

Case Report

# Intermittent Myokymia as a Pointer to Hemangioblastoma of the Cervical Spine: A Case Report

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## Keywords

Hemangioblastoma · Brainstem tumors · Myokymia

## Abstract

Hemangioblastomas represent 3% of all central nervous system (CNS) tumors. The majority of CNS hemangioblastomas are infratentorial, with the cerebellum being the most frequent location, while 13% are found in the brainstem. Symptoms of brainstem hemangioblastomas can be very subtle and might therefore be overlooked or misinterpreted. We report the case of a patient with a hemangioblastoma at the junction of the medulla oblongata and the cervical spine and provide a brief review of the literature.

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## Introduction

Hemangioblastomas are benign, highly vascularized tumors representing 3% of all central nervous system (CNS) tumors [1, 2]. They are mostly sporadic, while 20–30% are associated with the von Hippel-Lindau syndrome. Approximately 70% of hemangioblastomas are infratentorial, with the cerebellum being the most frequent location (90%), 13% are found in the brainstem, and about 3–13% originate at the level of the spinal cord [2]. A less common location is the junction of the medulla and upper spinal cord [1]. Based on their location, Fukushima et al. [3] divided brainstem hemangioblastomas into three categories: type A are

hemangioblastomas attached to the floor of the fourth ventricle, while type E tumors are partially embedded in the floor of the fourth ventricle, and type I manifest as intramedullary hemangioblastomas of the medulla oblongata. We report the case of a patient with a hemangioblastoma at the junction of the medulla oblongata and the cervical spine and provide a brief review of the literature.

### Case Presentation

A 58-year-old female patient complained of long-standing intermittent paresthesia, such as tingling and numbness, and dysesthesia at the level of the left heel and the dorsolateral aspect of the left foot triggered by walking, sitting, and running. At times, the paresthesia also affected the left distal lower extremity, the left thigh, the left pelvis, and the ulnar region of the left hand. The duration of the paresthesia was always relatively brief, ranging from minutes to 1 h. Initial neurological examination showed reduced sensibility with a patchy distribution at the left lower extremity with intact vibration sense. There were no motor deficits or atrophy, and Lhermitte's sign was negative. A thorax and lumbar spine X-ray showed minor spondyloarthritic changes at the L4/5 and L5/S1 levels. A CT of the lumbar spine did not show a disc herniation or protrusion. Multiple sclerosis was excluded given the long-standing fluctuating course of symptoms lacking spinal or cerebral symptomatology.

As paresthesia started abruptly and due to its patchy distribution, Wartenberg's migratory sensory neuropathy [4] was suspected initially. Differential diagnoses consisted of hereditary neuropathy with liability to pressure palsies, insertional tendinopathy, radiculopathy, multiple sclerosis, and restless leg syndrome. Amyotrophic lateral sclerosis was subsequently excluded, once the patient additionally started to complain of intermittent small jerks in the right periorbital region. Electrophysiological workup did not show any organic correlate to further support peripheral neuropathy. As the patient suffered from panic attacks and hyperventilation, psychosomatization was believed to cause her complaints and treatment with paroxetine was initiated. Neurological examination at our outpatient clinic showed a partial central left facial palsy, a slight hyperreflexia on the left side, and hypesthesia and hypoalgesia in the left S1 dermatome. Further, a twitching in the left orbital region (myokymia) was noted. Based on this single finding, a cranial and cervical MRI (with and without contrast) was performed that showed a solid-cystic mass posteriorly at the junction between the medulla oblongata and the cervical spine extending cranially into the fourth ventricle (Fig. 1). The mass compressed to a high degree the medulla and the cervical myelon at the level of the foramen magnum. Surgical treatment was offered. The cystic aspect of the tumor was fully resected, while the vascular malformations were clipped. Postoperatively, no substantiated improvement was noted. Paresthesia and dysesthesia of the left heel persisted less intensely, whereas dysesthesia of the left hand had increased instead. Subjectively, the patient felt reduced force of the upper and lower extremities. Concentration and memory were slightly reduced.

### Discussion

Symptoms of brainstem hemangioblastomas can be very subtle and nonspecific. They can be easily overseen or misinterpreted. Clinical findings are headache, vomiting, long tract signs, ataxia, food aversion, vertigo, coma, paresthesia, diplopia, and dysphagia [3, 5, 6]. Cranial nerve palsy is less frequent than expected [5]. Over the years, our patient had suffered from

intermittent paresthesia and dysesthesias of her lower extremities. As the focus in the search of the diagnosis was mainly based on the sensory alterations, subtle clinical findings such as the jerks in the orbital region were overlooked and not recognized as potential brainstem findings in the first stage of diagnostics. Establishing the diagnosis of a hemangioblastoma was seemingly difficult in our case. Moreover, Pavesi et al. [7] reported a long time span from symptom onset to diagnosis in 14 patients with lower brainstem hemangioblastomas, which ranged between several weeks and years.

#### *Hemangioblastoma at the Craniocervical Junction*

Krishnan et al. [8] reported a 68-year-old male patient with left-sided hearing loss and spontaneous, self-limiting hiccups. The hemangioblastoma was located at the craniocervical junction compressing the brainstem and the fourth ventricle. A second male patient with hemangioblastoma of the medulla oblongata displacing the whole brainstem ventrally complained of general weakness, visual deterioration, and anesthesia of the left face for 3 months. In a third patient, repetitive and periodical severe headaches, vertigo, and nausea for 1 year were reported; the hemangioblastoma was located at the fourth ventricle and the craniocervical junction.

#### *Hemangioblastoma at the Fourth Ventricle*

Agrawal et al. [1] described a 43-year-old female with a mass lesion at the level of the caudal part of the fourth ventricle and the dorsal medulla that caused an obstructive hydrocephalus. The caudal medulla was displaced ventrally, and the vermis was displaced dorsally and superiorly. Clinically, she presented progressive weakness, dysphagia, and staggering gait. An emergency suboccipital craniectomy and tumor resection were needed once the patient became progressively drowsy.

A hemangioblastoma located at the dorsal medulla and extending into the fourth ventricle by displacing the caudal medulla inferiorly and dorsally was described by Fukushima et al. [3]. Symptoms were headache, nausea, hiccups, unsteady gait, and anorexia. Neurological examination was remarkable for papilledema, horizontal nystagmus on the right, left lateral gazes, absent gag reflex, dysphagia, ataxic gait, and motor incoordination. In a second patient described by Fukushima et al., a hemangioblastoma was diagnosed at the level of the dorsal medulla protruding into the fourth ventricle leading to loss of appetite.

### **Conclusion**

Lower brainstem hemangioblastomas can show subtle symptoms that might easily be overlooked or misinterpreted. Neurological findings that cannot be promptly allocated need, in all circumstances, imaging of the whole CNS axis.

### **Statement of Ethics**

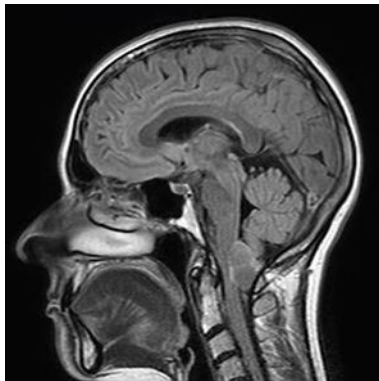
The authors certify that they have obtained the patient's consent. The patient has provided her consent for her images and clinical information to be published. The patient was informed that her name and initials will not be published and all information to the patient will remain anonymous.

## Disclosure Statement

The authors declare that they have no conflict of interest.

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**Fig. 1.** Sagittal MRI showing the tumor at the cervical spine.