

Case Report

Thoraco-lumbar high grade spinal cord astrocytoma: first documented case in a tertiary medical center and review of literature

John Patrick R. Marquez*, Niccolo S. Mamba

Department of Orthopedics, Cagayan Valley Medical Center, Cagayan, Philippines

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*Correspondence:

Dr. John Patrick R. Marquez,

E-mail: johnpatrickmarquez14@yahoo.com

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ABSTRACT

This presents the first documented case of high grade spinal cord astrocytoma in a pediatric patient treated at a tertiary medical center and gives a comprehensive review of literature on the most current modes of diagnosis, treatment, and prognosis. We report the case of a 7-year-old male with complaints of inability to ambulate and associated low back pain, which rapidly deteriorated to complete paraplegia. Magnetic resonance imaging (MRI) of his spine revealed an intradural intramedullary spinal cord lesion extending from T11 to L5 vertebra, with considerations of spinal astrocytoma and ependymoma. The patient underwent en bloc laminectomy on T9-L3, partial resection of the intramedullary cord tumor, and subsequent laminoplasty with mini plates and screws. Histopathologic studies showed spinal cord astrocytoma, World Health Organization (WHO) grade 3 (anaplastic type). The patient's neurologic status did not improve after the surgery and was advised to undergo chemo-radiation therapy but was lost to follow up. Ten months post-operation, patient sought consult due to severe back pain, generalized body weakness, and difficulty of breathing and was then admitted. Patient's condition worsened that eventually resulted to his demise.

Keywords: Spinal cord astrocytoma, Anaplastic spinal astrocytoma, Spinal cord tumor, Intramedullary astrocytoma, En bloc laminectomy, Thoraco-lumbar astrocytoma

INTRODUCTION

Primary spinal cord tumor is rare with an incidence of 0.74 per 100,000 persons per year. Among these reported cases, only 0.03 accounted for benign and malignant glial histologic features in international literature.¹ In the Philippines, there has been no reported cases of primary malignant spinal cord astrocytoma in the thoraco-lumbar region in the local literature.

Here we present a case report of high-grade intramedullary spinal cord astrocytoma in a pediatric patient.

CASE REPORT

This is the case of a 7-year old male who had bilateral lower extremity weakness accompanied by severe low

back pain. History revealed deterioration of his ambulatory status with bladder and bowel incontinence for a year, which rapidly declined a week prior to admission. There was no history of significant trauma nor comorbidities.

On physical examination, the patient did not show signs of distress. There were no obvious deformities in his limbs and spine and no impairment in his cognitive functions. Cranial neurologic examination was normal. Spinal neurologic examination showed impaired sensory function below xyphoid area bilaterally. Motor function of his upper extremities was normal while that of his lower extremities was graded 2/5 (MMT) bilaterally. There were no appreciable deep and pathologic reflexes.

Magnetic resonance imaging (MRI) of the thoracic and lumbar spine revealed an intradural intramedullary spinal

cord lesion extending from the T11 to L5 vertebra with multiple intratumoral syrinx formation (Figure 1).

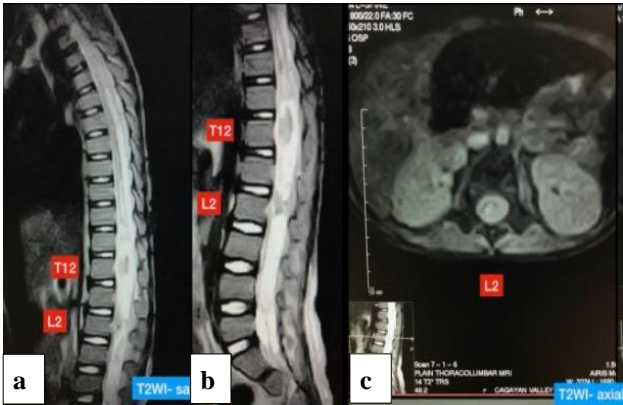


Figure 1: Sagittal sections T1WI of (a) thoracic and (b) lumbar spine, and (c) axial T1WI at L2 level showing enlarged and elongated spinal cord with an intradural intramedullary spinal cord tumor with multiple intra and peritumoral syrinx formation.

