Intramedullary spinal cord tumors (IMSCTs) are rare central nervous system neoplasm which account for 2% to 4% of all central nervous system tumors. Astrocytoma and Ependymoma are the most common intramedullary tumors. Astrocytoma is the most common among children and adolescents while ependymoma most often occur in adults. The management of spinal cord astrocytoma varies widely due to the assumption that astrocytomas are infiltrative tumors and that radical resection poses a high probability of inflicting neurological injury to the patient. There are no universally accepted guidelines for management of spinal cord astrocytoma. With advances in microsurgical technology such as the ultrasonic aspirator, surgical lasers, intraoperative ultrasound, and intraoperative neurophysiological monitoring, aggressive surgical resection has possibly become a safer. Here, we present a case of huge Intradural intramedullary spinal cord tumor which was managed with gross total resection of tumor without electrophysiological monitoring who improved neurologically after the resection.

Case Report

This 9-year-old girl presented with neck pain and torticollis for 4 month duration followed by weakness of right upper and lower limbs for 2 months and then weakness of left upper limb. She developed difficulty in swallowing for one month and was unable to walk and became bedridden for one month. There was no history of fever, loss of consciousness or trauma. Neurological examination revealed motor deficits in all limbs: motor sub scores- RUE 5/25, RLE 15/25 LUE 15/25 and LLE 15/25.
20/25, sensory could not be assessed, tone was increased in all the limbs with exaggerated reflexes and Babinski sign was positive bilaterally. Magnetic resonance imaging showed a heterogeneously enhancing intramedullary mass in cervicothoracic spinal cord with large cystic lesion extending into brainstem and fourth ventricle associated with syrinx formation in dorsal spinal cord (Figure 1). The preoperative differential diagnosis included astrocytoma and ependymoma.

Her preoperative functional status according to modified McCormick scale (for functional evaluation of patients with IMSCTs) was grade IV.

Following routine intubation the patient was kept in prone position with three point head fixation device with slight flexion of neck. Midline skin incision from inion to D3 spinous process was given, aponeurosis was divided, subperiosteal muscle dissection was done bilaterally. Cervico - upper dorsal laminectomy down to D3 level done. Dura opened and stay sutures applied, arachnoid opened and intramedullary dissection started with standard midline dissection approach. Pial retraction was obtained with tensionless 6.0 polypropylene sutures holding the median pia mater to the dural edges. After taking specimen for biopsy, tumor debulking was done from inside out, tumor spinal cord cleavage plane identified and gross total excision of the tumor was done. (Figure 2)

Post-operative Course

Although there was temporary decrement in motor deficit in immediate post operative period, her condition gradually improved and at 6 weeks follow up she was able to walk with support and able to eat food using her right hand. Her modified McCormick scale is grade III. MRI study performed after 6 weeks showed complete removal of tumor with regressing syrinx (Figure 3)

Discussion

Intramedullary spinal cord tumors are rare central nervous system neoplasms. Only 4 cases have been reported from Nepal in literature from Tribhuvan University Teaching Hospital, out of which 3 were ependymoma and only one was astrocytoma.10 The first successful resection of an intradural tumor, a fibro-myxoma, was accomplished in 1887 by Victor Horsley8 and the first successful resection of an intramedullary spinal cord tumor was performed in 1907 by Anton von Eiselsberg in Austria.6 However, the first report of an intramedullary tumor appeared in 1911 by Charles Elsberg in New York.7 Elsberg described a two-stage strategy for the removal of these tumors. At the initial operation a myelotomy would

![Figure 1: Preoperative MRI Image](image1)

![Figure 2: Intraoperative photographs showing excision of tumor (A), and after gross total resection and pial closure (B)](image2)

![Figure 3: Post operative MRI Images](image3)
Paudel et al. be performed, the surgeon would then return 1 week later to remove the spinal cord tumor. This technique allowed the neurosurgeon to remove only the extruded portion of an intramedullary tumor.

