

Pure Ventral Midline Long Segment Schwannoma of the Cervicodorsal Spine: A Case Report

Servikodorsal Omurganın Orta Hat Uzun Segment Şıvannomu: Bir Vaka Sunumu

ABSTRACT

Spinal intradural extramedullary schwannomas commonly occur posterolateral or anterolateral to the spinal cord. A case of a pure midline ventrally situated cervicodorsal extramedullary schwannoma in a 33-year-old female patient with subacute cord compression and sphincteric involvement is presented. Spinal MR imaging showed a C2-D3 midline ventrally situated extramedullary tumor with severe spinal cord compression. It was resected through a posterolateral approach. Histology was consistent with a schwannoma. There was a gradual improvement in left-sided spasticity but the patient had mild diaphragmatic paresis. MR imaging showed no evidence of the tumor at followup after 6 months. 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, Schwannoma, Spine, Surgery, Ventral location

ÖZ

Spinal intradural ekstramedüller şıvannomlar sıklıkla spinal kordun posterolateral veya anterolateralinde yerleşirler. Subakut kord kompresyonu ve sfinkter tutulumu olan 33 yaşında bir bayan hastada ventral orta hat yerleşimli bir servikodorsal ekstramedüller şıvannom sunulmuştur. Spinal MRG C2-D3 orta hat ventral yerleşimli şiddetli spinal kord kompresyonu yapan bir ekstramedüller tümör göstermişti. Bu tümör posterolateral bir yaklaşımla rezeke edildi. Histoloji bir şıvannomla uyumlu idi. Sol taraftaki spastisitede bir düzelme izlendi fakat hafif bir diyafragma parezisi vardı. Altı ay sonraki kontrolde MRG tümöre ait bir bulgu göstermedi. Bu tedavisi zor tümörlerin radyolojik özellikleri, patogenezi, ve tedavilerindeki cerrahi stratejiler tartışılmıştır ve ilgili literatür kısaca gözden geçirilmiştir.

ANAHTAR SOZCÜKLER: Ekstramedüller, İntradural, Şıvannom, Omurga, Cerrahi, Ventral yerleşim

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INTRODUCTION

Spinal schwannomas are common benign intradural extramedullary spinal neoplasms (5). They usually occur in the dorsal and the cervical region and present with subacute to chronic compressive myelopathy. They tend to extend along the nerve sheath into the neural foramen, widening it and if sufficiently longstanding, cause erosion of the pedicles. Modern MR imaging has helped immensely in their accurate preoperative identification and pathological diagnosis (6). 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. An appropriate surgical approach and strategy needs to be outlined to achieve a good outcome.

CASE REPORT

A 33-year-old female presented with progressively worsening neck pain and paraesthesias in both upper limbs for the past 3 years. She had progressive stiffness of both lower limbs, difficulty in walking and straining while micturition and constipation for the past 2 months. She required support while walking. There was no history of trauma, fever or Koch. There were no lower cranial nerve symptoms. On examination, she had spastic left hemiparesis grade 4. The grip was good. Reflexes were exaggerated bilaterally in both lower limbs and were 1+ in both upper limbs with absent supinators. Plantars were extensor. The sensory level was at D6. MRI of the craniovertebral region and cervical spine revealed a C2 to D3 midline ventral tumor measuring 13.2x1.2 cm. The tumor was hypointense on T1- and hyperintense on T2-weighted images with irregular thick peripheral enhancement (Figure 1A,B,C). The spinal cord was severely compressed and displaced posteriorly. A C2-D3 laminotomy by subsequent replacement of the laminae was performed. The tumor was midline ventral and the spinal cord was stretched out like a ribbon. The ligamentum denticulatum was cut. The tumor was yellowish-grey, lobulated and vascular, had a fleshy capsule and was partly suckable. The dissection was commenced from D3 and carried upwards. The tumor was adherent to the ventral surface of the spinal cord but no definite attachment of the tumor could be found. Intraoperative spinal cord monitoring was not performed. 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 (Figure 2A,B). Immediately following surgery, spasticity and power in left-sided limbs improved and the patient had mild diaphragmatic paresis at discharge. There was mild improvement in the sphincteric complaints. MR showed no evidence of residual tumor at follow-up after 6 months (Figure 3).

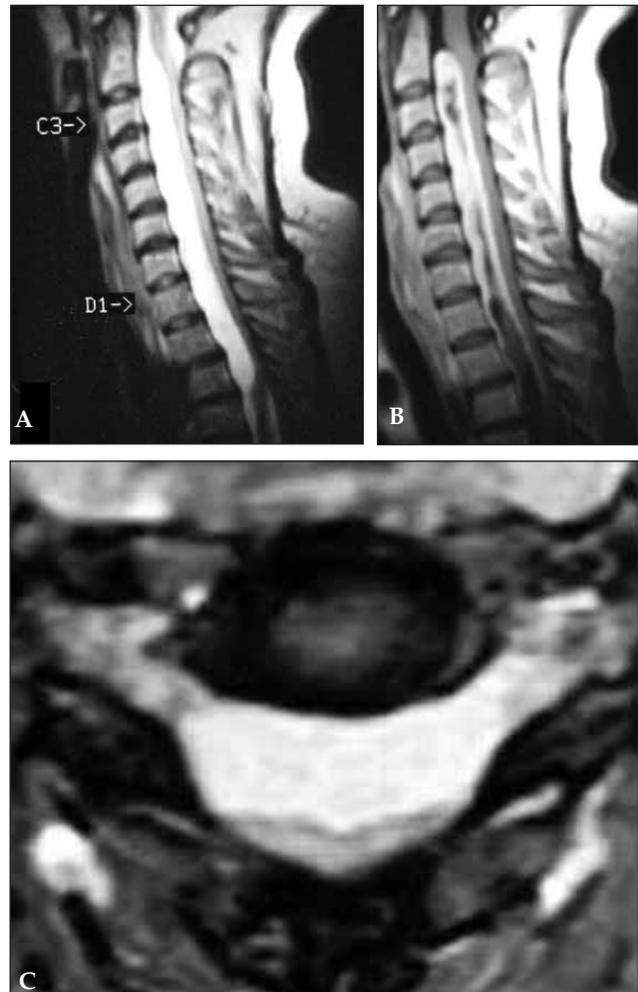


Figure 1: MRI showing ventrally situated extramedullary schwannoma extending from C2-D3.

