CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 26 (2016) 7–11

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

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

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ARTICLE INFO

Article history:
Received 20 April 2016
Received in revised form 4 July 2016
Accepted 4 July 2016
Available online 6 July 2016

Keywords:
Hemangioblastoma
Von-Hippel Lindau
Vascular tumor
Laminectomy
Myelotomy
Case report

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.

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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http://dx.doi.org/10.1016/j.ijscr.2016.07.002

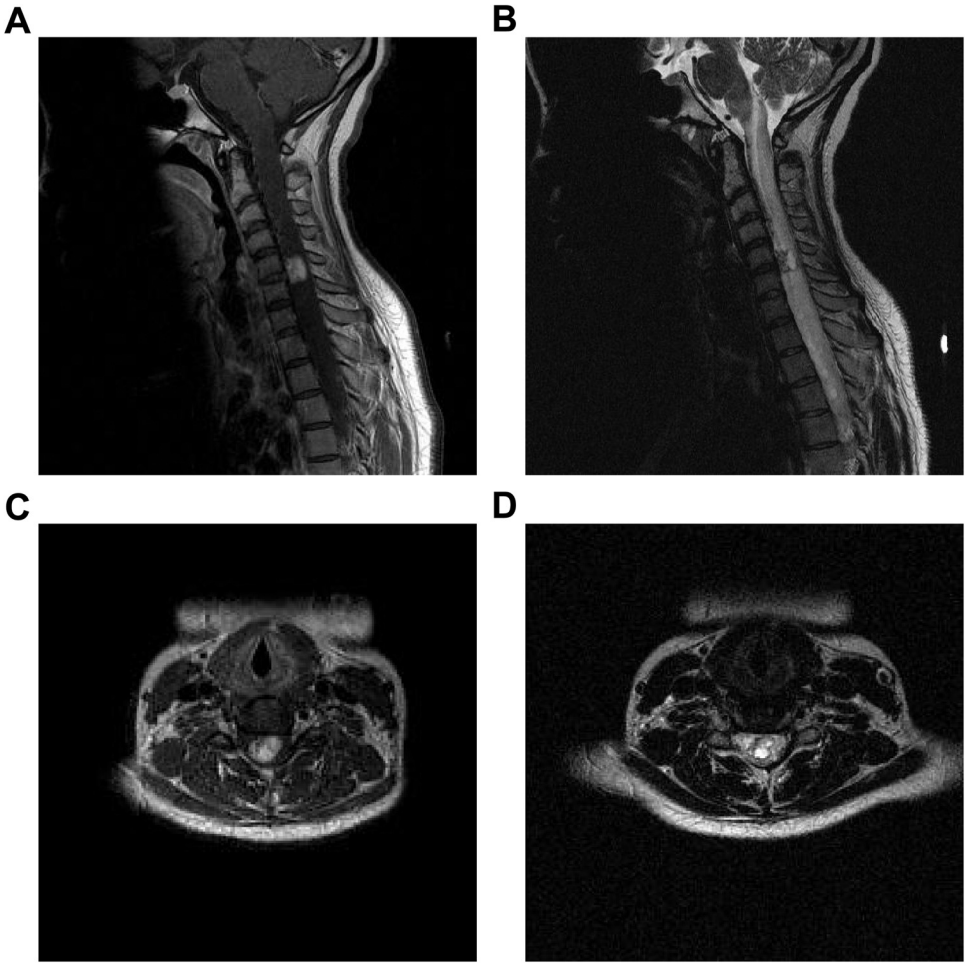

Fig. 1. Magnetic resonance imaging of the cervical cord of the hemangioblastoma upon admission. A 1.3 cm lesion is visualized in the posterior C5–C6 disc space, containing solid and cystic components with possible products of hemorrhage. An enlarged spinal cord with syrinx is visualized that tracks inferiorly 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_2 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_2 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

