Cervical Spine Ependymoma with Hematomyelia: Case Report and Review of the Literature

Hai Le¹, Rishi Wadhwa², Susan Le¹, Jennifer Cotter², Han Lee², Praveen Mummaneni¹ and Michael McDermott¹

¹Department of Neurosurgery, University of California, San Francisco, California, USA
²Department of Pathology, University of California, San Francisco, California, USA

Corresponding author: Hai Le, Department of Neurosurgery, University of California, San Francisco, California, USA, Tel: 415-353-7500; E-mail: Hai.Le@ucsf.edu

Abstract

Ependymomas are primary CNS tumors representing 3%-6% of all CNS tumors, and 34.5% of ependymomas occur in the spine. Spinal ependymomas occur most frequently in the cervical spine. Rarely, tumor-associated syringomyelia and hematomyelia may complicate cervical spinal ependymomas. Here, the authors present a case of a 37 year-old gentleman with cervical intramedullary WHO Grade II ependymoma with hematomyelia extending cephalad to the brainstem. The authors also detail their operative procedure using the OmniGuide CO₂ laser and review current literature on the management of cervical intramedullary ependymoma with tumor-associated syringomyelia and/or hematomyelia.

Keywords: Ependymoma; Syringomyelia; Hematomyelia; Intramedullary spinal tumor; CO₂ Laser; Omniguide

Abbreviations:

CNS: Central Nervous System; WHO: World Health Organization;
GTR: Gross Total Resection

Introduction

Ependymomas are primary CNS tumors representing 3%-6% of all CNS tumors; and 30-35% of ependymomas occur in the spine. Spinal ependymomas occur most frequently in the cervical spine [1-3].

Like many cervical spine lesions; intramedullary ependymoma of the cervical spine can compress the spinal cord and cause symptoms of cervical myelopathy. Infrequently; cervical spine ependymoma can coexist with syringomyelia. There have been several proposed pathomechanisms for the development of tumor-related syringomyelia. Syringomyelia may be caused by intramedullary ependymoma spreading cephalad and compressing on the foramina of Luschka. Another possible explanation is that tumor growth may cause increased intramedullary pressure and microcirculation impairment; thus weakening normal tissue architecture and leading to the development of degenerative cysts and syringomyelia [4]. Tumor necrosis and hemorrhage into the syrinx can cause hematomyelia [4-6].

To the best of our knowledge; there have only been two reported cases of primary cervical intramedullary ependymoma with hematomyelia. Kawakami and Mair first reported of a 68 year-old man on anticoagulation therapy for pulmonary embolism who was evaluated for cervical myelopathy. Workup showed a C6 tumor with hematomyelia from C3-T10; and histology confirmed the diagnosis of intramedullary ependymoma with neurilemmomas. The patient underwent cervical laminectomy without symptomatic relief; and he expired three weeks after admission [5]. Sato et al. reported of a 58 year-old man who presented with bilateral upper extremity motor and sensory disturbances. He was diagnosed with spinal tanyctic ependymoma at C2-C4 with hematomyelia extending caudally. The patient responded well to C2-4 laminectomy with subtotal resection of the tumor and evacuation of the hematoma [6].

Here, the authors present a case of a 37 year-old gentleman with cervical intramedullary WHO Grade II ependymoma with tumor-associated hematomyelia. Compared to the above-mentioned two cases; our patient was much younger; and the hematomyelia extended cephalad to the brainstem rather than caudally. We also detail our operative procedure using the OmniGuide CO₂ laser and review current literature on the management of cervical intramedullary ependymoma with tumor-associated syringomyelia and/or hematomyelia.

