusion [20].

It is also worth noting that in Opalski syndrome, hemiplegia occurs on the same side of the body as the brain lesion. This happens because the infarct extends downward, involving the corticospinal fibers located after the point where the fibers cross over (decussate) to the opposite side of the brain [21].

On the other hand, in other type of Wallenberg syndrome such as Babinski-Nageotte syndrome, there is hemiparesis (partial paralysis) on the opposite side of the body. This occurs because the pyramidal tract, which contains the corticospinal fibers, is affected before the point of decussation [22].The treatment options include conventional management and surgery. Decompression surgery has been shown to relieve microvascular compression and improve symptoms in some cases [11]. However, one case report found that symptoms did not improve after surgery [23]. Other studies suggest that conservative therapy without surgery may be effective and that surgical intervention should be used with caution [24]. In this specific case, the patient's symptoms improved and resolved within a month without surgery with complete resolution of symptoms at the follow-up.

The case report provides a detailed description of a patient's medical history, physical examination findings, and laboratory investigations.

The inclusion of imaging results, such as magnetic resonance imaging (MRI) and computed tomography (CT) scans, adds important diagnostic information to the report.

The report also includes a description of the treatment plan and the patient's response to it, which can be helpful for medical professionals treating similar cases. There were following limitations of this case report. The case report is limited to a single patient and therefore cannot be generalized to the broader population. There is no information on the patient's lifestyle or family history, which could potentially provide insight into the underlying cause of the condition. The report does not include information on long-term outcomes, such as the patient's ability to fully recover or any potential complications.

Opalski syndrome is an exceedingly rare form of Wallenberg syndrome, with only nine reported cases documented thus far. In fact, a review of the literature found only nine reported cases of Opalski syndrome worldwide, prior to the case reported from Pakistan. Therefore, the case reported from Pakistan is unique due to its rarity, and it is the first documented case of Opalski syndrome in Pakistan.

3. Conclusion

Lateral Medullary syndrome patients have very diverse clinical presentation and have one of rare variant known as Opalski syndrome which patients present with ipsilateral hemipersis. Opalski syndrome is rare variant of Wallenberg's syndrome which diagnosis is based upon presence of specific neurological symptoms i.e. hyperreflexia or Babinski sign. Proper neurological examination and correct radiological finding helps in diagnosis of this rare variant. Physiotherapy and medical management show improvement in patient condition but people with Opalski syndrome have poor prognosis and require regular monitoring.

Ethical Approval and consent to participate

NA.

Consent for publication

Written informed consent was obtained from the parent of patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.
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Data availability statement

No data was used for the research described in the article.

CRediT authorship contribution statement

Ubaid khan: Writing – review & editing, Writing – original draft. **Bilal Ahmad:** Data curation, Conceptualization. **Ayesha Aslam:** Funding acquisition, Formal analysis. **Aiman Muhammad:** Methodology, Investigation. **Javed Iqbal:** Supervision, Software, Resources.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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