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Cervicomedullary Cavernous Hemangioma Presenting as Spinal Shock Syndrome and Dysfunction: A Case Report and Review of Related Literature

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ABSTRACT

Introduction

Cavernomas in the brainstem and spinal cord are rare than their intracranial counterparts, and occurrences specifically at the cervicomedullary junction are infrequent. In this report, we present a case of a cervicomedullary cavernoma which manifested with spinal shock and dysfunction.

Case Report

We describe a patient who exhibited spinal shock syndrome and a stepwise decline in spinal cord function. A 33-year-old woman initially complained of right upper arm and occipital referred pain from the atlantoaxial region. Three days later, she experienced bowel and urinary incontinence. Subsequently, quadriparesis and numbness affecting all limbs developed after one week, followed by life-threatening respiratory depression after nine days. Magnetic resonance imaging revealed a lesion in the cervicomedullary junction, identified as a cavernous hemangioma. During the hospital stay, fragmented reflex activity gradually returned. Upon follow-up, the bowel and urinary incontinence, motor impairments, and sensory impairments showed improvement.

Conclusion

The proposed mechanism for the mass effect of this cavernous malformation on the spinal cord at the cervicomedullary junction was likely due to pressure effects caused by shifting dynamics. Understanding the natural history of cavernous malformations, regional neurovascular anatomy, safe entrance points to the brainstem, routes to the craniovertebral junction from the base of the skull, and specific microsurgical procedures for their removal are necessary for appropriate treatment. However, these considerations should be balanced against knowledge of the associated hazards and treatment recommendations.

Keywords: *Cavernoma, spinal shock, spine, cervicomedullary*

INTRODUCTION

Cavernous angiomas, also known as cavernomas, are vascular malformations primarily found in the central nervous system (CNS). They are more frequently observed in supratentorial locations compared to other regions of the CNS. However, spinal cavernomas (SC) are rare lesions, with an overall incidence rate of 0.04-0.05% in populations.[1] Among intramedullary cavernomas, the cervicomedullary junction is involved in 8% of cases, and the cervical region accounts for 32%.[2] Histologically, cavernomas are characterized by compact irregular sinusoidal vascular channels, lacking

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neuronal parenchyma between them. In this report, we present the case of a 33-year-old woman diagnosed with a cavernoma located at the cervicomedullary junction, based on her clinical and radiological evolution.

CASE REPORT

This is a case of a 33-year-old female patient who arrived at the Emergency department with a nine-day history of nape pain radiating to the right upper arm. The patient reported the onset of urinary and bowel incontinence, motor weakness, altered pain and temperature perception with bilateral distribution, which appeared three days after the initial symptoms. Subsequently, the patient experienced life-threatening respiratory depression on the day of the consultation. During the examination, the patient's blood pressure and cardiac rate were normal. The neurological exam showed well-preserved higher mental functions with no craniopathies. In motor examination, there was 1/5 muscle strength in the right upper extremities (C5-T1), 3/5 in the right lower extremities proximally (L2-L4), and 4/5 in the right lower extremities distally (L4-S1). Additionally, there was 4/5 muscle strength observed in both the left upper extremities (C5-T1) and lower extremities (L2-S1) in myotomal distribution during testing.

During the sensory system examination, the patient had a 30% deficit in altered perception of light touch, deep touch, pain, temperature on the right side of the body, and a 70% deficit on the left side. Vibration and proprioception were found to be intact. In reflex testing, the patient exhibited hyporeflexia in all extremities. However, fragmented reflex activity returned during the hospital stay, and upon follow-up, there was a resolution of incontinence, motor impairments, and sensory impairments.