Case Report

History

The patient was a 37 year-old Chinese male who presented on July 28, 2013 with progressively worsening left arm and left leg numbness; nausea; non-positional headaches and hiccups. He had initial workup at an outside hospital for non-radiating back pain exacerbated by heavy lifting in 2011; effectively managed with physical therapy. His back pain returned in June 2012; when MRI revealed a C1-T7 syrinx with features concerning for an underlying lesion. He did not pursue further workup at the time as he was asymptomatic except for intermittent back pain. The patient began developing progressively worsening left-sided numbness in February 2013; described as a loss of sensation to touch but not to pain. These symptoms had been accompanied by continual hiccupping; urinary frequency and two episodes of syncope. Past medical history was significant for two episodes of head trauma with loss of consciousness (in 2010 and 2012) from strikes to the head in karate. He denied any history of spinal cord trauma; and he was not on anticoagulation.
Examination

On physical exam; the patient was afebrile with normal vital signs. Cranial nerves were intact. He had normal muscle bulk; tone and strength throughout. He had decreased sensation to light touch in his LUE and LLE and increased sensation to pain in his LLE. Proprioception was intact. Reflexes were slightly diminished (1+) in biceps; triceps and Achilles bilaterally. Hoffmann and Babinski signs were negative. Rectal tone was normal. There was no tenderness to palpation over the spinous processes.

Imaging

Preoperative CT without contrast showed a round fluid attenuating mass in the foramen magnum at the inferior portion of the fourth ventricle causing expansion of the central spinal canal measuring approximately 1.4 cm in the transverse dimension (Figure 1). Preoperative MRI showed a multiloculated syrinx with multiple blood fluid levels extending from the obex to T5-T6; with intrinsic T1 hyperintensity at the inferior aspect suggestive of blood products. An enhancing 0.8 x 1.3 x 1.5 cm (AP by transverse by craniocaudal) mass was present centrally within the cervical spinal cord at C6-C7. Edema and blood products continued inferiorly within the cord to the level of T7 (Figure 2).

Figure 1: Preoperative axial unenhanced CT showing a fluid attenuating mass in the foramen magnum at the inferior portion of the fourth ventricle; suggestive of a syrinx.

Figure 2: Preoperative sagittal T2-weighted MRI of the cervical (A) and thoracic (B) spine showing a multiloculated large syrinx with the cavitary portion extending from the level of the obex to T5-T6 with an enhancing central mass at the C6-C7 level. Preoperative sagittal T1-weighted MRI (C) showing intrinsic T1 hyperintensity at the inferior aspect of the cavitary portion of the syrinx (T5-T6 level); likely secondary to blood products.

Operation

Given the patient’s clinical presentation and imaging; he was diagnosed with cervical myelopathy secondary to a C6-7 intramedullary spinal cord tumor with hematomyelia extending cephalad to the brainstem. The patient was taken to the operating room the next day for open C6-C7 laminectomy and microsurgical resection of the tumor. After steroids were administered; he was immobilized with Mayfield cranial fixture and positioned prone. The surgical field was prepped in a sterile fashion; and a linear skin incision from C5-T1 was made. Dissection down to the C6 and C7 laminae were carried out. Laminectomy troughs were created bilaterally using a high speed burr; and the laminae were elevated for laminectomy. Under the operating microscope; dorsal pial myelotomy was performed using the OmniGuide CO\textsubscript{2} laser; with the power setting at 5 watts and pulse setting at 200 milliseconds. The pia was tacked back to the dura using 6-0 Prolene suture. Dissection and exposure were carried out along the dorsal midline between the dorsal columns with the Rhoton #6 and #7 microdissectors. The tumor was found at the base of this plane. It was surrounded by a large syrinx containing old blood; which was thoroughly irrigated and evacuated. The tumor was carefully excised using Rhoton micro-instruments; Cavitron Ultrasonic Surgical Aspirator (CUSA) and bipolar. We first internally debulked the tumor with the CUSA. We then dissected the lateral margins using microdissectors and the CO\textsubscript{2} laser to take down the cord tumor interface. Finally; working along the ventral surface of the tumor; we divided the last of the vascular pedicles and removed the tumor completely. Throughout the case; motor evoked potentials (MEPs) confirmed that the spinal cord was functional with no changes from baseline. A gross total resection of the tumor was achieved. Frozen section returned as ependymoma. After hemostasis was achieved with bipolar; the dura was closed with running 6-0 Prolene suture. Valsalva maneuver did not show any CSF leak. The dura was covered with Tisseel. A JP drain was placed; and the skin was closed with running nylon suture. Clonidine; fentanyl and vancomycin powder were instilled subfascially. There were no intraoperative complications; and EBL was <200 mL.
Postoperative Course

