

## Cervicomedullary Ependymoma with Hemorrhage: A Case Report and Review of Literature

### Abstract

Ependymoma is a rare tumor central nervous system that arises from the ependymal lining of the ventricles or the central canal of the spinal cord. They are of neuroectodermal in origin and constitute about 30%–86% of tumors arising in the spinal cord. The occurrence of these tumors in the cervicomedullary region is very rare. Sudden symptomatic neurologic presentations due to hemorrhage in cervicomedullary ependymoma is very rare and so far have never been reported. Mostly presenting as neurologic deficits involving limbs, these tumors pose a technical challenge in their removal. We present a patient who presented with sudden-onset dysesthesia of the upper and lower limbs. On imaging, he was found to have a cystic medullary tumor extending to the cervical region with hemorrhage. We discuss the epidemiology, surgical challenges, and outcome along with review of literature of these rare tumors located in this precarious location.

**Keywords:** Cervicomedullary, ependymoma, hemorrhage

### Introduction

Intramedullary ependymoma in the cervicomedullary region is rare. These slow-growing benign tumors may pose serious neurologic deficits as a result of compression of vital brainstem structures. Sudden hemorrhage into a cervicomedullary ependymoma (CME) is an uncommon event, which can lead onto potential life-threatening symptoms. We present such a case of CME and its surgical management along with review of literature.

### Case Report

A 37-year-old male presented with dysesthesia of 4 months' duration involving his both upper limbs. On enquiring further, it was revealed that his symptoms started all on a sudden before 4 months and has been gradually increasing ever since. There was no history of neck pain. There was no history of trauma to the neck. On examination, general examination was unremarkable. There were no cranial nerve palsies. Motor system examination revealed that he had Grade 5 power in both upper and lower limbs and deep tendon reflexes were normal. There was sensory loss involving upper limbs from the nape of

the neck downward. Local skull and spine examination were normal.

He underwent a magnetic resonance imaging (MRI) of the brain along with cervical spine which showed an intra-axial tumor with a cystic component in the medullary region extending below to the second cervical vertebra [Figure 1a and b]. The tumor was mildly hyperintense on T1-weighted image and hyperintense on T2-weighted image. There was minimal contrast enhancement of the cystic wall of the tumor. There was fluid level in the cavity which was hypointense on both T1 and T2 weighted images suggestive of previous hemorrhage [Figure 1a-c]. Our preoperative differential diagnosis included pilocytic astrocytoma, hemangioblastoma, and ependymoma.

### Surgery

He was taken up for surgery under neuromonitoring. He underwent a posterior fossa midline craniotomy along with a laminectomy of the first cervical vertebra in the prone position. The tumor was exposed in the midline. The cystic part of the tumor was let out which contained xanthochromic fluid, suggestive of previous bleeding. The tumor was soft and moderately vascular which had a well-demarcated margin with that of

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### Access this article online

**Website:** www.asianjns.org

**DOI:** 10.4103/ajns.AJNS\_233\_19

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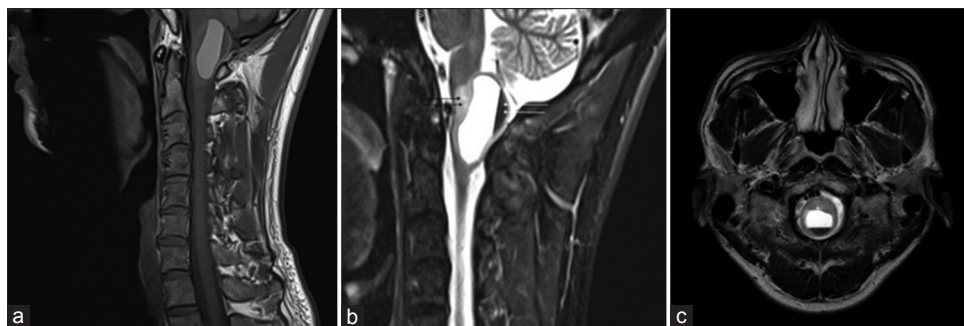
**How to cite this article:** Kutty RK, Ohmori K, Yamada Y, Kato Y. Cervicomedullary ependymoma with hemorrhage: A case report and review of literature. *Asian J Neurosurg* 2020;15:190-3.

