

# Cervicodorsal Intramedullary Ependymoma Presenting with Spontaneous Intratumoral Bleed and Acute Quadriparesis

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## ABSTRACT

Ependymomas are the most common intramedullary spinal cord tumors in adults and account for 1–5% of spinal tumors. The presence of hemorrhage within the spinal ependymoma is not uncommon on imaging studies, however an ependymoma of the cervicodorsal region presenting as sudden acute onset quadriparesis following intratumoral bleed has never been reported in world literature. This rare case of spontaneous hemorrhage into a cervicodorsal intramedullary ependymoma in a 40-year-old lady was immediately operated and showed excellent recovery.

**Keywords:** Acute quadriparesis, Ependymoma, Hemorrhage.

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## INTRODUCTION

Ependymomas are the most common intramedullary spinal cord tumors in adults and account for 1–5% of spinal tumors.<sup>1,2</sup> The presence of hemorrhage within the spinal ependymoma is not uncommon on imaging studies,<sup>1-4</sup> however, an ependymoma of the cervicodorsal region presenting as sudden acute onset quadriparesis following intratumoral bleed has never been reported in world literature. This rare case of spontaneous hemorrhage into a cervicodorsal intramedullary ependymoma in a 40-year-old lady was immediately operated and showed excellent recovery.

## CASE REPORT

A 40-year-old lady had presented with progressive stiffness of both the lower limbs and occasional imbalance while walking of 1-month duration. She had developed

sudden onset weakness of both the upper and lower limbs on the day of arrival to the hospital with an inability to walk without support along with the loss of bladder control. She had a history of normal menstrual cycle and was not on any anticoagulant medication nor did she have any history of lifting heavy objects.

On examination she was afebrile, pulse rate was 76/minute, Blood pressure was 124/70 mm Hg. She had no neck stiffness, was conscious and oriented with bilateral normal cranial nerves examination. Her upper limb tone was normal while both the lower limbs had increased tone.

Motor examination revealed bilateral 4-/5 power in the shoulder, elbow, wrist, and digits. Bilateral lower limb power was 4/5. Bilateral deep tendon reflexes were diminished in the upper limb and exaggerated in the lower limbs along with bilateral positive Babinski's sign.

She had decreased pain, temperature and touch sensation below cervical (C)<sup>5</sup> dermatome along with the impaired joint position in the lower limbs.

Her hematological parameters and the coagulation profile were within normal limits.

She underwent a magnetic resonance imaging (MRI) of the cervical spine which revealed an intramedullary lesion extending from cervical (C)<sup>4</sup> to thoracic (T)<sup>1</sup> along with cord widening. The lesion was iso to hyperintense on T1 weighted (wt) images (Fig. 1). The lesion with intratumoral acute bleed was hyperintense on T2 weighted Images and



**Fig. 1:** T1 weighted sagittal image of the cervical spine showing iso to hyper intense lesion in the cord (C3- T1) with widening of the cord from C4 to T1

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showed a blood fluid level (Fig. 2). On contrast imaging, the periphery of the lesion was enhancing moderately (Fig. 3).

The patient was taken up for emergency surgery. In prone position a cervical laminectomy C4–C7 was performed, after opening the dura, a midline myelotomy was done, and gross total excision of the tumor achieved. Intraoperatively there was intratumoral bleed (Fig. 4), the lesion was greyish, soft, vascular and could be dissected off the cord. Small bits of tumor adherent to the cord tissue was left behind.

Postoperatively the patient had recovered well and was able to walk with support 3 days after the surgery. At 6 months follow-up after the surgery the patients motor power had improved in the upper limbs to 4+/5 in the shoulder, elbow, and wrist. The muscle strength in the lower limbs had improved to 5/5, her joint position had improved, she was able to walk independently, and she had regained full bladder control.

## DISCUSSION

Amongst the intramedullary spinal tumors in adults, ependymomas account for 50–60%.<sup>1</sup> These are slow

growing tumors with the average time between presentation and diagnosis being more than 2 years.<sup>2,3,5</sup> Surgery is the primary modality of treatment. Achieving a gross total resection followed by adjuvant radiotherapy has been the accepted norm for optimal benefit and progression-free survival in patients with spinal ependymomas.<sup>1,6,7</sup> As per our literature search, there have been 16 case reports of patients with spinal ependymoma presenting with acute neurological deterioration due to spontaneous bleed within the tumor, of these only in 3 cases the cervical cord involvement has been reported. This is the first reported case of cervicodorsal-ependymoma presenting with acute onset quadripareisis following a spontaneous hemorrhage into the tumor.