The patient was started on initial intravenous dexamethasone at 4 mg once a day which was eventually increased to a maximum of 4 mg every six hours. There was rapid deterioration of his spinal neurologic function and he eventually became completely paraplegic within a few days of admission despite high doses of intravenous steroids.

We did a gross total resection of the intramedullary spinal cord lesion by removing the solid components at the T12 and L2 vertebral level, while leaving the syrinx to spontaneously resolve. On a prone position, the posterior bony elements were exposed from T10 to L4. Using a high-speed burr-drill, a laminoplastic laminotomy was created at the junction of the lamina and facets, taking care not to damage the pars interarticularis (Figure 2a). This was done from T11 to L3 and the laminae of these vertebrae was removed en bloc (Figure 2b). The en bloc laminae was wrapped in moist gauze to preserve viability.

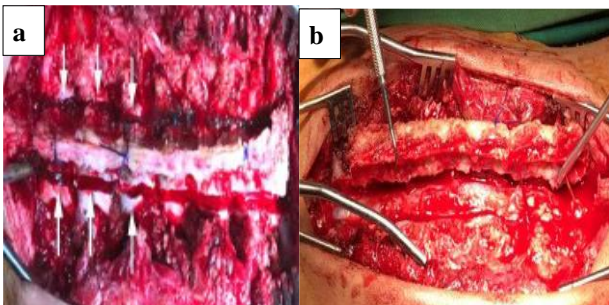


Figure 2: (a) Intra-operative photo showing bilateral laminoplastic laminotomy done on T11 to L3 vertebrae (white arrows) and (b) Lamina of T11 to L3 removed en bloc.

Under operating microscope view, the dura was opened and retained using tacking sutures (Figure 3).

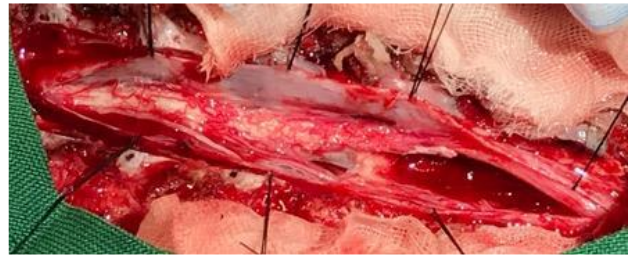


Figure 3: Dura opened and retained using tack sutures. Arachnoid membrane opened with spinal cord exposed.

The arachnoid membrane was opened. The spinal cord was inspected and work was initially focused on the L2 vertebrae level. It was noted that the cord at this level had an exophytic out pouching at its lateral aspect and had a sinus tract formation (Figure 4a and b). The pia membrane was opened using the sinus tract as a starting point. Pial tack sutures were put to expose the cord within. The tumor lesion was distinguished by having a brown-red color as opposed to the normal white colored spinal cord tissue (Figure 4c). It was easily friable and did not have a clear cleavage plane from the normal cord tissue and had a dispersed distribution as opposed to a single solid mass. After removing much of the tumor at this level, the lesion was judged to be malignant and no further dissection was done at T11 vertebral level. The dura was closed in a water tight fashion, and the En bloc laminae was fixed using titanium mini plates and screws (Figure 5).

