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## Isolated hemangioblastoma of the cervical spinal cord: A case report and literature review

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## ABSTRACT

**INTRODUCTION:** Hemangioblastomas are benign, slow growing but highly vascularized tumors of the central nervous system, with the most common location of occurrence in the posterior fossa. Hemangioblastomas usually have an associated with patients that have Von-Hippel Lindau disease, resulting a germline mutation in the *VHL* tumor suppressor gene. Isolated or sporadic occurrences of hemangioblastomas are much more infrequent and typically respond well after surgery.

**PRESENTATION OF CASE:** We present case of a 22 year old female with worsening shoulder pain, decreased sensation in the hands and feet, and decreasing strength and was found to have a hemangioblastoma of the cervical spine.

**DISCUSSION:** The patient was treated with surgery and responded well to treatment. We also present a review of the literature on isolated occurrences of hemangioblastomas of the spinal cord.

**CONCLUSION:** Isolated hemangioblastoma are a rare tumor of the central nervous system and can be managed with surgery.

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## 1. Introduction

Hemangioblastomas are highly vascularized but benign tumors of the central nervous system [1]. These tumors can contain endothelial cells, pericytes, and stromal cells. Many of these cases of hemangioblastomas are sporadic, but an estimated 10–40 percent are associated with a germline mutation of the Von-Hippel Lindau (*VHL*) tumor suppressor gene [2]. In patients with sporadic and isolated hemangioblastomas, only four percent of patients had a detectable mutation in the *VHL* gene; however, these mutations (C162Y, D179N and R200W (in two patients)) were deemed to be hemangioblastoma-only or low penetrance mutations [3].

Hemangioblastomas can occur anywhere along the central nervous system such as the brain [4–6], optic nerve [7], retina [8], and spinal cord [9]. Considering the slow growing nature of these tumors, for patients that are asymptomatic, patients can be followed solely by observation. For the symptomatic patient, surgery is the preferred option, allowing for a complete resection with accompanying pathological analysis. In this report, we present a

case of a 22 year old female who presented with bilateral upper extremity weakness and paresthesias and was found to have a 1.5 centimeter mass in the C5–C6 disc space with an extensive syrinx extending inferior to the level of T8. We present the clinical presentation of the patient, the operative/surgical management of the patient, and a literature review of sporadic hemangioblastomas of the spinal cord.

