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Case Report Open Access

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

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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 CO_2 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; CO_2 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 tanycytic 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 CO_2 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).

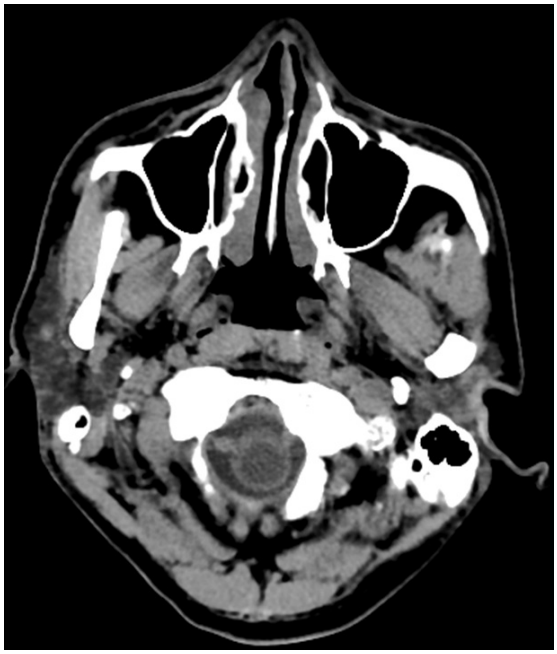

Figure 1: Preoperative axial unenhanced CT showing a fluid attenuating mass in the foramen magnum at the inferior portion of the fourth ventricle; suggestive of a syrinx.

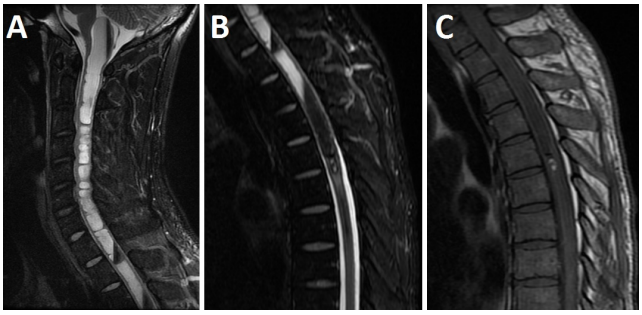

Figure 2: Preoperative sagittal T2-weighted MRI of the cervical (A) and thoracic (B) spine showing a multiloculated large syrinx with the cavitary portion extending from the level of the obex to T5-T6 with an enhancing central mass at the C6-C7 level. Preoperative sagittal T1-weighted MRI (C) showing 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_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_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 locked 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 synrix (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.

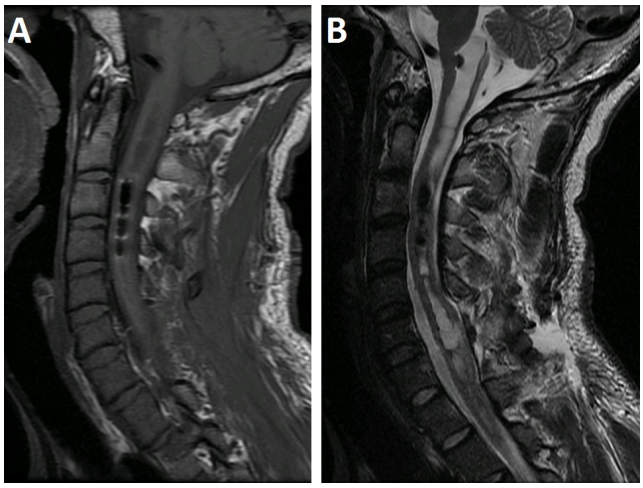

Figure 3: Postoperative T1-weighted (A) and T2-weighted (B) sagittal MRIs showing gross total resection of the ependymoma with interval decrease in size of the cervicothoracic synrinx.

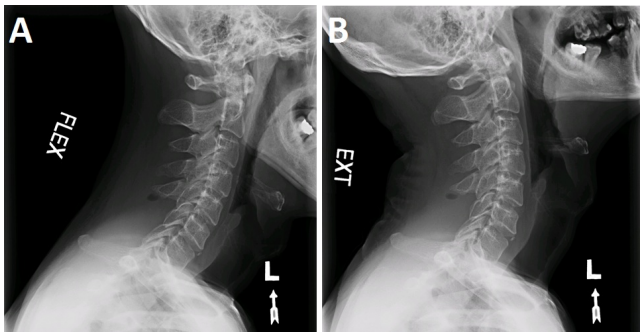

Figure 4: Postoperative dynamic x-rays showing expected post-laminectomy changes at C6-7 and good alignment without evidence of dynamic instability during flexion and extension.