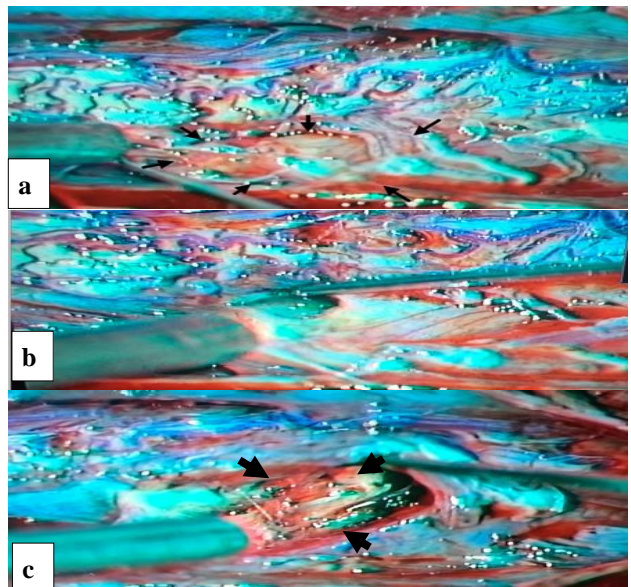


Figure 4: (a) Spinal cord showing outpouching mass (arrows) on its lateral aspect, (b) sinus formation located at junction of outpouching mass and normal cord (arrow), and (c) intramedullary tumor mass (arrows) having red-brown color compared to normal white colored cord tissue.



Figure 5: En bloc laminae of T11 to L3 reconstructed using titanium mini plates and screws.

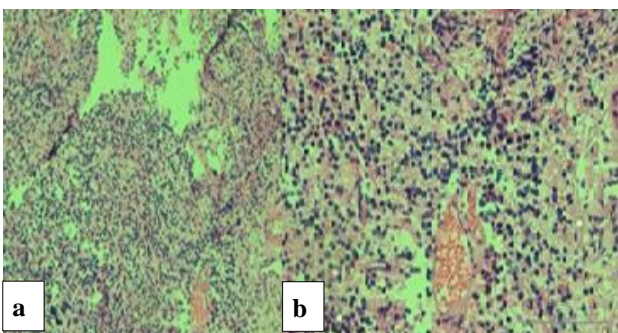


Figure 6: Photomicrographs of pathologic intramedullary spinal cord tumor tissue showing increased cellular density of neoplastic cells with increased nuclear pleomorphism and hyperchromatism at (a) mid power and (b) and high-power magnification. Final histopathologic diagnosis of astrocytoma, grade III.

Histopathology studies revealed increased cellular density of neoplastic cells with increased nuclear pleomorphism and hyperchromatism. There was increased mitotic activity observed without microvascular proliferation and necrosis. The specimens were signed out as malignant astrocytoma, World Health Organization (WHO) grade 3, anaplastic type (Figure 6). The patient was discharged after a few weeks, his wounds with satisfactory healing. His neurologic functions, however, did not improve and was still paraplegic with muscle grade of 0/5 (MMT) for both lower extremities. Bowel and bladder functions were likewise spastic with involuntary urination and defecation. The patient initially was lost to follow up after the removal of sutures and sought consultation 10 months post-op due to severe back pain, generalized body weakness and difficulty of breathing. Patient was pronounced dead 2 weeks after admission due to respiratory failure.

DISCUSSION

Astrocytomas are neoplasms of the central nervous system which arise from glial tissue supporting cells. They may arise anywhere from the brain and the spinal cord, more

common on the former with a 10:1 ratio.⁵ Primary spinal cord astrocytomas occur as intradural intramedullary tumors which have an infiltrative nature because of their location in the peripheral white cord tissue, making resection challenging and difficult. It is the most common intramedullary spinal cord tumor in the pediatric population and second in the adult population.⁴

The presentation of intramedullary spinal cord astrocytomas is variable and typically progresses over a period of months to years before a diagnosis is established. Earliest symptoms can include progressive back or neck pain which is attributed to the gradually enlarging tumor that expands the bony vertebrae. Myelopathy symptoms ensue such as unusual gait that progresses to difficulty in ambulation. Cervical lesions may also involve loss of upper limb dexterity and eventual loss of function. These are also accompanied by loss of active bladder and bowel control. Spinal deformities may be a sign of the disease due to the chronic muscle spasms which deform the growing spine. Symptoms may progress over years for more benign lesions but may be more rapid in malignant variants, which can be in a span of months with abrupt deterioration in a span of weeks.⁴

Radiographs may or may not show spinal deformities. MRI is the best diagnostic procedure which shows enlargement of the spinal cord in diameter and often length with the conus extending beyond the L2 vertebra. The tumor is seen as an intramedullary lesion having an isointense signal with the normal cord tissue on T1WI. On T2WI, the lesion has a mixture of iso and hyperintense structures because of the presence of intratumoral and peritumoral syrinx formations.⁴ Brain MRI is likewise needed to document that the lesion has not metastasized to the brain, or from the brain to the spinal cord.

