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Case Report

Giant Ventral Midline Schwannoma of Cervical Spine : Agonies and Nuances

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Pure ventral midline giant schwannoma is an extremely rare entity. Spinal intradural extramedullary schwannomas commonly occur posterolateral or anterolateral to the spinal cord. A case of a pure midline ventrally situated giant pan cervical extramedullary schwannoma in an 18-year-old male patient with compressive myelopathy and sphincter involvement is presented. Spinal MR imaging showed a midline ventrally situated extramedullary tumor with severe spinal cord compression extending from clivus to C7 vertebra. It was resected through a posterolateral approach. Histology was consistent with a schwannoma. Post operative MR imaging showed no evidence of the tumor. The radiological features, pathogenesis and surgical strategies in management of these difficult tumors are discussed and the relevant literature is briefly reviewed.

KEY WORDS : Extramedullary · Intradural · Giant schwannoma · Spine · Ventral location.

INTRODUCTION

Schwannomas are common benign intradural extramedullary spinal neoplasms[6]. These usually present with subacute to chronic compressive myelopathy. Modern magnetic resonance (MR) imaging has helped immensely in their accurate preoperative identification and pathological diagnosis[7]. Advanced instrumentation and microsurgical techniques have made surgical excision safe and radical. Surgical excision of midline ventral long segment intradural tumors can be formidable and can have potentially serious morbidity[19]. An appropriate surgical approach and strategy needs to be outlined to achieve a good outcome.

CASE REPORT

An 18-year-old male presented with progressively worsening neck pain and paraesthesias in both upper limbs for the past 3 years. He had progressive stiffness of both lower limbs, difficulty in walking and straining while micturition and constipation for the past 2 months. He required support while walking. There was no history of trauma, fever or tuberculosis. There were no neurocutaneus markers. There was weakness of spinal accessory nerves bilaterally in form of paresis of trapezius and sternomastoids. On examination, he had spastic quadriparesis of grade 4. Patient had hypertonia and hyperreflexia of all limbs. Plantars were extensor. MRI of the craniovertebral region and cervical spine revealed a midline ventral tumor extending from lower clivus to C7 vertebra. It was hypointense on T1 and hyperintense on T2-weighted image with irregular thick peripheral enhancement (Fig. 1). The spinal cord was severely compressed and displaced posteriorly. A C1-C7 laminotomy was performed. All lateral masses were preserved. The tumor was midline ventral and the spinal cord was stretched out like a ribbon. The ligamenta denticula were cut bilaterally. All roots were safeguarded. The tumor was yellowish-grey, lobulated, vascular and had a fleshy capsule and suckable. The dissection was commenced from C7 and carried upwards. The tumor was adherent to the ventral surface of the spinal cord in lower part but no definite attachment of the tumor to any nerve root could be found. A radical excision was achieved. The cord was seen to pulsate well at the end. Histological examination revealed features of schwannoma with extensive degenerative change (Fig. 2). Immediately following surgery, spasticity and paresis of all four

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limbs improved. Sphincteric complaints also showed delayed improvement. Post-operative MR showed no evidence of residual tumor (Fig. 3). At follow-up after 1 year patient is doing well.

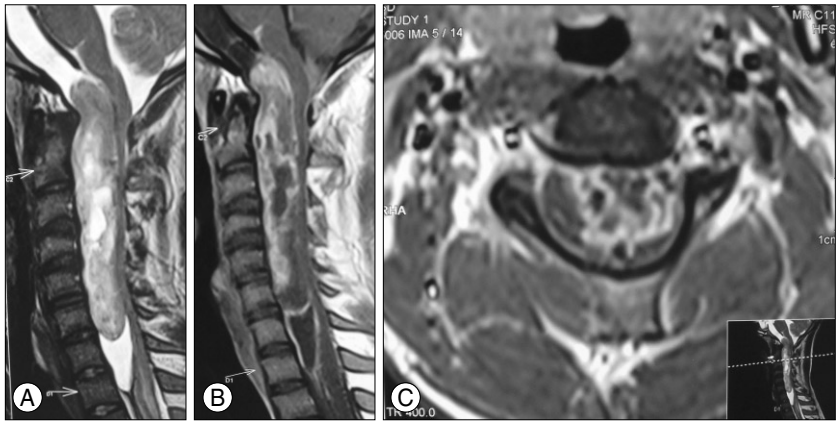

Fig. 1. MRI showing ventrally situated extramedullary schwannoma extending from lower clivus to C7. A : Sagittal T2-weighted image. B : Sagittal T1- contrast enhanced image. C : Axial T1-weighted contrast enhanced image.