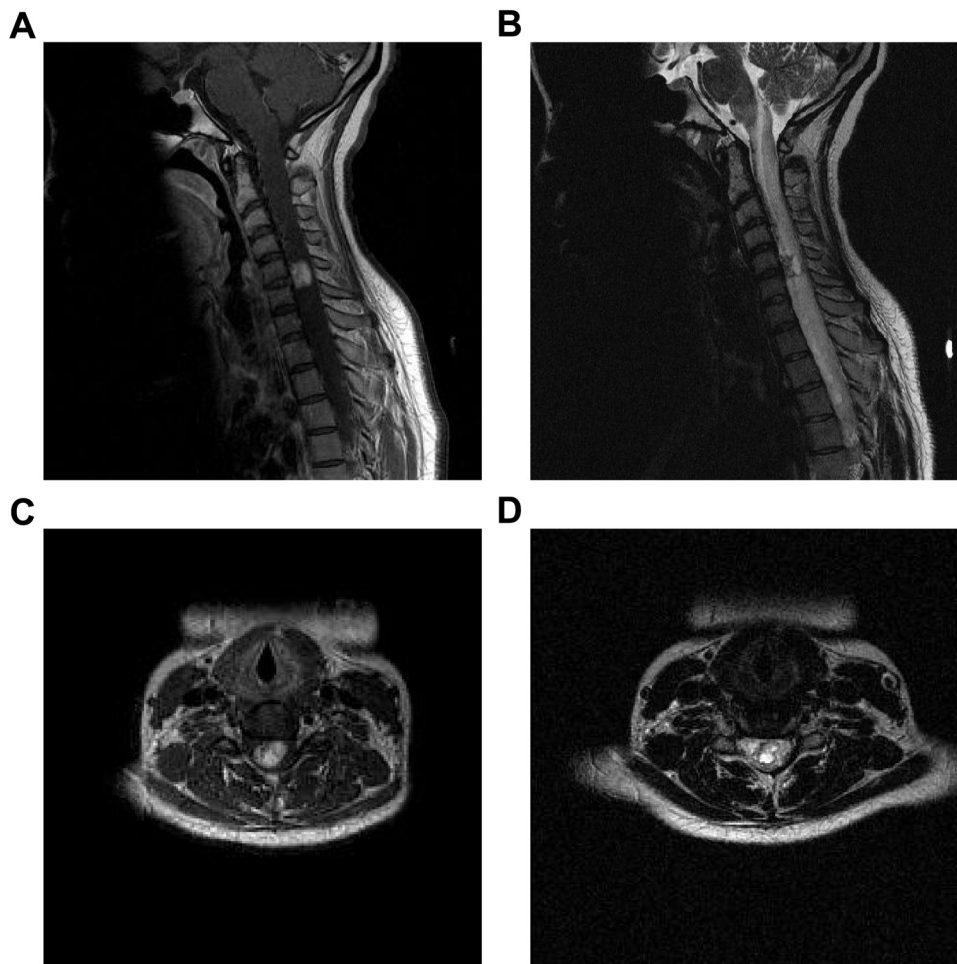
## 2. Case description

## 2.1. Preoperative information

A 22 year old African American female initially presented with decreased strength in her both hands, with the right worse than the left. She also endorsed pain in both of her shoulders. On examination, the patient had decreased sensation in both arms. The patient did not endorse any symptoms consistent with any lower spine pathology such as lower extremity weakness, bladder dysfunction, or sphincter dysfunction. The patient received a magnetic resonance imaging study of the cervical and thoracic spine that revealed a cervical canal/cord mass (Fig. 1). The mass is a focal lesion in the posterior C5–C6 disc space, measuring approximately in 1.3 × 1.0 centimeter, with extensive syrinx that terminates at the level of T8. The lesion appears to have a solid and cystic component with

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**Fig. 1.** Magnetic resonance imaging of the cervical cord of the hemangioblastoma upon admission. A 1.3 cm lesion is visualized in the posterior C5–C6 disc space, containing solid and cystic components with possible products of hemorrhage. An enlarged spinal cord with syrinx is visualized that tracks inferiorly to the thoracic cord. (A) Axial post-contrast T1 sequence at the level of C5; (B) Axial T2 sequence at the level of C5; (C) Sagittal post-contrast T1 sequence at the mid-sagittal line; (D) Sagittal T2 sequence at the mid-sagittal line.

possible products of hemorrhage. Magnetic resonance imaging of the lumbar spine and brain were largely unremarkable. The patient received an angiogram of the cervical spine that showed a markedly hypervascular intramedullary mass at the C5–6 level that corresponds to the abnormality seen on the magnetic resonance imaging with features, suggestive of ependymoma and hemangioblastoma.

## 2.2. Operative intervention and surgery

The patient then underwent a decompressive posterior cervical laminectomy of C4–C7 with fenestration of the syrinx, myelotomy with CO<sub>2</sub> laser, and resection of the tumor. A midline excision was made from the spinous process of C7/T1 and extended superiorly to approximately the level of C4. A C4 through C7 laminectomy was then performed after intraoperative fluoroscopy confirmed the location of the laminectomy. Then, the dura was opened in the midline, and the cervical cord was visualized to be extremely swollen. Using a CO<sub>2</sub> laser, a posterior myelotomy was performed, down to the medial inferior aspect of the syrinx. After resection and hemostasis was achieved, the dura was closed, and a dural regeneration matrix was overlaid. Then, a posterior lateral onlay fusion with bone marrow aspirate, autologous bone, demineralized bone matrix, and stem cell bone matrix was placed from the

level of C4 through T1. Somatosensory evoked response monitoring/electromyography showed baseline activities.

## 2.3. Surgical pathology

Tumor samples were collected in five pieces and were also reviewed at an outside institution. The surgical pathology returned as hemangioblastoma, World Health Organization grade I, with positive immunostaining for inhibin.

## 2.4. Postoperative course

After surgery, the patient was cared in the surgical intensive care unit with cervical spine precautions and placed on dexamethasone. The patient had 0/5 intrinsic strength in the bilateral upper extremity but was able to regain the majority of her strength in the immediate days after surgery. The patient received evaluation and treatment from physical therapy with adequate progress. Due to the socioeconomic status of the patient, the patient was discharged to a skilled nursing facility for continued rehabilitation, instead of her home. In the skilled nursing facility, the patient demonstrated improved strength with therapy. In the months following discharge, the patient continued to have improved strength and sensation. The patient was evaluated by medical and radiation

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