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

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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.[1,7] 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.
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...
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
|---------------------|---------|--------------|---------------------|------------------|---------------------|---------|-----------------|-----------------|----------------------|
| Luxon and Harrison, 1978 | 67 years/ Male | Sudden onset pain in tip of shoulder and occipital region | Mechanical strain | SAH | C3-C4 | C3-C5 Laminection and excision | Not mentioned | Not mentioned | No deficit | No deficit |
| Smith et al., 1985 | 74 years/ Female | Rapid progressive quadriparesis Neck and radicular pain | No | SDH, Intratumoral | C3-C5 | C4-T1 Laminection and excision | Not mentioned | Died(at 5 months- peritonitis) | V | III |
| De Divitiis et al., 1985 | 72 years/ Female | Sudden onset headache | No | SAH | C4-C7 | Schwannoma (Antoni type A predominance) | Not mentioned | Not mentioned | III | II |
| Chalf et al., 1990 | 56 years/ Female | Sudden onset scapular pain | No | SAH | C1-C2 | C1C2 laminection and excision | Not mentioned | No deficit | No deficit | No deficit |
| Mills et al., 1993 | 53 years/ Male | Sudden onset headache | No | SAH | C7-T1 | C6-T1 laminection and excision | Not mentioned | No deficit | No deficit | No deficit |
| Corriero et al., 1996 | 37 years/ Male | Sudden onset severe headache | After physical stress | SAH | C7-T1 | C6-T1 laminection and excision | Telangiectactic schwannoma, Antoni type B, S-100 positivity | 2 months | No deficit | No deficit |
| Ng, 2001 | 43 years/ Male | Neck and radicular pain | No | SDH, Intratumoral | C5-C7 | C5-C7 Partial laminection and excision | Mixed Antoni A and B pattern, ectatic hyalinized vessels | 3 months | IV | II |
| Ciappetta et al., 2008 | 44 years/ Female | Quadriparesis Sudden onset of quadriparesis, XI cranial nerve weakness | No | Intratumoral bleed | clivus to C5 | Far-lateral approach with laminectomy and excision | Compact cellular Antoni A pattern, S-100 positivity | 6 months | IV | II |
| Vazquez-Barquero et al., 2009 | 68 years/ Male | Rapidly progressive quadriparesis | No | SDH | C5 to C7 | C4-T1 Hemilaminection and excision | Compact Antoni A pattern, S-100 positivity | 2 months | V | II |
| Mohanty et al., 2015 | 44 years/ Male | Neck pain and weakness in all four limb | No | Intratumoral | C3 to C4 | C3-C4 laminection and excision | Mixed Antoni A and B pattern, S-100 positivity | Not mentioned | Not mentioned | Not mentioned |
| Prasad et al., 2015 | 40 years/ Male | Flaccid paraplegia | Minor fall | Intratumoral | C7-D3 | C7-D3 laminection and excision | Mixed Antoni A and B pattern | 6 months | V | II |
| Gandhoke et al., 2018 | 38 years/ Female | Acute onset quadriparesis | No | Intratumoral | C2-C4 | C2-C4 laminection and excision | Mixed Antoni A and B pattern, S-100 positivity | At discharge | IV | II |
| Jung et al., 2019 | 37 years/ Male | Acute onset quadriparesis | Physiotherapy | Intratumoral | C2-C3 | C2-C3 hemilaminection and excision | Cellular schwannoma (ectatic hyalinized vessels, degenerative changes) | 3 weeks | IV | II |
| Present case | 35 years/ Female | Acute onset quadriparesis | 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).

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**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

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**Figure 2:** Illustrated as – (a) Sections showing hypercellular Antoni Type A and hypocellular Antoni Type B areas (H and E, ×10), (b) with interspersed dilated, irregular shaped blood vessels (H and E, ×4), (c) cells are having oval to spindle shape nuclei, fibrillary cytoplasm with minimal atypia (H and E, ×40), (d) degenerative changes with smudgy chromatin, however, mitotic activity is not seen (H and E, ×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, 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). 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.

CONCLUSION

The suspicion of an ITH within a cervical schwannoma must be considered when patients develop the acute onset of quadriparesis/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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