We are unable to pinpoint the exact cause of the hemorrhage and sudden worsening in this patient as she did not have any predisposing factors like pregnancy, anticoagulation medication, lumbar puncture, heavy weight lifting, diabetes hypertension or thrombocytopenia. Amongst some of the previously reported acute neurological worsening in spinal ependymomas, the cause has been anticoagulant medication, pregnancy, and heavy lifting.<sup>1</sup>

On the magnetic resonance imaging (MRI) study, the lesion showed a blood fluid level with poor to moderate heterogeneously enhancing lesion in the C3 to T1 level with irregular borders whereas ependymomas are usually well-circumscribed lesions with a smooth and regular margin.<sup>1,8</sup>

Following emergency surgery, the patient had shown improvement in her motor power, joint position sense, subjective improvement in pain and temperature sensation along with bladder control. This has been usually seen in most previous cases with acute neurological worsening following a spontaneous bleed in spinal ependymoma. Although in a few cases, surgery delayed by a week to 1 month have also resulted in full recovery,<sup>1,2</sup>



Fig. 2A: T2 weighted sagittal and axial image

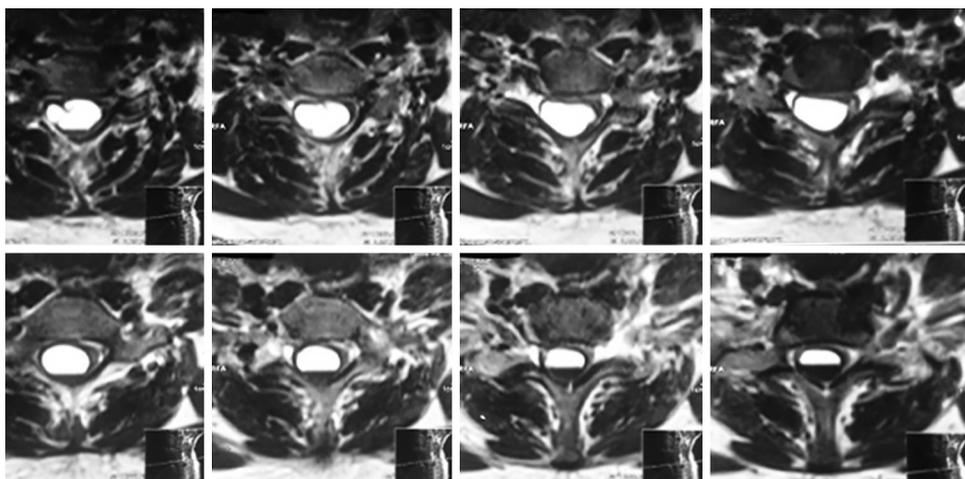
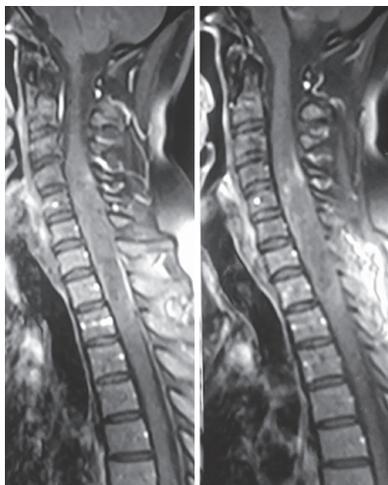


Fig. 2B: T2 weighted axial images with hyper intense signal of acute blood with fluid level within the cord



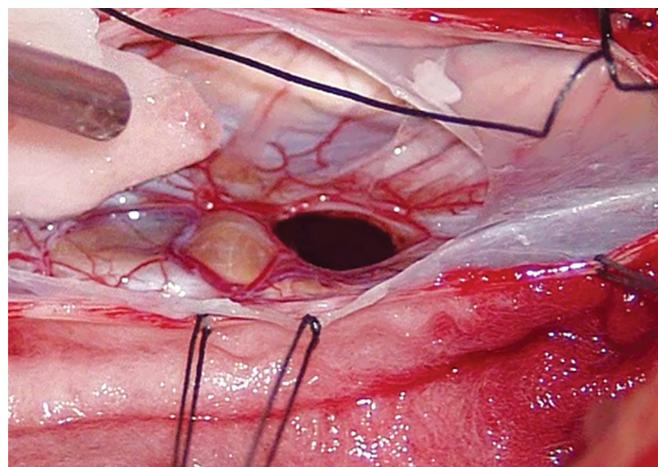
**Fig. 3:** T1 weighted contrast enhanced images of the cervical spine showing moderate heterogenous peripheral contrast enhancement of the intramedullary lesion