DISCUSSION

Intradural spinal schwannomas are usually dorsal, lateral or dorsolateral in position[6]. However, as much as 23% of cervical nerve sheath tumors may have an anterolateral component consistent with ventral root origin[8]. Giant spinal schwannomas are defined as those that extend over more than two vertebral levels. Giant spinal schwannomas in ventral midline location are rare[10,13,15,21]. We report the case of a pure midline ventral cervicodorsal intradural extramedullary schwannoma extending from clivus to C7. The origin of schwannoma may be difficult to judge in long segment schwannomas, as observed in our case. The location of the tumor in our case suggests an alternative site of Schwann cell derivation. Multiple theories exist to explain this aberrant location for a Schwann cell-based tumor. Some authors postulate that these arise from perivascular nerve plexi surrounding penetrating spinal cord vessels from the anterior spinal artery[2,17,18]. Alternatively, rests of neural crest progenitors may migrate improperly into the central nervous system parenchyma during embryogenesis[2,12,18,20]. We hypothesize that genesis of a midline ventral schwannoma without attachment to a ventral root requires an atypically located parent Schwann cell, such as those of the nervi vasorum of the anterior spinal artery[2,17]. Singer et al.[18] have reported a ventrally placed pigmented schwannoma of the C7-T2 region arising from the leptomeninges in the region of the anterior median septum, ventral to the dentate ligaments and anterior rootlets.

Various approaches to access the ventral cervical and cervicothoracic intradural tumors including schwannomas are : 1) anterior cervical or transthoracic approaches[3,16], 2) anterolateral approach[1], 3) posterolateral approach[6,11,13], and 4) endo-assisted posterolateral approach[4]. In most cases, there is no single approach that provides adequate surgical exposure across multiple spinal levels. O'Toole and McCormick[16] described anterior corpectomy and reconstruction for solid and small schwannoma apposed to the ventral midline surface of the spinal cord producing no significant rotation of the spinal cord and therefore presenting no tumor surface for lateral surgical access. Anterior approach has certain disadvantages like 1) inadequate access

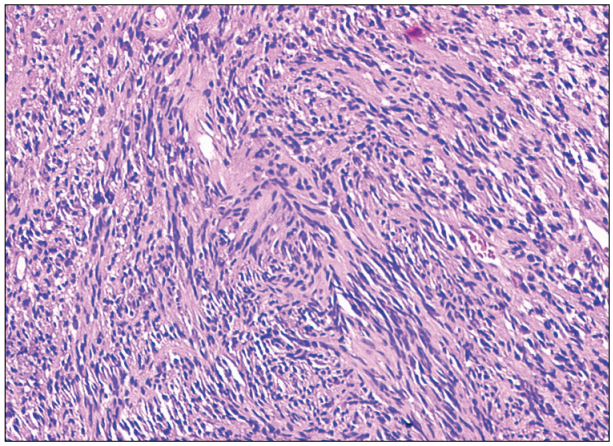

Fig. 2. Microphotograph showing densely packed spindle cells in interlocking fascicles (Antoni type A), intermingled with loosely textured tissue with extracellular clear spaces (Antoni type B). H&E, original magnification × 200.

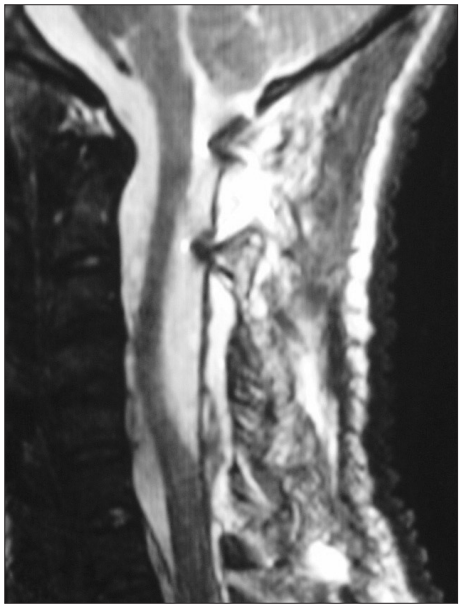

Fig. 3. Sagittal T2-weighted post-operative Modern magnetic resonance image showing no evidence of tumor.

