

Review Article

Hemorrhage in long segment cervical schwannoma; case report and literature review

Prashant Raj Singh¹, Nitish Nayak¹, Surendra Kumar Gupta¹, Raghavendra Kumar Sharma¹, Anju Shukla², Lokesh Suresh Nehete¹

¹Department of Neurosurgery, All India Institute of Medical Sciences, Raipur, Chhattisgarh, ²Department of Laboratory Medicine, Sahara Hospital, Lucknow, Uttar Pradesh, India.

E-mail: Prashant Raj Singh - prashantsingh2010@yahoo.com; Nitish Nayak - drnitishnayak@gmail.com; Surendra Kumar Gupta - drsurendrakrgupta@gmail.com; Raghavendra Kumar Sharma - sdr.raghavendra@gmail.com; Anju Shukla - anjushukla@gmail.com; *Lokesh Suresh Nehete - neloc23@gmail.com



*Corresponding author:

Lokesh Suresh Nehete,
Department of Neurosurgery,
All India Institute of Medical
Sciences, Raipur, Chhattisgarh,
India.

neloc23@gmail.com

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ABSTRACT

Background: Although hemorrhages associated with cervical and thoracic intraspinal schwannomas are typically localized to the subarachnoid hemorrhages (SAH) or subdural hemorrhages (SDH) compartments, rare intratumoral bleeds may also occur.

Methods: In the literature, we found and analyzed multiple factors for 13 cases (e.g., epidemiological, clinical, and pathological) of cervical schwannomas with intratumoral hemorrhages (ITH). We added the 14th case of a 35-year-old female with long segment cervical schwannoma with ITH who presented with acute quadriplegia and respiratory decompensation.

Results: These 14 patients averaged 51.77 years of age, 60% were male, and the tumor involved 2.83 segments. The incidence of SAH and ITH was noted in five cases each, while SDH's were very rare. The pathological characteristics were consistent with the diagnosis of cellular schwannomas with S-100 positivity. The clinical outcomes were good (100%) in all the cases, including the one presented (modified McCormick score III).

Conclusion: Cervical schwannomas with ITH are rare, and the surgical outcomes in such patients are good-excellent (>90%). The histopathology is always of prime importance and decisive in establishing and confirming the etiology of such ITH.

Keywords: Cellular schwannoma, Intratumoral bleed, Schwannoma with bleed

INTRODUCTION

Schwannomas are some of the most common peripheral nerve sheath tumors.^[1] They are classified, and outcomes/prognosis are closely tied to tumor size and location.^[3,7] Spinal schwannomas very rarely present with acute neurological deterioration secondary to intratumoral hemorrhages (ITH); other hemorrhagic sites include tumor-related subarachnoid hemorrhages (SAH) or subdural hemorrhages (SDH). Here, we present review of 13 cases of ITH attributed to cervical spinal schwannoma in the literature and have added our 14th cases to the series.

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MATERIALS AND METHODS

We utilized (2020) two electronic databases (e.g., U.S. National Library of Medicine, National Institutes of Health [PubMed], and EMBASE) to identify studies of cervical schwannoma with ITH [Table 1].

Literature review

We reviewed 13 cases from 13 articles regarding cervical ITHs attributed to spinal schwannomas, exclusive of our case. The following variables were studied: age, gender, predisposing factor, clinical presentation, size of the tumor, level of the tumor, hemorrhages, pathological tumor features, and outcomes (e.g., using the modified McCormick score).

Of the 13 previously reported cases of cervical schwannomas hemorrhages, spontaneous ITH were noted in five cases (38%), SAH in five cases (38%), and SDH with intratumoral bleed in two cases (15%), with one isolated case of SDH alone [Tables 1, 2]. Patients averaged 51.77 years of age; our patient was 35 years old. There were eight males (61%) and five (39%) females; our patient was a female. Only four cases (30%) had attendant trauma/spinal manipulation as predisposing factors to the hemorrhage. Tumors involved an average of 2.83 levels. Histopathological findings in six cases showed a predominant Antoni A pattern, one case had an Antoni B pattern, while four had mixed patterns as in our case; in two cases, no pathological features were discussed.

Outcomes were analyzed with the McCormick score; 12–13 cases showed significant improvement, while the outcome was not reviewed in one case.