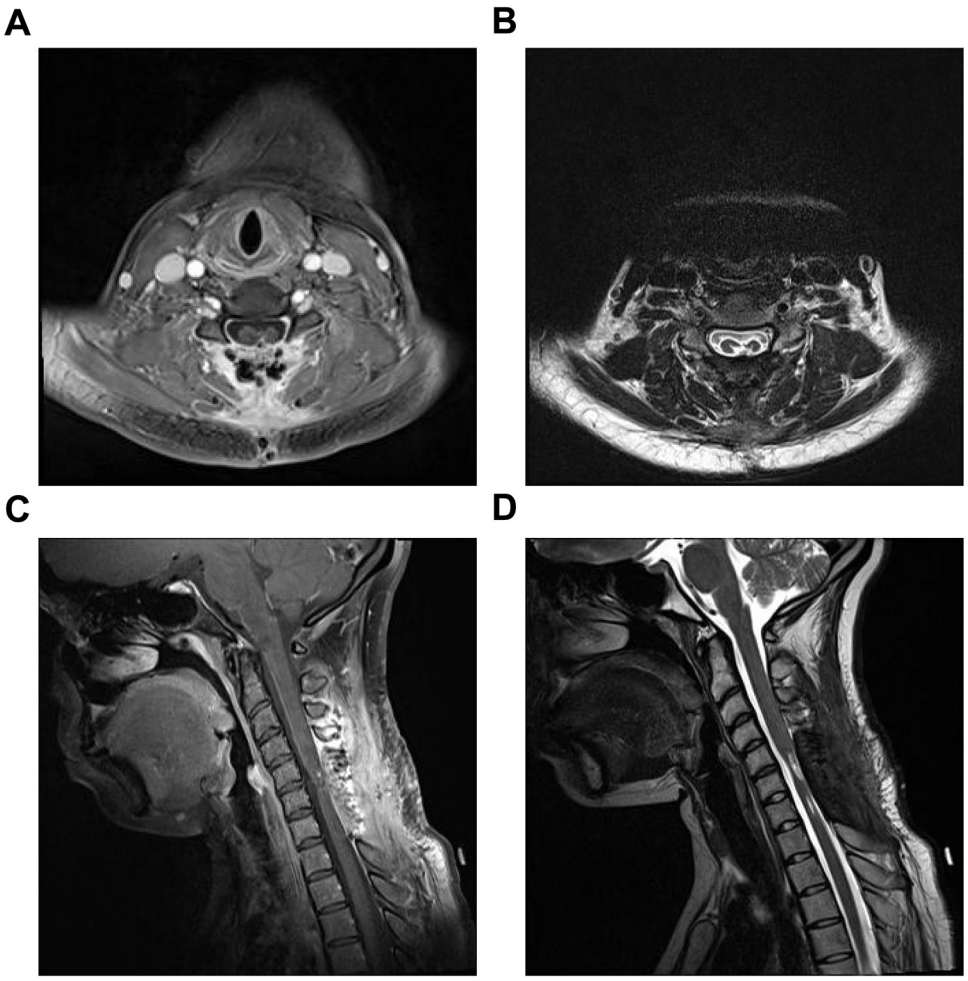

Fig. 2. Magnetic resonance imaging of the cervical cord of the hemangioblastoma, three months after surgery. Following imaging shows stable post-operative changes and resection of the C5–C6 mass, with a small 2 mm focus of enhancement in the posterior cord, unlikely to be a tumor recurrence and likely a post-operative artifact. (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.

oncology, who did not recommend adjuvant therapy, especially considering the slow growing nature of hemangioblastomas. A follow up magnetic resonance imaging of the cervical spine three months after surgery demonstrated stable post-operative changes without any features of residual or recurrent tumor growth (Fig. 2).

3. Discussion

Hemangioblastomas are a rare entity overall with the most common location for an occurrence of hemangioblastoma is in the posterior cranial fossa [10]. The second most frequent location is the spinal cord [11]. However, spinal cord hemangioblastomas only account for a small percentage of primary spinal cord tumors overall. Moreover, most descriptions of spinal cord hemangioblastomas are discussed in the context of a patient with Von-Hippel Lindau disease [12]. It is unknown how many isolated or sporadic spinal cord hemangioblastomas have occurred [13–19]. One study showed that spinal hemangioblastomas are sporadic in only approximately 20 percent of cases [20]. Patients with sporadic hemangioblastomas present much later in life, have less neurological symptoms, and have a better prognosis than their Von-Hippel Lindau counterparts [20,21]. Patients with Von-Hippel Lindau are at a higher risk of recurrence as well, requiring lifelong follow up and surveillance [21].

Current National Comprehensive Cancer Network guidelines recommend that patients who present with a primary spinal cord tumor undergo observation if the patient is asymptomatic but surgery if the patient is symptomatic. The rationale behind surgery is two-fold. First, patients who have primary spinal cord tumors and undergo surgical resection have acceptable rates of neurological improvements [22]. Second, surgery allows for the direct acquisition of tissue to confirm the diagnosis of hemangioblastoma. Imaging alone makes it difficult to ascertain neoplastic versus non-neoplastic conditions, requiring the reading radiologist to discern among different magnetic resonance signals [23]. Moreover, immunostaining can distinguish between metastatic renal cell carcinoma versus hemangioblastoma, especially in a patient with Von-Hippel Lindau disease [24,25]. Radiation therapy has also been proposed as a treatment modality, with a great deal of success [26–29]. However, we argue against radiation therapy since it would be difficult to treat a patient with certainty and curative intent without a definitive pathological diagnosis of the disease. Bevacizumab is a monoclonal antibody directed against vascular endothelial growth factor A that can block angiogenesis. It has been previously used in various oncological applications. One report described its use in a patient with a surgically unresectable cervical cord hemangioblastoma, showing significant tumor regression and clinical improvement [30].

4. Conclusions

Hemangioblastomas are a rare benign, slow growing vascular tumor of the central nervous system. We present a 22 year old patient who presented with bilateral upper extremity symptoms and had a sporadic hemangioblastoma of the spinal cord who was treated with surgery, not requiring any adjuvant therapy. The neurologic function improved in the days and months after surgery. We surmise that our experience is concordant with the few experiences reported in the literature and recommend expedited surgical and operative intervention for the treatment of these patients.

Consent

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

Conflicts of interest

The authors declare no conflicts of interest, relevant financial relationships, or any sources of support in the form of grants, equipment, or drugs.

Funding

No sources of funding are declared.

Ethical approval

Ethical approval has been given from the John Peter Smith Department of Neurosurgery.

Author contributions

Conception and design: Robert E. Germann, George F. Cravens; Acquisition of data: Robert E. Germann, George F. Cravens; Analysis and interpretation of the data: all authors; Drafting the article: Dat T. Vo; Critically revising the article: all authors; Approved the final version of the manuscript on behalf of all authors: Dat T. Vo; Study supervision: George F. Cravens.

Guarantor

All of the authors listed in the manuscript accept full responsibility for the work and conduct of this study.

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