Postoperative cervical MRI confirmed gross total resection of the solid enhancing intramedullary tumor at C6-C7 with interval decrease in size of the cervicothoracic syrinx (Figure 3). Dynamic x-rays showed expected postoperative changes and good alignment without evidence of dynamic instability during flexion and extension (Figure 4). Surgical pathology supported the diagnosis of intramedullary spinal cord ependymoma; WHO Grade II (Figure 5). His hospital course was insignificant; and he was discharged on postoperative day 4 in good condition. Follow-up visit at one month showed good recovery. Patient will need follow-up imaging surveillance but no adjuvant chemoradiation therapy at this time.

Discussion

Overview of cervical intramedullary spinal ependymomas

Intramedullary spinal ependymomas comprise only 15% of all spinal cord tumors; but they make up 60% of all intramedullary spinal cord tumors in adults [7,8]. In a comprehensive literature review; Oh et al. (2013) reported that spinal ependymomas occurred most frequently in the cervical spine (32.0%); followed by the conus plus cauda equina (26.8%); thoracic (16.3%); cervicothoracic (16.3%); thoracolumbar (5.1%); and cervicomedullary region (3.4%) [1]. Our patient had an intramedullary spinal cord tumor that involved the C6-7 levels.

Spinal ependymomas in the upper spinal regions (cervicomedullary; cervical; cervicothoracic) had significantly longer progression free survival compared with ependymomas in the lower spinal regions (thoracic; thoracolumbar; conus plus cauda equina) (p<0.001) [1].

Intratumoral hemorrhage is present (D; solid arrow); as are hemosiderin-laden macrophages (D; dotted arrow). These are nonspecific indicators of degenerative change; but in this case; may correlate with the clinical presentation of hematomyelia.

Citation: Le H, Wadhwa R, Le S, Cotter J, Lee HS, et al. (2014) Cervical Spine Ependymoma with Hematomyelia: Case Report and Review of the Literature. J Spine 3: 171. doi:10.4172/2165-7939.1000171
Operative management of cervical intramedullary spinal ependyomas

The underlying principle in the treatment of intramedullary spinal cord ependyomas is to achieve gross total resection (GTR). Surgical resection; usually through a posterior approach; is the treatment of choice for these lesions and should be performed as early as possible once the diagnosis is made or suspected. With GTR; the tumor recurrence rate is fortunately exceedingly low [1,10-14]. Radiation therapy or chemotherapy is considered surgical adjunct and is usually recommended in patients for whom gross total resection is not achieved [15]. Complications from spinal cord ependymoma surgery are not uncommon and include sensorimotor loss; dorsal column dysfunction; and bowel and bladder dysfunction [16].

The use of the OmniGuide CO₂ laser in spinal tumor resection

The OmniGuide CO₂ laser system has had good success in the fields of gynecology and otolaryngology. CO₂ laser technology for use in neurological surgery has been available for the past five decades but only recently gained popularity with the introduction of the flexible hollow core fiber system by OmniGuide that allows for greater surgical precision and is usually recommended in patients for whom gross total resection is not achieved [15]. Complications from spinal cord ependymoma surgery are not uncommon and include sensorimotor loss; dorsal column dysfunction; and bowel and bladder dysfunction [16].

In summary; syringomyelia and/or hematomyelia are rare complications associated with cervical intramedullary spinal ependyomas. The treatment of choice for these lesions is gross total resection with surgical evacuation of the fluid collection. The OmniGuide CO₂ laser offers great surgical precision with reduced tissue manipulation in tumor resection and should be more frequently utilized in spinal tumor surgery.