There is controversy regarding the optimal management of intramedullary astrocytoma. Since many of them are benign in nature, total resection of the tumor is potentially curable however infiltrative behavior of this tumor precludes total resection due to high risk of post-operative neurological deterioration. Constantini S et al.1 reported 164 cases of IMSCTs who underwent radical excision in patients 21 years of age and younger in whom gross-total resection was achieved in 76.8%; subtotal resections were performed in 20.1%. The majority of patients (79.3%) had histologically low-grade lesions. When comparing the preoperative and 3-month postoperative functional grades, 60.4% stayed the same, 15.8% improved, and 23.8% deteriorated. They found that long-term survival and quality of life for patients with low-grade gliomas treated by radical resection alone was comparable or superior to minimal resection and radiotherapy. Schneider C et al.14 assessed the Quality of life after surgical treatment of primary intramedullary spinal cord tumors in a small cohort of children who had undergone surgery for IMSCTs with a mean follow-up of 4.2 years. Total resection was achieved in 5 patients and subtotal resection in 7. The mean modified McCormick Scale score at the time of diagnosis was 1.7; at the time of follow-up, 1.5. The mean PedSQL 4.0 total score (Pediatric Quality of Life Questionnaire version 4) in the low-grade group was 78.5; in the high-grade group, 82.6. There was no significant difference in PedSQL 4.0 scores between the patient cohort and the normal population. Babu R et al.1 retrospectively reviewed 46 consecutive patients with spinal cord astrocytomas treated from 1992 to 2012. 67.4% underwent surgical resection, with the remaining only receiving biopsy. Of all patients, 37% worsened compared with their preoperative baseline. The occurrence of worsening increased with high tumor grade, patients having high-grade astrocytoma experiencing a higher incidence of neurological worsening than low-grade tumors (52.9% vs. 27.6%, \( P = 0.086 \)). Although the use of surgical resection resulted in a higher incidence of postoperative worsening than biopsy alone, this difference was not significant (41.9% vs. 26.7%, \( P = 0.31 \)). However, an increased extent of resection was seen to increase the rate of postoperative worsening, with 66.7% of patients who underwent subtotal or gross total resection being worse than their preoperative baseline compared with 18.8% of those who underwent partial resection (\( P = 0.0069 \)). Also resection did not provide a survival benefit compared with biopsy alone (\( P = 0.53 \)). They concluded that the resection of spinal cord astrocytomas should be reserved for select cases and should be used sparingly as surgical intervention is associated with a higher rate of neurological complications and lacks a clear benefit.

The utility of surgical resection on the outcomes of patients with spinal cord astrocytoma is currently unclear. Nakamura et al.13 evaluated 30 patients, those who underwent partial and total resection to have a significantly higher survival rate than those who only received excisional biopsies. However, most studies have not demonstrated a survival benefit with resection. A recent systematic review of the literature noted that of 14 studies that assessed the extent of resection, only 3 demonstrated improvements in progression or survival.2 In a large study by Minehan et al.15 which evaluated 136 patients with spinal cord astrocytoma during a 43-year period, biopsy alone was seen to be a significant good prognostic factor on multivariate analysis (HR, 0.45; 95% CI, 0.26–0.77; \( P = 0.004 \)). Although aggressive surgical resection of other intramedullary tumors such as ependymomas results in a clear improvement in outcomes with minimal risk of permanent neurological injury, the resection of infiltrative spinal cord astrocytoma remains an unproven therapeutic modality.1 Analysis of 225 patients with intramedullary tumor by Klekamp J9 showed that permanent morbidity was lowest after gross total resection, which was significantly correlated with surgical experience and the preoperative neurological state. In the long term, tumor recurrence rates for ependymomas and benign astrocytomas correlated significantly with the amount of resection. It concludes that intramedullary tumors should be surgically treated as soon as neurological symptoms appear. However, Surgery on intramedullary tumors should be performed by neurosurgeons who deal with these lesions on a regular basis as considerable experience is required to achieve high GTR rates and to limit rates of permanent morbidity.9

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<thead>
<tr>
<th>Grade</th>
<th>Modified McCormick Score</th>
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<tr>
<td>1</td>
<td>Intact neurologically, normal ambulation, minimal dysesthesia</td>
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<tr>
<td>2</td>
<td>Mild motor or sensory deficit, functional independence</td>
</tr>
<tr>
<td>3</td>
<td>Moderate deficit, limitation of function, independent w/ external aid</td>
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<tr>
<td>4</td>
<td>Severe motor or sensory deficit, limited function, dependent</td>
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<tr>
<td>5</td>
<td>Paraplegia or quadriplegia, even w/flickering movement</td>
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Conclusion

The total or gross total resection of the pilocytic astrocytoma of spinal cord should be the surgical goal because of its benign nature and potential for cure. Use of modern technology and intraoperative neurophysiological monitoring has made this goal a safer possibility. In our case we performed the gross total excision of pilocytic astrocytoma without neurophysiological monitoring who improved neurologically after the excision of tumor.
References