Case report

Case illustration

A 35-year-old female were presented with the sudden onset of quadriparesis over 1 week duration and 2 days of increased respiratory distress. On examination, she had a flaccid quadriparesis (motor 1/5 both upper extremities and 0/5 in both lower extremities) with a C7 sensory level and acute urinary retention. A contrast holo-neuraxis MRI showed a T1 hypointense, T2 heterogeneously hyperintense (e.g., with peripheral irregularity), and a non-enhancing central-large intradural extramedullary lesion from C2-T2 [Figure 1a-c]. Axial images showed extension into the right C2 neural foramen with widening/displacement of the spinal cord toward the left (e.g., suggestive of type III giant spinal schwannoma-Sridhar classification).^[3] Gradient-recalled echo T2-weighted images additionally documented “blooming” in the lesion’s central region suggestive of blood degradation products.

Surgery

The patient underwent a C3-T3 Laminoplasty. After opening the dura, a giant dark brown subarachnoid mass

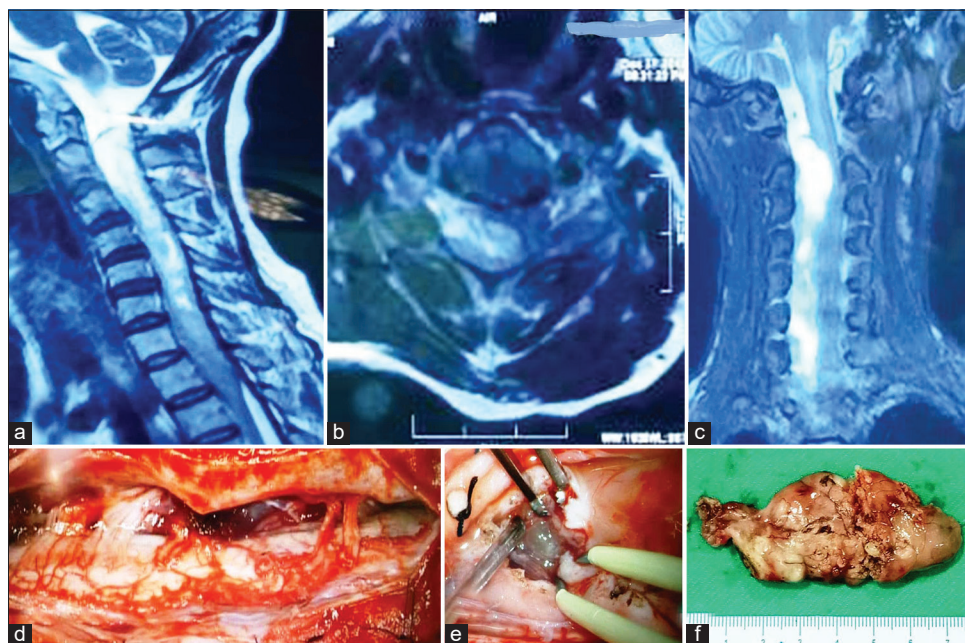


Figure 1: T2-weighted images of magnetic resonance imaging in sagittal (a), axial (b), and coronal (c) sections – a large mixed intensity intradural extramedullary spinal cord tumor from C2-D2 location and with significant compression of the spinal cord. A large brownish tumor anterolateral to the cord and pushing it to the left side (d), with intratumoral blood clots (e), and completely excised tumor (f).

Table 1: Previous reported cases of cervical schwannoma with bleed.

