Giant, solitary, ancient schwannoma of the cervico-thoracic spine: a case report and review of the literature

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Abstract

Introduction

Ancient schwannoma is a schwannoma subtype characterised by degeneration and diffused hypocellular areas. Advanced instrumentations, microsurgical techniques and intra-operative neurophysiological monitoring, have made tumour excision safe and radical. How-ever, in the third world where most of these are lacking, spinal tumour surgery carries a lot of risk. This is the first multisegmental, intradural tumour that we have successfully excised in our spinal unit. This paper presents a patient with giant, solitary, ancient schwannoma of the cervicothoracic spine.

Case Report

We present the case of a 29-year old Ugandan man who presented with pain, heaviness, weakness and numbness in the lower limbs. Magnetic resonance imaging showed an intradural, extramedullary tumour at spinal levels C6–T3. He underwent successful tumour excision without intra-operative neurophysiological monitoring through the posterior approach. He also had posterior instrumentation to prevent iatrogenic post-operative instability. Histopathological examination showed ancient schwannoma. The patient’s neurological status is steadily improving.

Conclusion

Giant ancient schwannomas at the cervico-thoracic spine are uncommon. These tumours can be successfully removed even in resource-limited settings as long as the technical expertise is available.

Introduction

Sridhar et al. defined giant spinal schwannomas as those that extend over more than two vertebral levels. Schwannomas are benign, slow-growing, nerve sheath tumours that arise from neural, crest-derived Schwann cells along the dorsal sensory spinal roots, but occasionally, they may arise from the ventral motor roots. They represent approximately 85% of all nerve sheath tumours. These tumours are evenly distributed throughout the spine and may involve multiple compartments including intradural extramedullary (72%), extradural (13%), intradural and extradural (13%), and intramedullary (1%)2. Most schwannomas are entirely intradural, but some extend intraoraminally as a dumbbell mass or are purely extradural. Dumbbell tumours are spinal tumours with contiguous intraspinal, foraminal and extraforaminal components. Presentation is usually in the third through fifth decades with no particular sex predilection3. Symptoms are related to a slowly enlarging mass. Pressure effects on nearby organs or sensory changes in the distribution of the affected nerve are the recognised symptoms. We present a case of a giant, ancient schwannoma which was successfully excised without intra-operative neurophysiological monitoring.

Case report

A 29-year old male Ugandan of Bantu ethnicity presented painful spasms and a feeling of heaviness in the lower limbs. He had been relatively well-up until 18 months prior to admission, when he started developing progressive weakness, heaviness and numbness in the right lower limb. Three months later, he felt a sudden sharp pain in the upper back that was piercing in nature, with radiation and paraesthesias to the lower back and right lower limb. This pain was exacerbated by walking and relieved by rest. He deteriorated over time and started experiencing intermittent painful spasms of the lower limbs, which did not permit him to walk normally, so he started using walking canes to aid his ambulation. He had normal bowel and micturition habits. He had no history suggestive of tuberculosis.

On clinical assessment, he was in a fairly good general condition with no external features of neurofibromatosis, but a spastic gait and ambulation with the aid of two walking canes. The upper limbs were neurologically intact; however, he had hypertonia and a sustained clonus in the lower limbs, more marked on the right side. He had weakness in the right lower limb muscles with bilateral knee and ankle reflexes. The upper limbs were neurologically intact, had a spastic gait and ambulated with the aid of two walking canes. The upper limbs were neurologically intact; however, he had hypertonia and a sustained clonus in the lower limbs, more marked on the right side. He had weakness in the right lower limb muscles with bilateral knee and ankle reflexes.

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Plain radiographs of the cervical and thoracic spine were normal. Magnetic resonance imaging (MRI) showed a linear, oval, heterogeneous signal intensity indicating an intradural, extramedullary mass that was approximately 6.6 cm × 1.3 cm in size, involving the anterior apical canal extending from C6–T2. The lesion was heterogeneously hypointense on T1 weighted imaging (Figure 1A), hyperintense on T2 weighted imaging (Figure 1B) and Short TI Inversion Recovery on cystic areas. The spinal cord had no altered signals and was compressed and displaced posteriorly (Figure 1C).

**Operation**

Tumour excision was done through a posterior approach. Wide multi-level laminectomies were done at C5–T2 levels (Figure 2A); the tumour was exposed through a dorsal myelotomy and enucleated enblock without intra-operative neurophysiologic monitoring (Figure 2B). Posterior stabilisation was then done to prevent post-surgical instability (Figure 2C).

**Histopathological examination**

Macroscopically, the mass was firm, irregular and yellowish as shown in Figure 3. Histopathological examination, as seen in Figure 4, showed...
encapsulated tissue comprising of spindle cells in compact and loose formations (Antoni A and Antoni B). Large, thick, hyalinised blood vessels together with areas of haemosiderin and scattered calcifications, were seen. A histological diagnosis of ancient schwannoma was made.

Post-operative follow up

Post-operatively, the patient had a transient deterioration in neurology (American Spinal Injury Association–ASIA D to C) from which he recovered by the third post-operative day. The patient was discharged in a wheelchair 10 days after surgery. On follow up after three months, he had a slight spastic gait and used a cane as a walking aid. He reported no back pain, no spasms and the left lower limb numbness had disappeared. He had hypertonia in the lower limbs; however, he could passively flex his knees. He had normal muscle power in the left lower limb but power grade 4/5 in the left lower limb for all myotomes L2–S1. Deep tendon reflexes were increased. His main problem was lack of post-operative rehabilitation since his home was located in a rural region, which had no physiotherapist available.