**Submission:** 21-07-2019      **Accepted:** 21-11-2019

**Published:** 25-02-2020

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**Figure 1:** Magnetic resonance imaging of the craniocervical junction. (a) T1-weighted image with contrast showing a well-defined iso-to-hyperintense lesion in the region of the medulla with predominantly cystic component. There is a fluid level seen posteriorly in the cyst suggestive of remote hemorrhage into the cyst. (b) T2-weighted sagittal image showing the tumor to be moderately enhancing with contrast in the anterior part with a fluid level in the posterior part. (c) Magnetic resonance imaging T2-weighted axial image showing the predominantly cystic tumor with a fluid level at the level of medulla

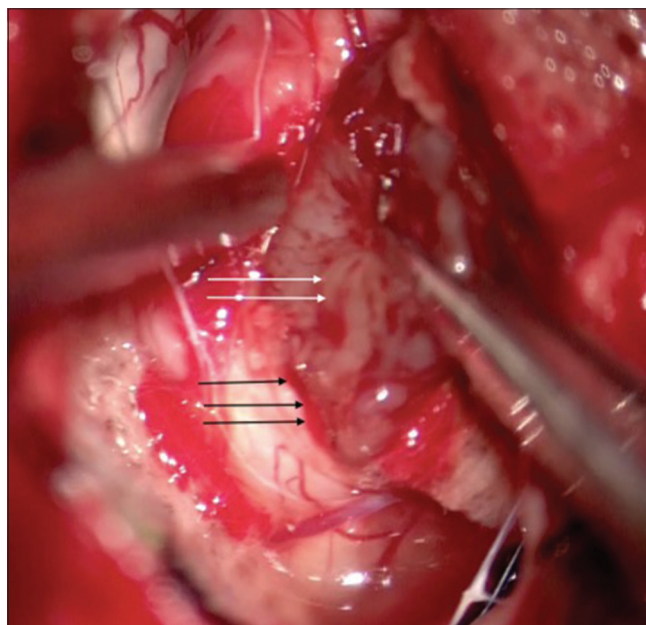
the normal cervical cord in the superficial part [Figure 2]. However, as the dissection progressed to the deeper parts, this demarcation was less well defined. The tumor was removed in piecemeal and intraoperative frozen section was done which was suggestive of high-grade glioma. The debulking of the tumor was performed during which profuse bleeding from the base was observed. A sudden drop in the transcranial motor evoked potentials as well as in blood pressure occurred associated with occurrence of arrhythmias after which the decision to proceed further was abandoned. Postoperatively, he was examined clinically and there were no new focal neurological deficits.

The histopathology of the tumor showed round cell tumor with rosette formation [Figure 3]. There was no evidence of calcification, necrosis, or neovascular proliferation. The MIB index was 3.7%. There was no mitosis. The tumor cells were positive for GFAP and EMA and negative for P53, synaptophysin, and NSE, suggestive of Grade 2 ependymoma.

A repeat MRI with contrast was taken [Figure 4] which showed residual tumor involving the base of the excision cavity. The option of a repeat surgery or wait and watch with serial imaging was discussed with the patient. After discussions with the family, the patient decided against surgery and is now on follow-up.

## Discussion

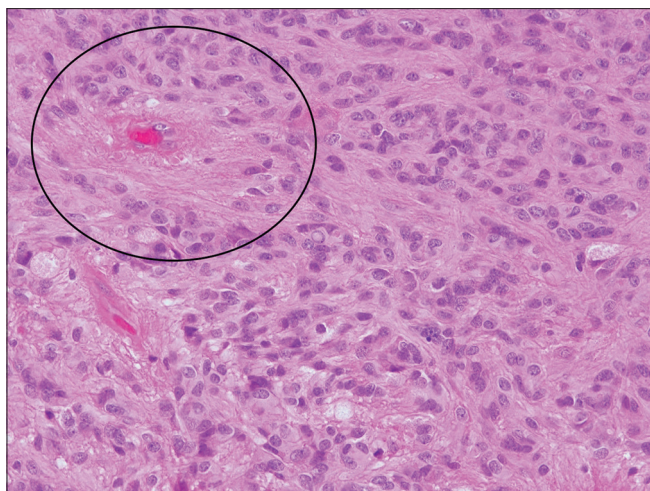
Intramedullary CMEs (ICMEs) are rare tumors of the neuraxis. "They constitute about 3.8%,<sup>[1,2]</sup> to 23%<sup>[3]</sup> of the cases in various reported series of ependymomas involving spinal cord. They are usually slow-growing and benign tumors, which manifest symptoms by compression of the brainstem or higher cervical cord. Those lesions arising predominantly in the medulla present with lower cranial nerve palsies along with motor and sensory deficits while lesions predominantly involving higher cervical cord present with features of myelopathy. In a series of 28 patients with CMEs by Ge *et al.*, the most common symptoms were in the form of motor deficits following by neck pain and sensory changes.<sup>[4]</sup> The clinical progression



**Figure 2:** Intraoperative picture. Fleshy tumor (white arrows) with hemorrhagic areas, after decompression of the cystic part. Note the distinct demarcation between the tumor and the normal medulla (black arrows)

was protracted in most of the cases ranging from 1 to 72 months. The precarious location of the lesions creates a significant challenge to their removal. However, these inherently slow-growing tumors tend to grow circumferentially around the pial structures than infiltrating the brainstem. This course of the tumor is explained by the obstruction it encounters due to the presence of the white matter tracts, viz., pyramidal decussation and medial lemniscus preventing their natural course into the pontomedullary junction.<sup>[5]</sup> Thus, they have a good plain of cleavage with the normal neural cord enabling their gross total removal most of the times.