Reported case study	Age/sex	Presentation	Predisposing factor	Location of bleed	MRI level of lesion	Surgery	Specific pathological feature	Follow up Duration	Modified Mc Cormick Grade Preoperative	Postoperative
Luxon and Harrison, 1978	67 years/ Male	Sudden onset pain in tip of shoulder and occipital region	Mechanical strain	SAH	C3-C4	C3-C5 Laminectomy and excision	Not mentioned	Not mentioned	No deficit	No deficit
Smith <i>et al.</i> , 1985	74 years/ Female	Rapid progressive quadriplegia	No	SDH, Intratumoral	C3-C5	C4-T1 Laminectomy and excision	Not mentioned	Died(at 5 months- peritonitis)	V	III
De Divitiis <i>et al.</i> , 1985	72 years/ Female	Neck and radicular pain	No	SAH	C4-C7	C3-D2 laminectomy and excision	Schwannoma (Antoni type A predominance)	Not mentioned	III	II
Chalif <i>et al.</i> , 1990	56 years/ Female	Paraparesis, Sudden onset headache	No	SAH	C1-C2	C1C2 laminectomy and excision	Hemorrhagic schwannoma	Not mentioned	No deficit	No deficit
Mills <i>et al.</i> , 1993	53 years/ Male	Sudden onset scapular pain	No	SAH	C7-T1	C6 - T1 laminectomy and excision	Ancient schwannoma (Degenerative nuclear atypia without mitosis), S-100 positivity	mentioned 6 months	IV	II
Corriero <i>et al.</i> , 1996	37 years/ Male	Sudden onset severe headache	After physical stress	SAH	C7-T1	C6-T1 laminectomy and excision	Telangiectatic schwannoma, Antoni type B, S-100 positivity	2 months	No deficit	No deficit
Ng, 2001	43 years/ Male	Neck and radicular Quadripareisis	No	SDH, Intratumoral	C5-C7	C5-C7 Partial laminectomy and excision	Mixed Antoni A and B pattern, ectatic hyalinized vessels	3 months	IV	II
Ciappetta <i>et al.</i> , 2008	44 years/ Female	Sudden onset Quadripareisis, XI cranial nerve weakness	No	Intratumoral bleed	divus to C5	Far-lateral approach with laminectomy and excision	Compact cellular Antoni A pattern, S-100 positivity	6 months	IV	II
Vazquez-Barquero <i>et al.</i> , 2009	68 years/ Male	Rapidly progressive paraparesis.	No	SDH	C5 to C7	C4-T1 Hemilaminectomy and excision	Compact Antoni A pattern, S-100 positivity	2 months	V	II
Mohanty <i>et al.</i> , 2015	44 years/ Male	Neck pain and weakness in all four limb	No	Intratumoral	C3 to C4	C3-C4 laminectomy and excision	Mixed Antoni A and B pattern	Not mentioned	Not mentioned	Not mentioned
Prasad <i>et al.</i> , 2015	40 years/ Male	Flaccid paraplegia	Minor fall	Intratumoral	C7-D3	C7-D3 laminectomy and excision	Mixed Antoni A and B pattern	6 months	V	II
Gandhoke <i>et al.</i> , 2018	38 years/ Female	Acute onset quadripareisis	No	Intratumoral	C2-C4	C2-C4 laminectomy and excision	Mixed Antoni A and B pattern, S-100 positivity	At discharge	IV	II
Jung <i>et al.</i> , 2019	37 years/ Male	Acute onset quadripareisis	Physiotherapy	Intratumoral	C2-C3	C2-C3 hemilaminectomy and excision	Cellular schwannoma (ectatic hyalinized vessels, degenerative changes)	3 weeks	IV	II
Present case	35 years/ Female	Acute onset quadripareisis	No	Intratumoral	C2-D2	C3 -D2 Laminoplasty and excision	Mixed Antoni A and B pattern, degenerative nuclear atypia without mitosis, S-100 positivity.	At discharge	V	III

was found attached to the right C2 nerve root and the spinal cord was displaced toward the left. The tumor was

soft in consistency and accompanied by a lobulated dark-brown hemorrhage (e.g., with the altered thickness of blood clots of different ages). The tumor was easily suckable, and a good cleavage plane could develop circumferentially around the tumor, which was approximately 7.5 cm in size [Figure 1d-f]. The tumor was first internally decompressed under intraoperative monitoring in a piecemeal manner using the cavitron ultrasonic aspirator, finally facilitating gross total, *en bloc* removal.

Histology

The histological examination revealed a cellular schwannoma with mixed Antoni A and Antoni B cells and degenerative atypia; however, mitotic activity was not discernible [Figure 2a-d]. There were areas of hemorrhage and fibrin deposition. Sections were further subjected to immunohistochemistry, which showed strong and diffuse staining for S 100 [Figure 2e], while the EMA was negative [Figure 2f].

Postoperative course

The postoperative course was uneventful as the patient's status improved; motor power became 3/5 in both upper and lower extremities, and her respiratory distress regressed. By the time of discharge, she could walk with one person's support (e.g., motor power still 3–4/5). Her neurological status remained mostly unchanged at the 3 months follow-up (modified McCormick score III).

Table 2: Characteristics of study cohort of previous reported cases.