The treatment goal for these lesions is to resect the tumor en bloc if possible, to prevent recurrence, while keeping in mind that preservation of neurologic function is of utmost importance. Studies have shown that the degree of functionality is directly correlated to survival rate. Patients who retain their ability to ambulate have a greater life span than those who do not.⁴ The accepted algorithm for treatment is once the spinal cord is opened, an intraoperative rush frozen section biopsy is done. If the results show benign findings, the tumor is grossly resected en bloc with macroscopic clear margins. This has shown to increase the survival rate for patients in combination with radiotherapy and chemotherapy.^{2,9} But if findings show malignant features, no further resection is done and the wound is closed.

The role of chemotherapy and radiotherapy has remained controversial particularly for malignant variants of spinal cord astrocytoma which have very poor prognosis despite of treatment. Santi reported that malignant spinal cord astrocytoma has dismal prognosis regardless of the kind of surgery, whether gross resection or biopsy alone, or whether adjuvant chemo and radiotherapy is given or not.

In their series of 36 cases of primary malignant spinal cord tumors between 1962 and 2000, the median overall survival period was only 17 months.⁸ Minehan also reported similar results of 136 patients treated from 1962 to 2005, where 69 were benign pilocytic astrocytoma and 67 were malignant variants. Patients with pilocytic tumors survived significantly longer than those with infiltrative astrocytomas (median overall survival, 39.9 versus 1.85 years). The study concluded that the histologic type is the most important prognostic variable affecting the outcome of spinal cord astrocytoma. In this study, they showed radiation therapy somehow improved the lifespan of malignant variants despite their relatively dismal prognosis (24 versus 3 months).⁹ The study conducted by Adams, wherein 135 patients all with primary malignant spinal cord astrocytomas were treated between 1973 to 2000, also showed that the prognosis of these patients did not change over decades despite the advancement of imaging technology and surgical instrumentation. Chemotherapy and radiotherapy did not alter their prognosis. The median survival rate for anaplastic type (WHO grade 3) versus glioblastoma type (WHO grade 4) is 17 versus 10 months.¹⁰

Our patient exemplifies a typical case of a primary malignant astrocytoma. History and physical examination showed sudden rapid deterioration of symptoms to complete paraplegia, which are all supported by literature on MRI and confirmed by histologic studies of anaplastic astrocytoma. We decided to do only partial resection on the lesion based on findings of malignant-like features intraoperatively. As expected, the patient did not improve neurologically because he was already in complete paraplegia state preoperatively. What was achieved in the procedure was determining the true nature and prognosis of the patient. As based on literature review, instituting radiotherapy with possible chemotherapy may possibly improve the patient's lifespan, even for a few months, despite the dismal prognosis.

Limitations

MRI scans on the brain could have helped to determine if this is truly a case of primary spinal lesion or if it has metastasized from the brain. We did not find the need to do it at the time of treatment because the patient did not show signs of cerebral involvement, up to the last follow up. Nevertheless, a brain MRI should be standard in investigating spinal cord intramedullary lesions.

Also, we were not able to do an intraoperative rush frozen section to determine the histopathologic malignancy of our specimen, due to some technical error in our facilities at the time. We mainly depended on our intraoperative findings and judged the lesion to be malignant, which was confirmed by postoperative formal histopathologic studies. An intraoperative rush frozen section biopsy should be standard whenever surgical decision making depends on the benignity or malignancy of a lesion.

CONCLUSION

This first documented case of thoraco-lumbar malignant spinal cord astrocytoma at a local tertiary medical center shows the classic features of this disease entity based on the literatures written on this subject over the decades. Not much has changed over the years as it still persists to be a challenging entity to treat given its dire prognosis despite the advancement in imaging studies, surgical techniques, instrumentation, radiotherapy and systemic chemotherapy.

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