Acute neurologic presentations of ICME are very rare. Although ependymomas involving the spinal cord in the thoracolumbar,<sup>[6-8]</sup> lumbar,<sup>[9-16]</sup> thoracic,<sup>[17,18]</sup> and cervicothoracic<sup>[19]</sup> regions have been reported to show acute hemorrhagic manifestation, such instances in ICME have never been reported. A thorough search on

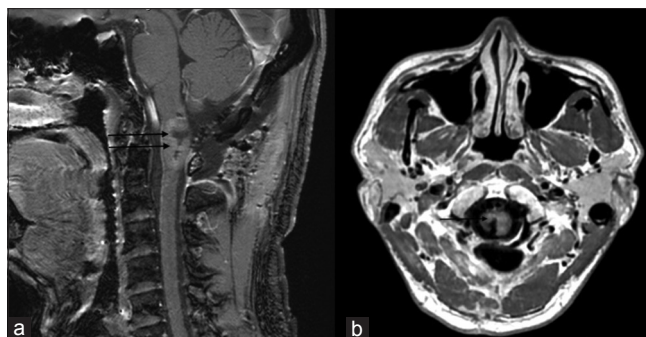


**Figure 3: Microphotograph showing pseudorosette arrangement of the cells of ependymoma (H and E, x40)**

the popular medical search engines such as PubMed and Google Scholar was done to confirm this finding. Our patient had an episode of acute bleeding into the ICME which manifested as severe dysesthesia which mitigated over 1-month duration.

During the surgery, there was a well-defined plain of cleavage between the tumor and the brain stem. The intratumoral hemorrhage was seen as xanthochromic fluid after opening the cystic part of the tumor. The frozen section examination of the tumor by the pathologist was described as a high-grade malignancy. The tumor-brainstem interface was followed to resect most of the tumor, but as the dissection progressed, this plain of cleavage was ill-defined in the deeper regions. During the course of this deeper dissection, there was a gross change in the motor evoked potential which hindered further removal. This was accompanied by transient fluctuations in the blood pressure as well as cardiac rhythm. In this situation, considering the aggressive nature of the lesion, it was decided to not to further perceive overzealous resection compromising the safety of the patient. Postoperatively, the patient did not develop any further new neurologic deficits. The final histopathology of the tumor came as Grade 2 ependymoma after which the need for repeat surgery versus radiotherapy was discussed along with option of maintaining close follow-up with imaging clinically. Such cases of low-grade ependymoma has been shown to be stable without progression for months on follow-up.<sup>[20,21]</sup> Many reports suggest radiotherapy for such subtotal resections in cases of ependymoma.<sup>[4,21]</sup> However, in our patient, we considered to follow him up with serial imaging considering the precarious location of the tumor. Studies have reported patients ending up in respiratory paralysis needing ventilatory assistance in cases where surgeons have chased the tumor aiming for gross total resections in cervicomedullary ependymomas.<sup>[4]</sup>

The prognosis of spinal ependymomas is overall better than their cranial counterparts. The most important prognostic



**Figure 4: Magnetic resonance imaging of the craniocervical region. (a) T1-weighted with contrast sagittal – residual tumor is seen as enhancing areas in the anterior part of the cystic cavity (horizontal black arrows). (b) Magnetic resonance imaging T1-weighted axial image with contrast-excision cavity along with the contrast enhancing areas in the anterior half of the medulla, suggestive of residual tumor (horizontal black arrow)**

factor in the outcome of spinal ependymoma is the gross total resection.<sup>[4]</sup> Recurrence rates can vary from months to years. Moreover, radiological recurrences may not be necessarily accompanied by neurological deficits. Hence, various options including surgery or radiotherapy should be discussed along with the patient in case of recurrences on follow-up.

## Conclusion

Ependymoma in the cervicomedullary region is a rare tumor which can present with hemorrhage. Intraoperative neuromonitoring can guide to the safe removal of this lesions. Every attempt should be made for gross total resection with preservation of vital functions because they are indolent and benign, slow growing tumors with good long-term survival.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

## Financial support and sponsorship

Nil.

## Conflicts of interest

There are no conflicts of interest.

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