

Case Report

# Acute hemorrhage within intradural extramedullary schwannoma in cervical spine presenting with quadriparesis

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Abstract

Schwannoma with acute hemorrhage is rarely seen. A 44-years-old male patient presented with complaint of neck pain and acute onset of quadriparesis. Magnetic resonance imaging (MRI) of his cervical spine revealed evidence of an intradural extramedullary tumor with intratumoral acute hemorrhage. He was operated in emergency and the mass was found to be schwannoma with acute hemorrhage. Post operatively the patient improved significantly. Though schwannomas show microscopic intratumoral hemorrhage and necrosis at times, schwannoma with acute hemorrhage resulting acute onset of neurological deficit is very uncommon.

**Key words:** Hemorrhage, intradural extramedullary, schwannoma

INTRODUCTION

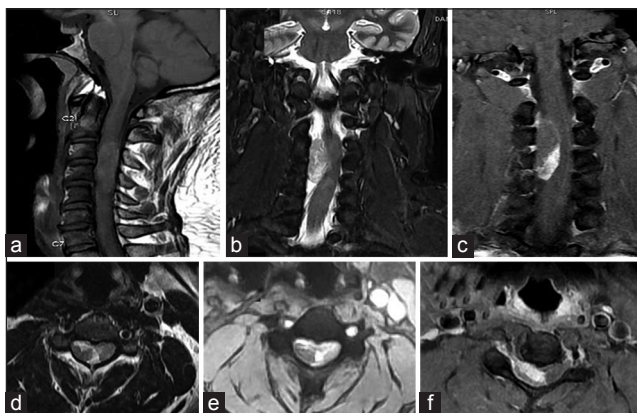
Spinal schwannomas are slow growing benign tumors of spinal nerve sheath and are diagnosed incidentally on imaging or after causing symptoms such as back pain and progressive neurological deficit. <sup>[1]</sup> These tumors are seen commonly in intradural extramedullary location and in fourth to sixth decade of life. They rarely presents with acute emergency. <sup>[2]</sup> We report a case of cervical intradural extramedullary schwannoma with acute intratumoral hemorrhage in an adult male patient who had acute onset of quadriparesis.

CASE REPORT

A 44-years-old male patient, teacher by profession, attended the neurosurgery outpatient department with complaint of

weakness in both upper and lower limbs for one day and neck pain radiating to shoulder for two days duration. He was nonsmoker and non-alcoholic and was not suffering from diabetes, tuberculosis, hypertension or any chronic illness before. No history of drug abuse or trauma was present. On clinical examination, his vitals were within normal limit. Higher functions and cranial nerve examination were normal. Motor examination revealed grade 3/5 power in left side limbs, grade 4/5 powers in right side limbs, weak bilateral hand grip, exaggerated tendon reflex on left side limbs, and bilateral extensor plantar reflex. Bowel and bladder function was normal. Sensory examination showed decrease pain sensation on the left side limbs and decrease touch sensation on the right side limbs. There were no skin and subcutaneous nodules or *café au let* spot. Clinically cervical myelopathy was suspected and magnetic resonance imaging (MRI) of cervical spine was advised. Contrast-enhanced MRI of cervical spine revealed a lobulated enhancing intradural extramedullary mass of size about 35 × 13 mm on right posterolateral aspect of spinal cord from C3 to C5 vertebral body levels showing intratumoral macroscopic hemorrhage [Figure 1]. Radiological diagnosis was nerve sheath tumor with intratumoral hematoma and necrosis. Patient was operated on emergency. Laminectomy from C3 to C4 was

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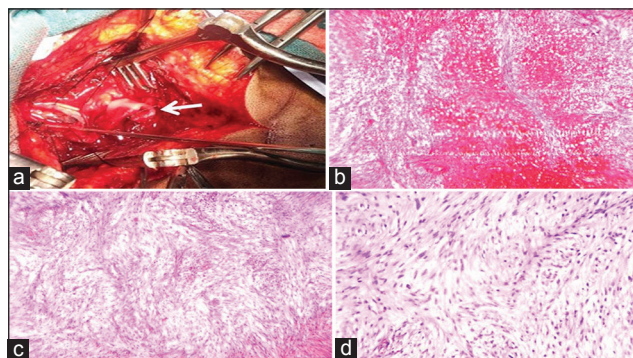
**Figure 1:** The mass shows T1W hetero hyperintense (a), T2W iso to hyperintense (b and d), hyperintense and hypointense signal intensity in gradient image (e), no enhancement of hemorrhagic component and patchy enhancement of non-hemorrhagic component (c and f)

done. After opening the dura, a chocolate colored lobulated mass was seen on posterolateral aspect of spinal cord. Under operating microscope, the arachnoid mater was cut open and the tumors bulged out. Intratumoral clots are removed. The mass was excised along with its capsule [Figure 2a]. Wound was closed in layers. Post operative period was uneventful. His motor and sensory functions were normal on third post operative day. Histopathological examination of the excised mass confirmed schwannoma with intratumoral macroscopic hemorrhage [Figure 2b-d]. The patient was asymptomatic on follow-up.

