

Case Report

Cervical C2 to C4 schwannoma with intratumoral hemorrhage presenting as acute spastic quadriparesis: A rare case reportC. S. Gandhoke, S. K. Syal¹, D. Singh, V. Batra², Y. Nallacheruvu²Departments of Neurosurgery and ²Pathology, Maulana Azad Medical College, Lok Nayak Jai Prakash Narayan Hospital, Guru Nanak Eye Centre and G. B. Pant Institute of Postgraduate Medical Education and Research (G.I.P.M.E.R.), New Delhi, ¹Department of Paediatrics, Sri Guru Ram Das Institute of Medical Sciences and Research, Amritsar, Punjab, IndiaE-mail: *C. S. Gandhoke - charandeepsingh2008@gmail.com; S. K. Syal - simransyal100@gmail.com; D. Singh - drdaljit@hotmail.com; V. Batra - vvbatra9@gmail.com; Y. Nallacheruvu - sswini@gmail.com

*Corresponding author

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Abstract**Background:** Spinal schwannomas are slow growing, benign nerve sheath tumors. These may be asymptomatic or may present as backache with radicular pain, slowly progressive neurological deficits, but rarely with acute spastic quadriparesis attributed to intratumoral hemorrhage.**Case Description:** A 38-year-old male presented with the chief complaint of neck pain radiating to the left upper extremity for the last 8 months. On admission, he exhibited diffuse hyper-reflexia but had no motor or sensory deficit. Magnetic resonance imaging showed a solid-cystic intradural extramedullary (IDEM) C2 to C4 mass severely compressing the spinal cord. The same day the patient acutely developed a spastic quadriparesis. Immediately, a partial C2, C3, and C4 laminectomy was performed for tumor excision; within 5 postoperative days, he fully regained neurological function. The final histopathology was consistent with a "schwannoma showing areas of congestion and hemorrhage."**Conclusion:** Spinal schwannomas rarely present with intratumoral hemorrhage and acute spastic quadriparesis. Immediate operative decompression may lead to excellent postoperative neurological recovery.**Key Words:** Cervical, intratumoral, hemorrhage, quadriparesis, schwannoma

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**INTRODUCTION**

Spinal schwannomas are slow growing, benign nerve sheath tumors. These may be asymptomatic, present as backache/radicular pain, or slowly progressive neurological deficits. Patients rarely present with acute spastic quadriparesis attributed to acute intratumoral hemorrhage.

CASE REPORT

We report the case of a 38-year-old male who presented with neck pain radiating to the left upper extremity

for the last 8 months. On admission, he exhibited isolated hyper-reflexia without a motor or sensory

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deficit. The cervical magnetic resonance imaging (MRI) showed on T1, a hypointense, and on T2, a heterogeneously hyperintense solid-cystic intradural extramedullary (IDEM) C2 to C4 mass severely compressing the spinal cord; that heterogeneously enhanced with contrast [Figure 1]. The same day the patient acutely developed severe neck pain and an acute spastic quadriplegia (power was 2/5 in all 4 limbs; C5 sensory level). The patient immediately underwent a partial C2, C3, and C4 laminectomy for tumor excision; the IDEM mass exhibited intratumoral hemorrhage [Figure 2]. *En bloc* removal of the mass was performed and required resection of the dorsal nerve root to which it was attached [Video 1].

The hematoxylin and eosin sections showed numerous congested and dilated blood vessels, with both hypercellular Antoni A and hypocellular Antoni B areas composed of spindle cells. Immunohistochemistry was also positive for S100 protein, leading to the final histopathological diagnosis of “schwannoma showing areas of congestion and hemorrhage” [Figure 3]. The patient recovered well and was discharged on the 5th postoperative day with no motor or sensory deficits.

DISCUSSION

Schwannomas are typically round or oval, lobulated, and encapsulated tumors arising eccentrically from a nerve root sheath. They are most commonly intradural extramedullary (58%) in location, followed by extradural (27%), dumb-bell shaped (15% both intradural/extradural), and rarely have intramedullary (less than 1%) spinal involvement.^[11] Patients usually present in 4th or 5th decades of life.