Discussion

The histological diagnosis of this case was ancient schwannoma. Ancient schwannoma, a degenerative neurilemmoma, is a schwannoma subtype characterised by degeneration and diffused hypocellular areas. These changes are believed to occur because it takes a long time for schwannomas to develop. The term 'ancient schwannoma' was first reported by Ackerman et al. to describe schwannomas with calcification. Their histology generally displays a biphasic pattern with areas of highly ordered cellularity (Antoni type A) and less cellular areas where a highly myxoid matrix predominates (Antoni type B). They are characterised by prominent vessels with thick hyaline walls, degenerative changes typified by perivascular hyalinisation, calcification, haemosiderin deposition, cystic necrosis, relative loss of Antoni type A tissue and degenerative nuclei that may be misinterpreted as sarcomatous pleomorphisms. Calcification is the usual degenerative change, but ossification is a rare degenerative variant. These degenerative changes are thought to be the result of the long-term progression of this tumour. They are usually located deep in the head and neck, thorax, retroperitoneum, pelvis and extremities of elderly patients.

The signs and symptoms of intradural, extramedullary tumours are not specific to tumours and are similar to those caused by any spinal disorder that produces symptoms of spinal cord or nerve root compression. Because of the slow growth of these tumours, symptoms may be subtle and progress slowly over time before diagnosis. The most common symptom is pain, either localized or radicular. Because of the multifactorial and nonspecific nature of back pain, a thorough and detailed neurological examination is required to assess patients presenting with pain for any signs of myelopathy or abnormal reflexes.

MRI has made the pre-operative planning of giant tumours easier than when myelography was the most commonly used neurodiagnostic tool. The entire extent of the lesion is seen in all three planes, and the relationship of the tumour to the neural elements, vascular structures and other organs, is clearly delineated. The imaging protocol should include sagittal and axial T1-weighted and T2-weighted sequences including...

Figure 3: Tumour gross appearance: the yellowish, firm and irregular, giant schwannoma.

Figure 4: Histopathological findings: encapsulated tissue comprising of spindle cells in compact and loose formations (Antoni A and Antoni B). Large, thick, hyalinised blood vessels and scattered calcifications. The pathological diagnosis was ancient schwannoma. H&E staining ×40.
The surgical approach used in this case was posterior, where laminectomies were done from C5–T2 and the tumour resected with preservation of the nerve roots. Much as was desired, we did not do intra-operative neurophysiological monitoring because it was not available. Intra-operative electrophysiological assessment can be useful in deciding on the surgical procedures. The surgeons can decide upon total resection of a schwannoma without consequent neurological deterioration when it is demonstrated that motor and sensory nerve roots involved in the schwannoma are already less or not functional. In patients with good functioning nerve roots, there is the risk of recurrence of the tumour but partial resection may be indicated to avoid post-operative neurological deterioration. The appropriate surgical approach must be based on an understanding of the underlying anatomy and the type and location of the lesion. Though the tumour was ventrally located, we avoided the anterior surgical approach. Reluctance to use anterior approaches to ventrally located spinal cord tumours has stemmed largely from concerns about 1) inadequate access to the tumour because of a deep and constrained operative field of view, 2) excessive bleeding from the Batson’s epidural venous plexus, 3) the need for spinal reconstruction and 4) post-operative cerebrospinal fluid fistula formation through the anterior dead space created during surgical dissection.

It is important to plan for reconstruction once tumour removal has been achieved. Removal of the posterior elements with the facet joint at more than one level is likely to leave the spine unstable. To avoid iatrogenic instability, we did supplemental posterior instrumented fixation. This is particularly applicable for surgery performed across the cervicothoracic or thoracolumbar junctions. The decision on the use of hardware to stabilise the spine depends on whether the tumour removal has been radical or not. Some surgeons do not routinely stabilise the spine following tumour removal. In a case series by Sridhar et al., only two of 10 patients with giant schwannomas were stabilised following tumour removal. Their indication for stabilisation was erosion of more than 25% of the vertebral body. However, they opined that if the lesion is in the cervical or upper-lumbar spine, early mobilisation might be hazardous if the spinal column has not been stabilised. Fear of destabilising the spine should not compromise the exposure required to safely remove these tumours.

The detection of calcification within a mass is important to estimate the difficulty of resection or post-operative prognosis as well as to narrow the differential diagnosis. Evidence of calcification makes surgical removal difficult because of adhesions to the spinal cord and because calcification complicates the operative procedure. The additional manipulations required for dissecting the tumour, increase the risk of a poor neurological outcome. The advised treatment for schwannoma is total microsurgical resection. Neurological deficit should improve after surgery. Prognosis is good and tumours do not typically recur, except in cases of neurofibromatosis-2 and schwannomatosis. A solitary schwannoma will rarely show malignant degeneration. The risk is higher for patients with neurofibromatosis-2.

**Conclusion**

Giant, solitary, ancient schwannomas at the cervico-thoracic spine are uncommon. These tumours can be successfully removed even in resource-limited settings as long as the technical expertise is available. Without intra-operative neurophysiological monitoring, chances of nerve and spinal cord damage are high. However, in situations where there is an absolute surgical indication, it is worth trying even if the surgical approach is posterior.
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References