Cauda equina schwannoma presenting with intratumoral hemorrhage and intracranial subarachnoid hemorrhage

Case report

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The authors report the case of a spinal intradural schwannoma presenting with intracranial subarachnoid hemorrhage (SAH). Cerebral angiography did not show any intracranial lesion; however, MRI revealed two separate tumors in the lower segment of the spinal cord. The proximal lesion arising from the conus medullaris was well circumscribed and homogeneously enhanced, whereas the tumor in the cauda equina revealed hemorrhagic signals on MRI. This case also illustrates an unusual presentation of intracranial SAH simultaneously with intratumoral hemorrhage in a spinal cord schwannoma. The absence of hemorrhagic changes in the lesion arising proximal to the cauda equina region supports the mechanical theory proposed for the pathogenesis of hemorrhagic complications in spinal cord tumors.

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KEY WORDS • spinal cord schwannoma • subarachnoid hemorrhage • intratumoral hemorrhage • cauda equina • oncology

Intracranial subarachnoid hemorrhage (SAH) arising from spinal pathology is a rare event. Spinal trauma, arteriovenous malformation, and saccular aneurysms are common pathologies of the spine that are associated with SAH; however, the spontaneous onset of SAH has also been described.1,10,13 Spinal cord tumors, primary or metastatic, presenting as intracranial SAH have been occasionally reported.1,12,14 Spinal schwannomas are rare causes of SAH, especially without any spinal or nerve root compression symptoms.12 Intratumoral hemorrhage in spinal cord schwannoma, another unusual feature, has also been documented.1,8,15 We report a rare case of spinal schwannoma with the simultaneous presentation of intracranial SAH and intratumoral hemorrhage. We have also described the details of the microsurgical removal of this tumor and discussed the pathogenesis of intracranial SAH in cauda equina spinal cord tumors.

Case Report

History and Examination. A 47-year-old man presented with a history of seizure episodes occurring 3 months earlier. There was no history of headache, vomiting, or visual changes associated with the seizures. He was treated at an outside hospital, and the seizures did not recur; however, left leg pain developed and became aggravated a few days before his presentation to us. No abnormality was detected on physical examination. The patient was evaluated for seizures, and CT studies of the head revealed SAH in the suprasellar anterior interhemispheric cistern, sylvian fissures, and basal cisterns (Fig. 1). Factors predisposing to spontaneous SAH, such as chronic medical conditions or anticoagulant therapy, were ruled out. Three-dimensional cerebral angiography failed to demonstrate the intracranial source of the SAH. Magnetic resonance imaging of the spine revealed an irregularly shaped lesion at the L1–2 level, which had cystic portions with areas of bleed within and outside the tumor (Fig. 2). The tumor enhanced heterogeneously on T1-weighted postcontrast images. There was another lesion superiorly at the adjacent level (T-12), which was hypointense on T1- and hyperintense on T2-weighted images with homogeneous enhancement on postcontrast images. Spinal angiography was deferred given obvious

Abbreviation used in this paper: SAH = subarachnoid hemorrhage.
evidence of hemorrhage within the spinal tumor and the absence of flow voids on MRI. Degenerative changes were seen in the lumbar spine at and below the level of the tumors.

Diagnosis of SAH as a result of spinal cord tumor was made, and resection was planned in the same inpatient setting.

Operation. The patient underwent T-12, L-1, and L-2 laminectomy, and after opening the dura mater, a well-organized hematoma was encountered. Blood clots were removed, and a large hemorrhagic mass was seen at the L1–2 level along with another separate lesion superiorly, as already depicted on MRI, at the T-12 level. Hemorrhagic tumor was separated from the surrounding neural tissues using standard microsurgical technique (Fig. 3). Residual blood clots and tumor tissues were gently separated from the cauda equina (Video 1).

Postoperative Course. The patient had an uneventful inpatient stay and was discharged on the 5th postoperative day. Nimodipine was given as a vasospasm prophylaxis at the time of presentation and was continued for 3 weeks’ duration. At the 3-month follow-up, the patient reported significant improvement in his leg pain without any new episode of seizure; hence, subsequent angiography was deferred. The patient continued to use anti-seizure medications, which will be tapered over the next few months.

Discussion

Spinal causes are responsible for up to 1.5% of all cases of SAH, although a lower incidence has also been reported.7,16 Patients with SAH from spinal pathology present mainly in their 3rd decade.12 Of the primary spinal cord tumors, ependymoma of the conus medullaris–cauda equina region is the most common cause of SAH, followed by schwannoma and rarely hemangioblastoma.2,4,9,11 In their review, Parmar et al.12 identified 20 cases of spinal cord schwannoma, 7 cases of which presented with intracranial symptoms only. Patients with intracranial SAH usually present with meningism, headache, and mental tissue and excised, with sacrifice of the nerve root. A nonhemorrhagic proximal lesion was removed in a piecemeal fashion. Copyright Anil Nanda. Published with permission. Click here to view with Media Player. Click here to view with Quicktime.

