

Case Report

Hemangiopericytoma invading the craniovertebral junction: First reported case and review of the literature

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Abstract

Occurrence of hemangiopericytoma (HPC) in the central nervous system is rare. Spinal HPCs with intramedullary involvement are even more unusual. We present a case of a craniovertebral intradural HPC with both extra- and intra-medullary extensions. Though the patient presented with vague cervical symptoms, imaging was indicative of an intradural lesion from the occiput to C4 and a second smaller, subclinical lesion, at the T2-3 level. He underwent gross total surgical resection of the craniovertebral lesion and did well post-operatively. The thoracic lesion was treated with radiosurgery and the patient is neurologically at baseline 5 years later. Gross total resection of HPCs is the recommended treatment when possible. Histopathology is crucial for diagnosis due to both its rarity and similar characteristics to other tumors on physical and radiographic evaluations. Recognizing that these uncommon tumors can occur with both extra-medullary and intra-medullary locations are important for diagnosis and treatment recommendations. Future studies using national surgical databases that contain histology will be needed to understand the long-term clinical outcomes.

Key words: Cervical, extramedullary, hemangiopericytoma, intradural, spinal

INTRODUCTION

In 1942, Stout and Murray described a vascular neoplasm originating from capillary pericytes and called it hemangiopericytoma (HPC).^[1] Later, in the early 1990s, The World Health Organization (WHO) would categorize this neoplasm as a “mesenchymal non-meningothelial” tumor (grade II and III).^[2] More recently, the updated WHO tumors of soft-tissue and bone, classified HPCs as fibroblastic without evidence of pericytic differentiation,^[2] signifying their similarity to solitary fibrous tumors.

Although more commonly found outside the central nervous system (CNS), CNS HPCs do occur and are usually intracranial. Tumors occurring in the spine are extremely rare and usually extradural. In terms of intradural cervical tumors, only 14 cases have been reported.^[3] Only two of these reports have documented intramedullary involvement. We present a rare case of a HPC originating from the craniovertebral junction with intradural and extra-medullary elements, spinal cord invasion and thoracic drop metastasis.

CASE REPORT

History

This study reports a case of a 56-year-old fairly healthy male patient presented complaining of neck pain that radiated into his head and both shoulders. His pain was present for several months and no longer relieved by anti-inflammatory medications.

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Examinations and findings

On neurological exam, he had intact motor and sensory responses bilaterally, with normal reflexes. Romberg, cerebellar and gait examinations were normal as well. The gadolinium-enhanced cervical spine magnetic resonance imaging (MRI) revealed an intradural extra-medullary lesion extending from the occiput to C4 in the ventrolateral position with central stenosis and cord compression [Figure 1a]. In addition, there was a small enhancing lesion seen at the upper thoracic spine [Figure 1a]. Both neural foramina were normal. MRIs of the brain and lumbar spine were both within normal limits.

Operation

A suboccipital craniectomy and C1-4 complete laminectomy were performed. With the aid of the microscope, a midline dural incision was made to expose a solid, red tumor in the dorsolateral right gutter, displacing the spinal cord to the left and bordered by clear arachnoid.

The tumor was removed in several large fragments and debulked away from a number of nerves in its proximity with a Cavitron. It was found to be densely adherent to the spinal cord at the C1 level around dentate ligaments, extending ventrolaterally. Multiple lobules of tumor were removed in a clear arachnoid plane except where there was adherence to the spinal cord at the C1 level. There was pial invasion on one side, which with cauterization of the pia, the residual tumor was removed and a gross total excision was attained. Careful hemostasis and closure were achieved without issues.

Pathology

Grossly, the tumor was a large hemorrhagic mass. The pathologic specimens were designated as intradural extra-medullary and partially invasive intradural intramedullary at

the C1 level. On microscopy, the tumor cells had widened nuclei with spotted chromatin and mitotic figures (up to 30 per 10 high-power fields). There were foci of hemorrhage, but no areas of necrosis. There was minimal reticulin deposition. Immunostaining revealed the tumor was positive for CD34 and B-cell lymphoma 2, but negative for smooth muscle actin, epithelial membrane antigen, synaptophysin and glial fibrillary acidic protein and a Ki-67 proliferative index of 12% [Figure 1]. On electron microscopy, the tumor was highly vascular and contained numerous extravasated erythrocytes and other cellular blood elements. Final histology was a tumor borderline grade between HPC WHO grade II based on morphology and anaplastic HPC WHO grade III based on elevated mitotic count and intratumoral hemorrhage.

Post-operative course

The post-operative course was uneventful and there was complete resection of the tumor as evidenced on MRI. Initially, the patient experienced mild right deltoid weakness without tingling, numbness, headaches or other neurologic symptoms. A post-operative MRI at 6-months revealed the remaining enhancing drop metastasis at the T2-3 level. At an outside facility, the patient underwent successful radiosurgery of the area. His reported right periscapular shoulder girdle atrophy has been responding to physical therapy and other treatments. Five years post-operatively, he is neurologically at baseline and follow-up MRI [Figure 1] reveals no evidence of recurrence in either the cervical or thoracic regions.