On MRI, spinal schwannomas are typically hypointense on T1; on T2-weighted studies, they may be hyperintense and/or heterogeneous with focal areas of hyperintensity/hypointensity corresponding to cyst formation, hemorrhage, dense cellularity, and/or collagen deposition.^[4,5] With contrast they may exhibit homogeneous or patchy/inhomogeneous enhancement; the latter is more common in spinal schwannomas with cystic or necrotic components and/or intratumoral hemorrhage.^[4,5,10]

Intratumoral hemorrhages in spinal schwannomas

Intratumoral hemorrhages are occasionally reported with cranial schwannomas, but rarely with spinal schwannomas. There are various theories regarding the etiology of these acute intratumoral bleeds. According to the vascular theory, hyalinized ectatic vessels inside the schwannoma may undergo spontaneous thrombosis resulting in distal necrosis and hemorrhage.^[8] The mechanical theory is that traction on the tumor vasculature during movement may lead to disruption of blood vessels and hemorrhages.^[2,3,7,8]

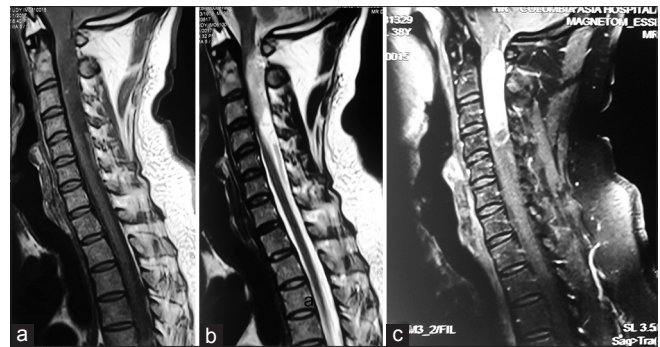


Figure 1: MRI cervical spine suggestive of (a) T1 hypointense (b) T2 heterogeneously hyperintense solid - cystic intradural extramedullary (IDEM) C2 to C4 mass severely compressing the spinal cord and pushing it anteriorly and to the right. (c) The lesion shows heterogeneous post contrast enhancement.

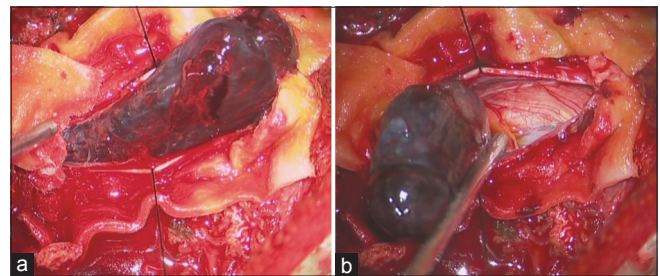


Figure 2: (a) Intra-operative photograph showing cervical schwannoma with intratumoral bleed. (b) The lesion is causing severe compression of the spinal cord

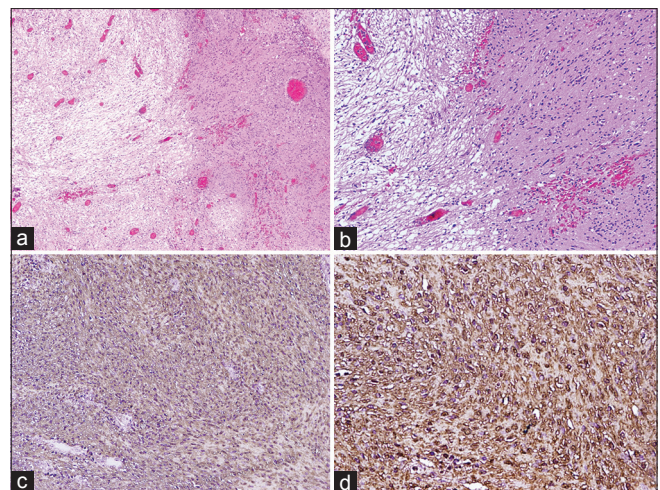


Figure 3: (a) Hematoxylin and eosin 40x magnification: Low power view of the tumor showing numerous congested and dilated blood vessels. (b) Hematoxylin and eosin 200x magnification: Both hypocellular and hypercellular areas of schwannoma seen with the hypercellular areas showing the tumour to be composed of spindle cells. (c and d) S100 staining, 100x and 200x magnification respectively: S100 stain at low and high power respectively showing cytoplasmic positivity in tumour cells proving the neuronal origin of the cells

Other causes of hemorrhage include central ischemic necrosis associated with tumor growth and/or malignant transformation with neovascularisation.^[1,9]

Treatment of acute spinal intratumoral hemorrhages

Treatment options for acute intratumoral hemorrhages in spinal schwannomas include complete excision of the tumor and its capsule. Early diagnosis and emergent removal of these lesions may lead to partial/full resolution of the acute neurological deficits.^[6] Here, the patient acutely presented with a quadriparesis of several hours duration that fully resolved within 5 days of intraspinal C2-C4 tumor removal.

CONCLUSION

Spinal schwannomas rarely present with intratumoral hemorrhage leading to acute spastic quadriparesis. In this setting, immediate operative intervention may lead to complete neurological recovery.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their 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.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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