but none of them had lesions in the cervical region and most of them, in fact, were intradural extra medullary lesions in the lumbar spine. In patients where surgery was delayed by 1 week or more after intratumoral hemorrhage, authors have reported extensive adhesions between the tumor and the normal spinal cord resulting in increase in difficulty and the length of the procedure.<sup>1,2</sup> Also surgeons should not stick to total removal in infiltrated and adhering tumors as subtotally resected tumors with postoperative radiotherapy have not always recurred.<sup>9</sup>

Though a series of surgical management of spinal ependymomas have earlier been published, early diagnosis and treatment are associated with a favorable outcome whereas patients with sphincter dysfunction have a poor outcome.<sup>3,5,10</sup> Heuer et al.<sup>2</sup> have shown excellent results with complete recovery following a delayed surgery in 2 patients with intratumoral bleed in the lumbar spine; however, emergency surgery should be advised and is recommended to avoid encountering adhesions during surgery.

## CONCLUSION

This report is intended to highlight the importance of recognizing spontaneous hemorrhage in a cervical intramedullary spinal tumor as a cause of acute neurological deterioration in young adults. Although emergency surgery is recommended in acute neurological worsening following a spontaneous hemorrhage in ependymomas, we suggest all patients with intramedullary cervical tumors should be counseled and advised early intervention.



**Fig. 4:** Intraoperative image of the cervical cord showing intratumoral hemorrhage

## REFERENCES

1. Lee S-H, Park DJ, Jeun S-S. Acute Paraplegia as a Result of Hemorrhagic Spinal Ependymoma Masked by Spinal Anesthesia: Case Report and Review of Literature. *Brain Tumor Research and Treatment*. 2016;4(1):30-34.
2. Heuer GG, Stiefel MF, Bailey RL, Schuster JM. Acute paraparesis from hemorrhagic spinal ependymoma: diagnostic dilemma and surgical management. Report of two cases and review of the literature. *J Neurosurg Spine* 2007;7:652-655.
3. Martinez-Perez R, Hernandez-Lain A, Paredes I, Munarriz PM, Castaño-Leon AM, Lagares A. Acute neurological deterioration as a result of two synchronous hemorrhagic spinal ependymomas. *Surgical Neurology International*. 2012;3:33.
4. Yoshii S, Shimizu K, Ido K, Nakamura T. Ependymoma of the spinal cord and the cauda equina region. *J Spinal Disord* 1999;12:157-61.
5. Schweitzer JS, Batzdorf U. Ependymoma of the cauda equina region: Diagnosis, treatment, and outcome in 15 patients. *Neurosurgery* 1992;30:202-207.
6. Safae M, Oh MC, Mummaneni PV, Weinstein PR, Ames CP, Chou D, et al. Surgical outcomes in spinal cord ependymomas and the importance of extent of resection in children and young adults. *J Neurosurg Pediatr* 2014;13(4):393-399.
7. Klekamp J. Spinal ependymomas. Part 1: Intramedullary ependymomas. *Neurosurg Focus* 2015;39:E6.
8. Kucia EJ, Bambakidis NC, Chang SW, Spetzler RF. Surgical technique and outcomes in the treatment of spinal cord ependymomas, part 1: intramedullary ependymomas. *Neurosurgery* 2011;68(1 Suppl Operative):57-63; discussion 63.
9. Sakai Y, Matsuyama Y, Katayama Y, Imagama S, Ito Z, Wakao N. Spinal myxopapillary ependymoma: neurological deterioration in patients treated with surgery. *Spine (Phila Pa 1976)*. 2009 Jul 1;34(15):1619-24
10. Sasanelli F, Ordesi G, Campo B, Mariani G. Altered bladder function as the only and late presenting symptom of ependymoma of the filum terminale. *The Italian Journal of Neurological Sciences*. 1987;8(4):387-389.