## DISCUSSION

Peripheral nerve sheath tumors comprise neurofibroma and schwannoma. Schwannoma, neurinoma, and neurilemoma are synonyms. Schwannomas and neurofibromas are different entities. Schwannomas generally are solitary, do not envelop the adjacent nerve root, which usually is the dorsal sensory root. In contrast, neurofibromas envelop the dorsal sensory root, frequently are multiple, and usually are associated with neurofibromatosis, even when single.<sup>[3]</sup> Schwannoma along with neurofibroma are most common intraspinal lesions, representing 16–30% of all intraspinal masses and are commonly seen in the fourth decade of life. They are most commonly intradural extramedullary in location (58%) followed by extradural (27%), dumbbell shaped with both an extradural and an intradural component (15%), and, rarely, intramedullary (less than 1%). In children, most common location is in the cervical region, followed by the lumbar and thoracic regions.<sup>[4]</sup>

The clinical presentation of intraspinal schwannoma depends on the size and location of the tumor. The clinical symptoms and sign are vague due to slow growth of the tumor. Conti and colleagues<sup>[5]</sup> demonstrated an average duration of symptoms for schwannomas of 55 months, with a range of 1-360 months. The most common presurgical symptom is



**Figure 2:** The intraoperative picture (a) shows the schwannoma with intratumoral hemorrhage (arrow mark). Histopathological study of the mass shows macro hemorrhage within the mass (b) cellular area (Antoni A) and loose myxoid area (Antoni B) in low (c) and high magnification (d) image

segmental pain, followed by radicular pain and variable motor deficits.

Schwannomas are well-circumscribed encapsulated lobulated masses attached to periphery of the nerve and may have areas of cystic and xanthomatous change. On microscopic examination, tumors show a mixture of two growth patterns such as the Antoni A pattern and Antoni B pattern. In Antoni A pattern, elongated cells with cytoplasmic processes are arranged in fascicles in areas of moderate to high cellularity with little stromal matrix. In the Antoni B pattern of growth, the tumor is less densely cellular with a loose meshwork of cells along with microcysts and myxoid changes. Axons are largely excluded from the tumor.<sup>[6]</sup> Acute macro hemorrhage can occur inside schwannoma rarely. Exact mechanism of acute intratumoral hemorrhage is not known. One hypothesis suggests that hyalinized ectatic vessels inside schwannoma undergo spontaneous thrombosis resulting distal necrosis and hemorrhage. Second theory supports the traction of tumor vasculature during movement.<sup>[7]</sup> Trauma or history of anticoagulant intake may cause intratumoral acute hemorrhage. In our case there was no history of trauma or anticoagulant therapy. The cause of hemorrhage can be explained by the first theory. Intradural extramedullary schwannoma may cause intraspinal subdural hemorrhage and intraspinal subarachnoid hemorrhage.<sup>[7]</sup>

Posterior scalloping of the vertebral bodies and widening of the neural foramina are the common radiological findings in plain radiograph study of spine. The schwannomas show decreased attenuation in computed tomography of spine and are well differentiated from spinal cord and thecal sac in computed tomography myelogram study. On MRI of spine, schwannomas have typical increased signal intensity than muscle in T1-weighted image, markedly hyperintense (due to high water content of the lesion) in T2-weighted sequence and shows intense homogeneous enhancement in post contrast study.<sup>[4]</sup> Sometimes they shows inhomogeneous T2-weighted signal with areas of hyperintense and hypointense signal corresponding to cyst formation, hemorrhage or dense collagen deposition.<sup>[8]</sup> Patchy enhancement is

seen in schwannoma with hemorrhage and or necrosis.<sup>[9]</sup> Intratumoral hemorrhage inside schwannoma shows different signal in MRI depending on the ages of blood product. Acute hemorrhage shows hyperdense attenuation in computed tomography (CT) scan image. MRI shows T1W isointense, T2W hypointense signal in acute hemorrhage, T1W hyperintense and T2W hypointense signal in early subacute hemorrhage and T1/ T2W hyperintense signal in late subacute hemorrhage and T1/T2W hypointense signal in chronic hemorrhage.<sup>[10]</sup> In our case the intra tumoral hematoma was late subacute in nature.

Radiological differentials of intradural extramedullary schwannoma are neurofibroma and meningioma. Schwannoma are usually encapsulated (70%) and attached to periphery of nerve while neurofibroma surrounds the nerve fibre and uncommonly encapsulated (30%).<sup>[11]</sup> Meningioma shows often calcification and dural tail sign unlike schwannoma while schwannoma shows more heterogeneous hyperintense signal on T2W image than meningioma.<sup>[12]</sup>

The complete excision of the tumor along with capsule prevents recurrence. Early diagnosis and emergency removal of hemorrhagic schwannoma are factor for successful post surgical outcome.<sup>[13]</sup> In our case, the patient showed marked improvement of symptoms due to acute presentation, early diagnosis and operation.

## CONCLUSION

The intradural extramedullary schwannoma with acute intratumoral hemorrhage presents early due to rapid onset of neurological deficit and is considered as neurosurgical emergency. Contrast enhanced MRI of spine is the investigation of choice for intraspinal tumors. Complete excision of hemorrhagic tumor prevents recurrence rate.

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