References

1. Oh MC, Kim JM, Kaur G, Safaee M, Sun MZ, et al. (2013) Prognosis by tumor location in adults with spinal ependyomas. J Neurosurg Spine 18: 226-235.
2. Gilbert MR, Ruda R, Soffietti R (2010) Ependymomas in adults. Curr Neurol Neurosci Rep 10: 240-247.
3. Chamberlain MC (2003) Ependymomas. Curr Neurol Neurosci Rep 3: 193-199.
4. Szpak GM, Lewandowska E, Schmidt-Sidor B, Pasennik E, Modzelewska J, et al. (2008) Giant cell ependymoma of the spinal cord and fourth ventricle coexisting with syringomyelia. Folia Neuropathol 46: 220-231.
5. Kawakami Y, Mair WG (1973) Haematomyelia associated with anticoagulant therapy, an intramedullary ependymoma and Schwann cells. Acta Neuropathol 26: 253-258.
6. Sato K, Kubota T, Ishida M, Handa Y (2005) Spinal tanyctyic ependymoma with hematomyelia—case report—. Neurol Med Chir (Tokyo) 45: 168-171.
7. Chang UK, Choe WJ, Chung SK, Chung CK, Kim HJ (2002) Surgical outcome and prognostic factors of spinal intramedullary ependyomas in adults. J Neurooncol 57: 133-139.
8. Balériaux DL (1999) Spinal cord tumors. Eur Radiol 9: 1252-1258.
9. Louis DN, Ohgaki H, Wiestler OD, Cavenee WK, Burger PC, et al. (2007) The 2007 WHO classification of tumours of the central nervous system. Acta Neuropathol 114: 97-109.
10. Epstein FJ, Farmer JP, Freed D (1993) Adult intramedullary spinal cord ependymomas: the result of surgery in 38 patients. J Neurosurg 79: 204-209.
11. Lee TT, Gromelski EB, Green BA (1998) Surgical treatment of spinal ependymoma and post-operative radiotherapy. Acta Neurochir (Wien) 140: 309-313.
12. Hoshimaru M, Koyama T, Hashimoto N, Kikuchi H (1999) Results of microsurgical treatment for intramedullary spinal cord ependymomas: analysis of 36 cases. Neurosurgery 44: 264-269.
13. Klekamp J (2013) Treatment of intramedullary tumors: analysis of surgical morbidity and long-term results. J Neurosurg Spine 19: 12-26.
14. Oh MC, Tarapore PE, Kim JM, Sun MZ, Safaee M, et al. (2013) Spinal ependymomas: benefits of extent of resection for different histological grades. J Clin Neurosci 20: 1390-1397.
15. Lee TT, Gromelski EB, Green BA (1998) Surgical treatment of spinal ependymoma and postoperative radiotherapy. Acta Neurochir (Wien) 140: 309-313.
16. Nagasawa DT, Smith ZA, Cremer N, Fong C, Lu DC, et al. (2011) Complications associated with the treatment for spinal ependymomas. Neurosurg Focus 31: E13.
17. Ryan RW, Wolf T, Spetzler RF, Coons SW, Fink Y, et al. (2010) Application of a flexible CO2 laser fiber for neurosurgery: laser-tissue interactions. J Neurosurg 112: 434-443.
18. Kim JS, Lee SH (2009) Carbon dioxide (CO2) laser-assisted ablation of lumbar discal cyst. Photomed Laser Surg 27: 837-842.
19. Ahn Y, Moon KS, Kang BU, Hur SM, Kim JD (2012) Laser-assisted posterior cervical foraminotomy and discectomy for lateral and foraminal cervical disc herniation. Photomed Laser Surg 30: 510-515.
20. Kim JS, Oh HS, Lee SH (2012) Usefulness of carbon dioxide laser for recurrent lumbar disc herniation. Photomed Laser Surg 30: 568-572.
21. Desai SK, Paulson D, Thompson BJ, Patterson J, Mohanty A (2012) The role of flexible hollow core carbon dioxide lasers in resection of lumbar intraspinal lipomas. Childs Nerv Syst 28: 1785-1790.