Discussion

Cavernomas are rare congenital benign vascular malformations of the brain, accounting for 5-15% of all brain vascular malformations.[4,5,7] They originate from arrested development of blood vessel progenitors and are more commonly found in the supratentorial brain parenchyma 1 . Brain stem cavernomas (BSC) make up 10-15% of all intracerebral cavernomas, with the majority located in the pons and midbrain (95% of cases), while medullary cavernomas contribute only 5% to BSC.[3] The overall incidence rate of CNS cavernomas, including both the brain and spinal cord, is approximately 1.9 cases per 100,000 persons per year, with spinal cord lesions accounting for 3-5% of these cases.[8]

In this case report, the hemangioma was located at the caudal medulla, extending to the C4 region within the intramedullary location. Cavernomas exhibit a characteristic appearance on MRI, appearing as a well-circumscribed lesion with a heterogeneous core resembling popcorn. T1-weighted images show a dark rim of hemosiderin surrounding the lesion (Figure 2). However, the diagnosis can be challenging due to the potential presence of blood products that can obscure the typical MRI features during initial imaging (Figure 1).

Cavernomas are typically not visible on formal angiograms due to their slow blood flow, hence their designation as angiographically occult. Around 5% of patients with an intracranial cavernoma will also have a spinal cavernoma, while 45% of patients with a spinal cavernoma will have an accompanying intracranial cavernoma. It is exceptionally rare for patients to have multiple spinal cavernomas without any concurrent intracranial cavernomas.[9] Therefore, if a spinal cavernoma is suspected or identified, additional MRI imaging of the head should be considered. In the present case, a cavernoma was solely detected at the cervicomedullary junction.

In large retrospective reviews of patients with symptomatic spinal cord cavernomas, it has been observed that 36% to 58% of cases present with spinal cord hemorrhage.[12, 13] A more recent retrospective review of 33 patients found that the rate of symptomatic hemorrhage was 1.6% per lesion per year.[14] These estimates align closely with

Figure 1. MRI Cranial Axial DWI Image (A) GRE (B) showed susceptibility signal in cervicomedullary junction (CMJ) as well as in Axial SWI showing peripheral blooming (E, F). Axial ADC (C). T2 Axial Flair (D), showing hyperintense lesion in the caudal medulla.

