Hemorrhage within the Schwannoma of Thoracic Spinal Cord presenting as Acute rapidly Progressive Paraplegia: A Rare Case

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ABSTRACT

Spinal schwannomas are benign, slow-growing nerve sheath tumors presenting as insidious lesions. Acute presentation is rare with spinal schwannomas. An intratumoral bleed causing acute presentation with paraplegia and its recovery after surgical intervention is rarely reported in the literature. We report one such case and discuss the possible mechanism for such presentation and the available literature on the subject.

Keywords: Acute paraplegia, Hemorrhagic schwannoma, Thoracic cord.

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INTRODUCTION

Nerve sheath tumors of the spinal cord are slow-growing, benign lesions having an indolent course of presentation with pain and slowly progressive neurological symptoms. Acute presentation of schwannoma as acute rapidly progressive neurological deficits is rarely reported in the literature. We present a case of hemorrhage within the schwannoma of thoracic spinal cord, precipitated due to traumatic jolting, presenting as acute rapidly progressive paraplegia.¹

CASE REPORT

A 48-year-old man, bus driver by occupation, presented with the history of acute onset of bilateral lower limb weakness which got precipitated following a powerful jolting movement of the speeding bus that headed against a speed breaker. He perceived an acute severe shooting pain over the upper back and subsequently noticed gradually progressive weakness and numbness of both lower limbs ascending from feet till the level of the nipples. The weakness worsened rapidly within 24 hours to a stage where he could not move his either legs along with retention of urine.

His neurological examination of lower limbs revealed an increased tone, a power of 0/5, an exaggerated knee, and ankle jerk with clonus, with 80% loss of sensations in both lower limbs, right more than left extending up to the nipples.

He was evaluated elsewhere with a noncontrast magnetic resonance imaging (MRI) of thoracic spine (Figs 1A and B), which revealed an intradural extramedullary lesion at the level of D2-3 on the left side (Figs 2A, B and 3A), causing significant shift in the cord and cord compression.

In view of rapidly worsening neurological status, a contrast MRI was not done and the patient was taken up for immediate surgery. He underwent a left D2-3 hemiamyotomoy, and on dural opening, the tumor was seen arising from left D3 dorsal root as a greyish blue (likely due to the hemorrhage inside), capsulated, extramedullary lesion. The tumor was excised in toto, microsurgically.

A cut section (Fig. 3B) of the gross specimen revealed the hemorrhage within the tumor. The histopathological report (Fig. 3C) confirmed schwannoma with hemorrhage.

Postoperatively, the patient was gradually mobilized with physiotherapy, and his power improved to 4/5 and was ambulant with minimal support.

DISCUSSION

Spinal cord tumors constitute 15% of central nervous system tumors, of which 90 to 95% are benign, primary tumors arising from nerve sheath or meninges. Nerve sheath tumors in the general population are uncommon with a reported annual incidence of 0.3 to 0.4/10,000. They usually occur between 4th and 6th decade with a male preponderance as against the intracranial or peripheral locations where females predominate.

Schwannomas are the major subtype of nerve sheath tumors, usually solitary, sporadic, and may be associated with neurofibromatosis type II.
Schwannomas are usually intradural but may be partly or wholly extradural. They are slow-growing tumors and 75% arise from the dorsal rootlets. Patient typically presents with a dull-aching local pain and a radicular pain along the dermatome of the nerve root from which it arises. Progressively, the patient may present with compressive myelopathy or cauda which develops over months. However, an acute presentation of a schwannoma of the spinal cord is rarely reported in the literature.

Mahadewa et al. reported a case of acute complete paraplegia due to a lumbar schwannoma, which after surgery recovered gradually over 6 months. The lesion was a schwannoma with solid and cystic components and did not reveal any signal changes suggestive of bleed within the lesion. Intraoperatively, there was no hematoma within the lesion and gross and histopathological examination of the specimen did not reveal any bleed within the lesion. They postulated that microscopic enlargement of the tumor in a small spinal canal would have precipitated acute neurological worsening and claimed that a vascular insult is unlikely, as the patient made a significant recovery.

Hemorrhage within schwannomas is very rare. Vestibular schwannomas have an estimated risk of less than 1% hemorrhage occurrence within the tumor and the various causes proposed include large size (>3 cm) with increased vascularity, chronic hypertension, anticoagulant therapy, and rarely trauma. Hemorrhage within spinal schwannomas is even rarer, with only very few case reports available in the literature. Spinal schwannomas with associated subdural and subarachnoid bleed are also reported.

Ichinose et al. reported, in a 64-year-old man, a spinal schwannoma of cauda equina with intratumoral bleed, presented as acute paraplegia which was operated and the patient made recovery. In their review, they identified only two other cases of such intratumoral bleed. They proposed that the ectatic and hyalinized vessels within the tumor may thrombose and subsequently go for necrosis and hemorrhage. They also proposed that schwannomas are highly vascularized tumors and...
a mechanical or physical stress to the tumor due to traction during mobility along the spinal axis during acute movements of the spine could also precipitate such hemorrhages within the tumors.\(^5\)

Yeh et al\(^6\) reported on a long segment schwannoma of the thoracic cord with hemorrhage presented as acute complete paraplegia. Sahoo et al\(^7\) reported a cervical schwannoma with hemorrhage presenting as acute quadriplegia.

In an MRI, schwannomas are typically homogenously hyperintense in both T1 and T2 weighted images and are intensely contrast enhancing. An inhomogeneous or heterogeneous appearance of the lesion in T2 weighted images suggests a cyst, hemorrhage, or dense collagen accumulation. An intratumoral hemorrhage shows different signal intensities depending on the age of the bleed. An understanding of the history and radiological features can help in early decision-making and an appropriate intervention, which can significantly affect the outcome of patients in this acute setting.

**CONCLUSION**

Spinal schwannomas can present with acute neurological worsening and can be due to mechanical aggravating factor. When operated immediately can have good neurological recovery.

**REFERENCES**