Patient	Results
Average age	51.77 years
Range	37–74 years
Sex distribution	
Male	8 (61%)
Female	5 (39%)
Tumor level of involvement (average)	2.83
Range	2–6 vertebral levels
Hemorrhage	
SAH	5 (38%)
SDH	1 (9.09%)
SDH with intratumoral bleed	(15.4%)
ITH	5 (38%)
Predisposing factor (only physical stress/spinal manipulation)	
Yes	4 (30%)
No	9 (70%)
Histopathological findings	
Antoni A pattern	6 (46.2%)
Antoni B pattern	1 (9.09%)
Mixed pattern (both)	4 (30.76%)
Not mentioned	2 (15.4%)
Outcome (modified McCormick grade)	
Excellent (I-III)	12 (92.3%)
Not mentioned	1 (9.09%)

SAH: Subarachnoid hemorrhage, SDH: Subdural hemorrhage, ITH: Intratumoral hemorrhage

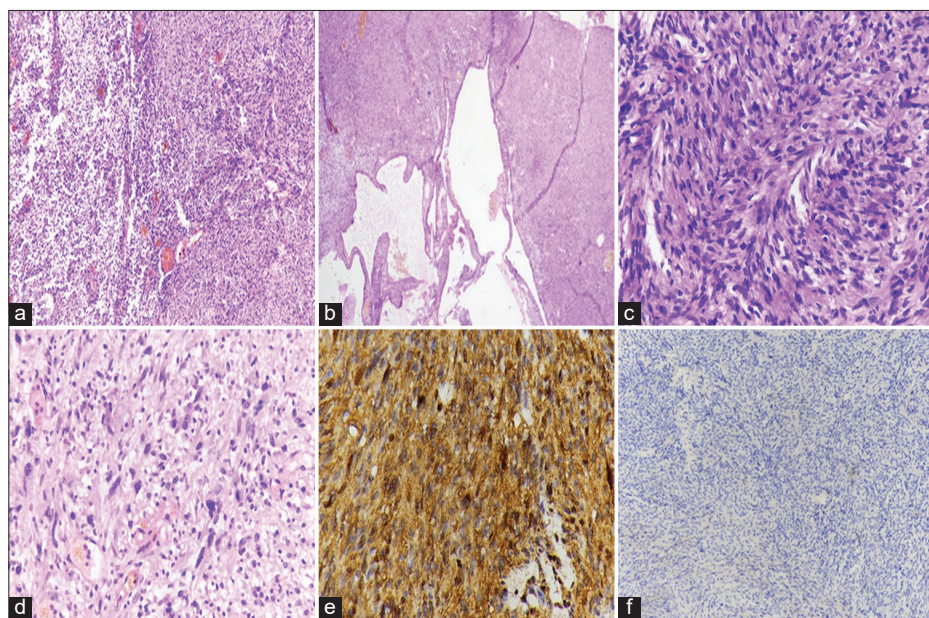


Figure 2: Illustrated as – (a) Sections showing hypercellular Antoni Type A and hypocellular Antoni Type B areas (H and E, $\times 10$), (b) with interspersed dilated, irregular shaped blood vessels (H and E, $\times 4$), (c) cells are having oval to spindle shape nuclei, fibrillary cytoplasm with minimal atypia (H and E, $\times 40$), (d) degenerative changes with smudgy chromatin, however, mitotic activity is not seen (H and E, $\times 40$), (e) immunohistochemistry showing diffuse and strong S100 positivity and (f) While EMA is negative.

DISCUSSION

ITH occurring within the cervical spinal are very rare. We reviewed 13 prior cervical schwannomas, five of which were ITH. Of interest, cervical schwannomas equally present with ITH and SAH, while SDHs are rare. Outcomes of ITH within cervical schwannomas appear independent of size, age, or type of bleed, and preoperative neurological status.

The mechanism of hemorrhage in such tumors is variously reported as attributed to; ectatic hyalinized vessels undergoing spontaneous thrombosis^[5] distal tumor necrosis, hemorrhage, endothelial proliferation in vascular lesions, and/or recanalization by meningeal vessels within the necrotic tumor, or traction along the spinal axis (e.g., on nerve roots' vascular attachments).^[2,3,5] The pathological factors have also been studied previously with different names as ancient schwannoma to cellular schwannoma, which has explained the degenerative and hemorrhagic nature of certain pathologically distinct schwannomas.^[3-6]

CONCLUSION

The suspicion of an ITH within a cervical schwannoma must be considered when patients develop the acute onset of quadriplegia/plegia without an attendant history of coagulopathy, hypertension, or spinal manipulation/trauma. The histopathology is the final determinant of the lesion.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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