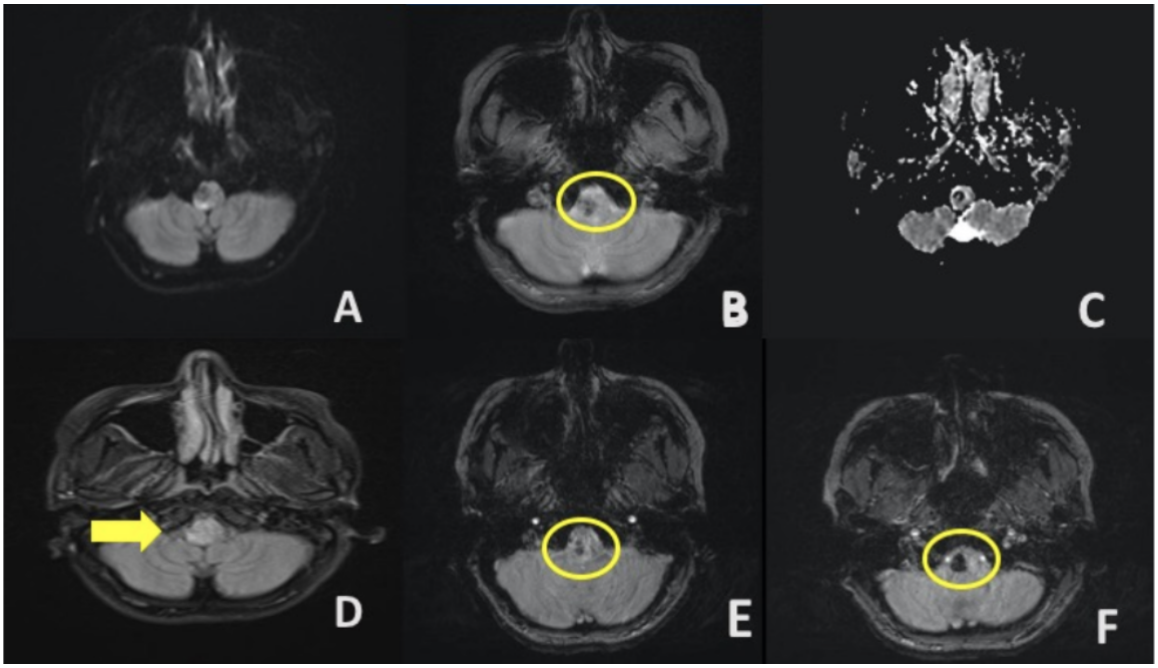
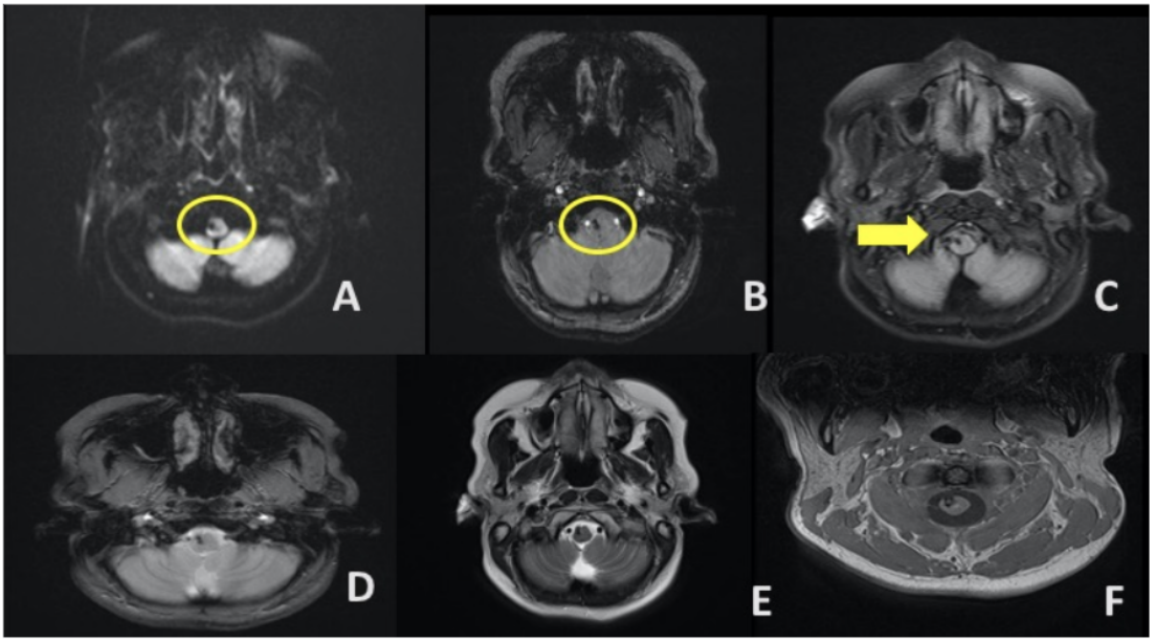


Figure 2. Follow-up study after 3 months: MRI Cranial Axial DWI Image (A) GRE (B) still showed a susceptibility signal in the cervicomedullary junction (CMJ). T2 Axial Flair Image (C), now shows a hypointense lesion in the caudal medulla. Axial T2 TRA (E) and T1 VIBE TRA (F).



the risk of bleeding from intracranial cavernomas, which ranges from 0.7% to 2.5% per lesion per year.[11]

The term "spinal shock" encompasses all phenomena related to the physiological or anatomical transection of the spinal cord, leading to temporary loss or depression of spinal reflex activity below the level of the injury. Typically, spinal shock is caused by a traumatic injury and manifests immediately. However, there have been reports of spinal shock occurring gradually over several hours in cases of progressive injury mechanisms. Spinal cord reflex arcs immediately above the level of injury, may also be severely depressed, exhibiting the Schiff-Sherrington phenomenon. The end of the spinal shock phase in spinal cord injury is indicated by the return of elicitable abnormal cutaneospinal or muscle spindle reflex arcs.