due to deep and constrained field of view, 2) excessive bleeding from epidural venous plexus, 3) need for ensuring spinal stability and meticulous bony reconstruction, and 4) postoperative CSF fistula[16]. However, Acosta et al.[1] described anterolateral corridor using modified paramedian transpedicular approach and spinal reconstruction for ventral intradural tumors including schwannomas of the cervical and cervicothoracic spine. Acosta et al.[1] state that removal of the posterolateral elements, and the dorsal vertebral body combined with angulation of the operating table exposes the ventral-most thecal sac and creates a potential space for delivery of tumor without spinal cord retraction or rotation. This approach avoids the difficulties of anterior approach[1].

Traditionally, the approach for resection of intradural spinal tumors has been by posterior laminectomy with or without facetectomy and/or pediculectomy. This is largely a reflection of the posterior and lateral orientation of most intradural tumors. Extensive bone excision and reconstruction is avoided in this approach[13].

Barami and Dagnew[4] reported endoscopic assisted removal of ventral intradural spinal tumors through a posterior approach. They postulate that judicious use of endoscope during tumor removal allows minimal retraction of the cord and inspection of the corridors which are difficult to visualize with the microscope. We believe that an endo-assisted posterolateral approach might be the best option for such difficult tumors in near future.

We believe that these tumors form a special subset of schwannomas with different origin and presentation. These manifest with myelopathy rather than radiculopathy due to aberrant origin, therefore present after achieving giant size unlike simple schwannomas originating from roots. Literature on consistency of giant ventral schwannomas is lacking due to very rare occurrence. We have realized that giant ventral schwannomas have softer consistency due to extensive degenerative changes (might be due to relative ischemia caused by huge size) which facilitates the fall of the tumor into the vision after decompression. Surgical approach is determined primarily depending on the location of tumors in spinal canal. However, tumor consistency and pathology should be also considered. Solid ventral midline schwannomas are optimally managed by anterior and anterolateral approaches[1,3,16]. However; tumors with softer consistency may be approached from posterolateral approach with satisfactory outcome as evidenced by our case.

On preoperative MR imaging, schwannomas usually show hypointense or isointense signal on T1-weighted and hyperintense on T2-weighted images[6,7]. Homogenous enhancement is common but occasional heterogeneous enhancement revealing intratumoral hemorrhage, cyst or necrosis is seen suggesting an atypical or aggressive nature[6,7]. Meningioma and tuberculous granulomas are important differential diagnosis[5,14]. The hyperintense signal on T2-weighted MR images was suggestive of a relatively soft tumor consistency. MR imaging is helpful to indicate consistency of the tumor, thereby suitability of the posterolateral approach.

A piecemeal decompression bilaterally in between the nerve roots allowed the tumor to express into the operative field and relieved the cord compression in our case. Gentle and slow micro dissection, minimal use of cautery and no retraction of cord parenchyma are the keys to the successful surgery. The use of cautery was kept minimal to avoid thermal injury to the roots and cord. Preservation of roots and lateral masses is essential for long term good functional outcome. The upper third of the tumor was free in the subarachnoid space while the tumor in the middle and the lower third was adherent to the ventral surface of the spinal cord suggesting a caudocranial growth of the tumor.

Laminoplasty reduces the risk of post-operative kyphosis[9]. However, Kim et al.[10] have emphasized that despite of multilevel laminoplasty for huge craniocervical cord tumor the patient may develop swan neck deformity. To prevent this kind of complication, they stressed anterior cervical fixation in addition to posterior laminoplasty or laminectomy. In our patient, although there was no immediate cervical spinal instability, we are closely monitoring the patient. Long term follow up is essential for such complication.

CONCLUSION

Pure midline ventral giant schwannomas are very rarely encountered. These manifest late with myelopathy after achieving giant size due to aberrant origin and location. Surgery for these tumors is potentially formidable. A posterolateral approach with piecemeal micro dissection is feasible in such tumors. It remains unclear whether these schwannomas represent a special variant of schwannomas or an entirely distinct entity. Larger series may answer this question in the future.

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