The tumor was attached to a nerve root, which was sacrificed. The resected tumor was ovoid in shape and cystic in consistency with a tense glistening capsule. The tumor at the T-12 level was also completely resected in a piecemeal fashion.

On histology, the section of the hemorrhagic mass confirmed the diagnosis of schwannoma with a combination of Antoni A and B architecture. There were areas of fresh hemorrhage and focal necrosis. Immunohistochemistry was positive for S100 protein. The section of the proximal lesion also showed the characteristics of schwannoma (Fig. 4).

Fig. 1. Axial CT scans of the brain revealing diffuse SAH in the suprasellar anterior interhemispheric cistern, sylvian fissures, and basal cisterns.

Fig. 2. Postcontrast T1-weighted MR image (A) showing two separate hypointense lesions at the T12–L1 level. The superior lesion enhanced homogeneously, whereas the inferior mass had irregular enhancement with a lobulated appearance. Sagittal T2-weighted (B) and T2 gradient echo (C) MR images showing areas of heterogeneous hypointensities within (white arrows) and outside (white arrow with black outline) the tumor, suggestive of acute hemorrhage.
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status changes; however, the clinical presentation can be extremely variable, ranging from nearly asymptomatic to sudden death. The patient in our case did not have any cranial symptoms, although his history of seizures could have been associated with the intracranial hemorrhage caused by the undetected intraspinal schwannoma.

Many theories, both vascular and mechanical, have been proposed to understand the mechanism of hemorrhage from spinal tumors into the subarachnoid space. According to the vascular theory, ecstatic and hyalinized vessels of the tumor may undergo spontaneous thrombosis, which results in tumor necrosis and hemorrhage. Mechanical factors also explain the unusual presentation of SAH in spinal tumors, particularly at the cauda equina region. The mechanical theory states that the abnormal movements between spinal tumor and cauda equina produce traction forces, which stretch the most superficial vascular structure, resulting in hemorrhage. This may explain the appearance of symptoms as a result of exertion in some cases. In our case the SAH spontaneously developed, which may be explained by the mechanical factors described in the cauda equina region. The absence of hemorrhagic signs in the T-12 tumor also supports the mechanical theory. Degenerative changes of the spine in our patient also may have contributed to the mechanical stresses on the tumor, which has been discussed in an earlier report.

Intratumoral hemorrhage in spinal schwannoma is a rare occurrence. Ichinose et al. reviewed only 3 cases of solitary intratumoral hemorrhage, cases that presented exclusively with spinal symptoms and neurological deficit in the lower limbs. In our case, there was clinico-radiological and histological evidence of intratumoral hemorrhage with simultaneous intracranial SAH, which, to the best of our knowledge, has not been previously described. Magnetic resonance imaging gives useful information about

**Fig. 3.** Intraoperative images. **A:** Intrathecal hematoma seen after opening the dura. **B:** Two lesions (white arrows) are visible with intervening normal spinal cord. **C:** The inferior ovoid hemorrhagic mass was carefully separated from the surrounding nerve roots and excised. **D:** A superior well-circumscribed nonhemorrhagic lesion was also completely resected.

**Fig. 4.** Histopathological findings. **Left:** Section of nonhemorrhagic lesion shows a cellular area consisting of elongated spindle cells with Antoni A and B structures. **Right:** Section of hemorrhagic mass also reveals typical characteristics of schwannoma along with areas of fresh hemorrhage and microcystic changes. H & E, original magnification × 200.
the duration and type of hematoma. In contrast to the hyperintense well-defined nonhemorrhagic schwannoma at the T-12 level, the hemorrhagic tumor had heterogeneous hypointensities on T2-weighted and T2 gradient echo images, which is suggestive of acute intratumoral hemorrhage. Low signal intensity seen on T2-weighted images was mainly due to the presence of deoxyhemoglobin. Exacerbation of leg pain may have been associated with a recent hemorrhagic event within the tumor.

Conclusions

We suggest that spinal causes should always be ruled out in patients with angio-negative intracranial SAH. Magnetic resonance imaging is a sensitive imaging modality to detect intratumoral hemorrhage, and signal intensity within the tumor helps to assess the age of the hemorrhage. We also support the mechanical theory, which describes the abnormal stresses on cauda equina tumors leading to SAH, as in our case in which the lesion situated proximal to the cauda equina did not develop signs of hemorrhage. Degenerative changes in the spine may also play a role in the pathogenesis of intracranial SAH in spinal cord tumors.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Nanda, Kukreja. Acquisition of data: Kukreja, Sharma. Drafting the article: Kukreja, Ambekar. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version: all authors.

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