Autonomic reflex arcs, which involve relays to secondary ganglionic neurons outside the spinal cord, may be variably affected during spinal shock, and their recovery after the abatement of spinal shock is unpredictable. The returning spinal cord reflex arcs below the level of injury undergo irreversible changes and serve as the foundation for rehabilitation efforts.[6] Spinal cavernomas are less common compared to their intracranial counterparts.

Hemorrhage from an intramedullary cavernoma typically leads to sudden neurological decline, although microhemorrhages can also manifest as progressive myelopathy, as observed in this patient's case. Hematomyelia is primarily caused by intramedullary cavernomas but can also arise from extramedullary intradural cavernomas.

In the retrospective study by Ming-Guo Xie et al. that analyzed 53 cases of medullary cavernous malformations, achieving a high rate of gross total resection (98.1%). Postoperatively, respiratory dysfunction was observed in 43.4% of the patients, and dysphagia or cough was reported in 30.2% of the patients. The majority of patients (84%) had favorable outcomes, with a mean Modified Rankin Scale (MRS) score of 1.7, and 68% showed improvement in their condition. Age and prolonged tracheal intubation after surgery were identified as adverse factors for long-term outcomes.

On the other hand, the study by Si Zhang et al. focused on cavernous malformations involving the medulla oblongata in a smaller sample of 19 patients. Gross total resection was achieved in 17 cases (89.5%), while 2 cases had subtotal resection. Postoperatively, new onset or worsening neurologic deficits occurred in 31.6% of the patients. After a mean follow-up of 45.8 +/- 22.2 months, 52.6% of the patients showed improvement in neurological status, and 36.8% remained stable. The mean modified Rankin scale (mRS) score improved from 2.58 preoperatively to 1.84 at the recent follow-up. No hemorrhage or recurrence was observed during the follow-up period.

In comparing the two studies, both focused on the surgical treatment of cavernous malformations, but with variations in the specific location and sample size. Based on their findings, both studies suggest that surgical treatment can lead to favorable outcomes for patients with medullary cavernous malformations or cavernous malformations involving the medulla oblongata. Achieving gross total resection appears to be associated with better outcomes. However, the studies differ in terms of the specific outcomes evaluated, such as respiratory dysfunction and dysphagia in the study by Ming-Guo Xie et al., while focusing more on neurologic deficits in the study by Si Zhang et al.

Although surgical outcomes reported in the literature have been satisfying, surgical interventions have become increasingly contraindicated due to the benign clinical course observed in brainstem cavernomas, as in the case of this patient in whom conservative treatment was given. This consideration, combined with the high risk of surgical morbidity, has led to a shift away from surgical intervention.

Recommendations for surgical resection of symptomatic cavernoma involving the medulla oblongata emphasize the use of optimal approaches, feasible entry zones, and meticulous microsurgical techniques. The goal is to achieve safe resection and favorable outcomes. However, it is important to note that clinical features, surgical indications, timing, and microsurgical techniques of this special entity should be distinctive from the brainstem cavernous malformation in other sites.[15] In cases where the cavernoma is located in proximity to the pial or ependymal surface of the brainstem or where lesions are accessible through safe entry zones, surgical intervention is advisable. This is particularly true for lesions that have caused multiple significantly symptomatic hemorrhages and can be considered aggressive.[9]

However, the treatment approach remains controversial for deep-seated lesions that are located away from the surface of the brainstem and are inaccessible through safe entry zones.

Alternative treatments, such as radiosurgery and medications, are still subjects of debate and may be considered for lesions amenable to these approaches but pose a high risk with surgery.

CONCLUSION

This case and review of the literature discuss critical teaching points that help in understanding the pathology and unique clinical features of brain stem and spinal cavernomas, which exhibit a more dynamic course compared to supratentorial cavernomas.

Cervicomedullary cavernomas are rare and can manifest with varied clinical symptoms.

Cavernomas should be considered as a differential diagnosis in cases presenting with spinal shock and dysfunction. Surgical intervention should be performed only when there is a clear indication, as the surgery itself carries a significant risk of morbidity.

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