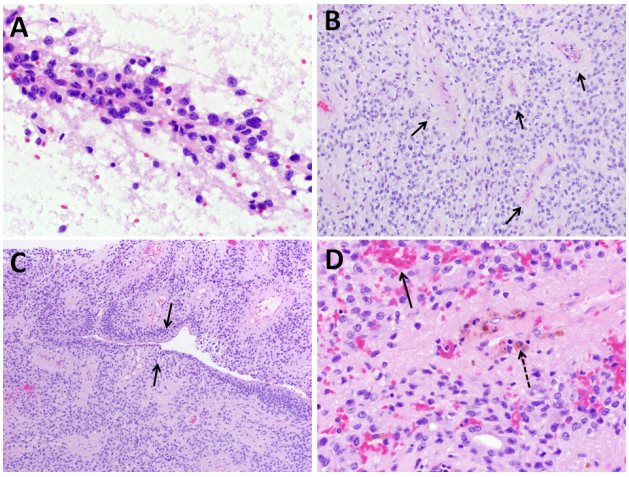

Figure 5: Smear preparation (A) shows a relatively bland population of tumor cells with elongated fibrillary processes and oval nuclei with coarse chromatin. Classic features of WHO grade II ependymoma include perivascular pseudorosettes with anuclear zones surrounding small vessels (B) and true ependymal canals; in which columnar cells line an open lumen (C). 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.

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 are histologically classified by the WHO classification into three different grades. Grade I consists of myxopapillary ependymomas and subependymomas; Grade II consists of classic ependymomas; and Grade III consists of anaplastic ependymomas [1,9]. Greater than 75% of ependymomas occurring in the cervical or thoracic regions were WHO Grade II [1]; which was the tumor grade observed in our patient. Classic features of WHO grade II ependymoma include perivascular pseudorosettes with anuclear zones surrounding small vessels and true ependymal canals; in which columnar cells line an open lumen (Figure 5).

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].

Operative management of cervical intramedullary spinal ependymomas

The underlying principle in the treatment of intramedullary spinal cord ependymomas 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 CO_2 laser in spinal tumor resection

The OmniGuide CO_2 laser system has had good success in the fields of gynecology and otolaryngology. CO_2 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 dexterity and precision (Omniguide Inc.; Cambridge; MA) [17]. Kim and Lee applied this technology to ablate lumbar discal cyst in 14 patients and reported that the back VAS score improved by 2.4 and leg VAS score improved by 6.4 ($p<0.001$). The ODI score improved from 46.47% to 14.81% ($p<0.001$) [18]. Ahn et al. used CO_2 laser to perform posterior cervical foraminotomy and discectomy in 24 patients and observed that the arm VAS score improved by 5.59 while the NDI score improved from 47% to 10.46%; both outcomes reaching statistical significance [19]. In a retrospective study by Kim et al.; CO_2 laser also provided statistically significant improvements in leg VAS score and ODI score in 21 patients undergoing revision microdiscectomy for recurrent lumbar disc herniation [20]. The outcomes of these studies suggest that the OmniGuide CO_2 laser could be more routinely use to provide neurological spine surgeons greater precision with less tissue manipulation.

The aforementioned studies were performed for non-tumorous pathology of the spine. There have not been many studies evaluating the application of CO_2 laser in patients with spinal tumors. Desai et al. used the CO_2 technology to resect 8 lumbar intraspinal lipomas with subtotal to near-total resection in all cases. None of the patients developed any new sensorimotor deficits; but 3 patients did have postoperative CSF leakage [21]. At our institution; we have been utilizing the OmniGuide CO_2 laser to facilitate resection of spinal tumors. Our clinical/surgical outcomes are comparable or better in cervical spinal cord tumor resection using this CO_2 laser technology compared to resection using conventional surgical instruments.

Conclusion

In summary; syringomyelia and/or hematomyelia are rare complications associated with cervical intramedullary spinal ependymomas. The treatment of choice for these lesions is gross total resection with surgical evacuation of the fluid collection. The OmniGuide CO_2 laser offers great surgical precision with reduced tissue manipulation in tumor resection and should be more frequently utilized in spinal tumor surgery.

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