DISCUSSION

HPCs are rare, locally-recurring, aggressive malignant tumors with metastatic potential. Although they have been reported throughout the spinal cord, the cervicothoracic region has the greatest incidence.^[3] Intradural tumors are even rarer and have been less commonly associated with pain than extradural tumors. In the review of prior cervical HPCs in the literature, none of the 14 reported cases had pain complaints involving intradural lesions, though our patient presented with a pain complaint. Dural-based lesions are often well-circumscribed and sharply enhance, which may confound the diagnosis.^[4,5] Radiographically, most findings are nonspecific and show similar characteristics as other intracranial or intraspinal tumors such as schwannomas, meningiomas, neurofibromas and sometimes as ependymomas, lymphomas and sarcoidoses.^[3-5] Lesions are generally hyperintense on computed tomography with homogeneous contrast enhancement and isodense on T1- and T2-weighted MRI images with heterogeneous enhancement and commonly contain flow voids.^[6,7]

Surgical treatment with gross total resection (GTR) has been reported in the literature to provide the best prognosis and overall survival.^[4,8] Our patient has done well during the 5 years of follow-up after GTR. Our case was unique anatomically in that from C2-C4 it was intradural extra-medullary, but at C1 it was felt to be intradural intramedullary. Tumors with

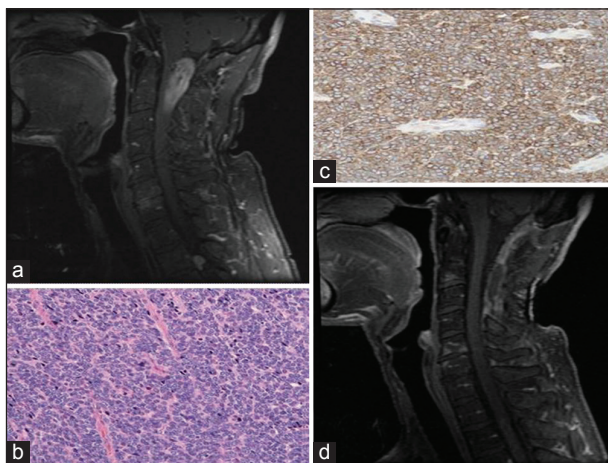


Figure 1: Pre-operative T1 magnetic resonance imaging (MRI) with gadolinium sagittal (a) MRI showing an enhancing lesion at the occiput to C4 levels with severe spinal cord compression and a small enhancing lesion seen at the upper thoracic spine (T2-3). H and E staining (b) with dense cellularity and oval shaped nuclei forming fascicles. CD34 staining (c) demonstrating the classic staghorn vascular channel, original magnification $\times 200$. 5-year post-operative sagittal T1 MRI with gadolinium (d) showing no evidence of cervical or thoracic disease

intramedullary involvement reportedly have a recurrence rate of 0% and an 3-year overall survival of 100%.^[3] In addition, intradural tumors seem to have better prospects than extradural tumors. For instance, Betchen *et al.* reported that extradural tumors recurred earlier than intradural tumors (2.6 years vs. 6 years, respectively).^[4] Nonetheless, bigger studies with long-term follow-up are required before significant conclusions can be made. Radiotherapy has been studied with inconclusive results and mostly for intracranial tumors.^[8] In terms of metastasis, it occurs in about 29% of patients and timing of recurrence can vary widely from 2 to 18 years after initial treatment, which demonstrates the need for frequent follow-up. Our patient had a drop metastasis in the thoracic area. At 6-months post-operatively from the cervical surgery, he underwent successful radiosurgery without recurrence 5 years later.

Currently, the advised treatment for spinal HPCs is en bloc resection of the tumor with its surrounding dura. When this is not possible, HPC tumors should undergo embolization followed by debulking. Based on the available research, radiotherapy is not recommended following GTR except for cases involving subtotal resection and recurrences where repeat surgery is contraindicated.^[3] No therapy has been shown to be effective in preventing metastasis, thus far.

Future studies using national surgical databases that contain histological grades for HPC spinal cases will likely elucidate the expected long-term clinical outcomes.

CONCLUSIONS

Diagnosis of HPCs is challenging due to its rarity and indiscriminant findings on neurological examination and imaging. Our case emphasizes the exceptionally infrequent occurrence of HPC at intradural extra-medullary and

intramedullary sites and the importance of histopathology for diagnosis. Surgical GTR is the standard of care with long-term follow-up to monitor for drop metastases. In conclusion, recognizing that these rare tumors can occur intradural extra-medullary and intra-medullary will aid in accurate diagnosis and treatment of HPC.

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