A. Sagittal T2-weighted image.

B. Sagittal T1-contrast enhanced image.

C. Axial T1-weighted contrast enhanced image.

DISCUSSION

Spinal schwannomas are usually dorsal, lateral or dorsolateral in position and about 10 to 15% extend through the dural root sleeve into the intervertebral foramen producing a dumbbell-shaped appearance (5). Giant spinal schwannomas have been reported

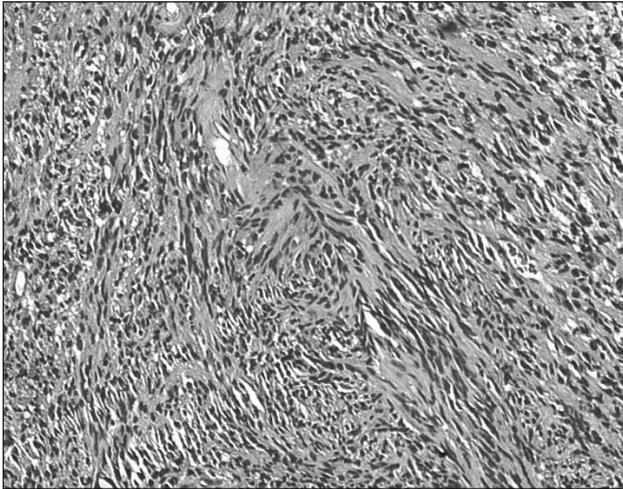


Figure 2: Photomicrograph showing densely packed elongated spindle cells in interlocking fascicles (Antoni type A), intermingled with loosely textured tissue with extracellular clear spaces (Antoni type B). H&E, original magnification X 200.



Figure 3: Sagittal T2-weighted MR image showing no evidence of tumor.

but pure ventral midline intradural schwannomas are rare (12, 13). They are restricted to one region of the spine and usually extend over two or three spinal segments. We report the first case of a C2 to D3 pure midline ventral cervicodorsal intradural extramedullary schwannoma. Intradural schwannomas usually arise from the posterior rootlets (80%) and few arise from the anterior rootlets (20%) (5,12). The origin of schwannoma is easier to identify if it extends over a short segment but may be difficult or impossible to judge in long segment schwannomas, as observed in

our case. It is also known to arise in the extraarachnoid space, nervi vasorum of the anterior spinal artery and the leptomeninges in the region of the anterior median septum, ventral to the dentate ligaments and anterior rootlets (7,9,10). Schwannomas usually show hypointense or isointense signal on T1-weighted and hyperintense on T2-weighted images and homogenous contrast enhancement on preoperative MR imaging (2). Meningioma and tuberculous granulomas are

important differential diagnoses (4,8). It may rarely mimic a glioma (13).

Surgery for a dorsal or dorsolaterally situated schwannoma is relatively straightforward since the spinal cord is pushed anteriorly or anterolaterally (1, 5, 12). The anterior approach is suited best for ventral midline intradural tumors which are small in size, apposed to the ventral surface of the spinal cord and produce no significant displacement or rotation of the spinal cord (2, 9, 11). However, the acceptance is modest due to scarce experience and literature on the subject. The surgical considerations in our case included the pure midline ventral situation of the tumor spanning multiple spinal segments. The posterior compression and displacement of the spinal cord is a potential risk for inadvertent injury and enhanced postoperative morbidity. The midline ventral location of the tumor would probably dictate an anterior approach. However, the posterolateral approach was more suitable since a wider exposure would be required to achieve total control whilst removing the tumor. The predominant bulk of the tumor was in the anteroposterior direction than lateral. In addition, the spinal cord was compressed and displaced posteriorly as well as flattened against the lamina which provided a posterolateral corridor access for internal debulking of the tumor. The uniform hyperintense signal on T2-weighted MR images was suggestive of a relatively soft to firm tumor consistency. A rapid and radical internal decompression allowed the tumor to express into the operative field and relieve the midline ventral cervical spinal cord compression. The upper third of the tumor was free and lying loose in the subarachnoid while the tumor in the middle and the lower third was adherent to the ventral surface of the spinal cord suggesting caudocranial growth. Transverse dural incisions, sectioning of dentate ligaments and extensive posterolateral bone removal in the posterolateral approach all need attention. Longterm followup is essential in our patient although there was no immediate cervical spinal instability. Endoscopic assisted removal of ventral intradural spinal tumors through a posterior approach allows minimal retraction of the spinal cord as well as inspection of the corridors which are difficult to visualize with the microscope (3).

In conclusion, surgery for pure ventral midline intradural cervicodorsal long segment schwannoma is formidable. Anterior surgical approach is best

suited for small sized midline ventral tumors extending over a short spinal segment and in close approximation to the ventral surface of the spinal cord. A posterolateral approach is feasible in a pure midline ventrally located large sized tumor spanning over multiple spinal segments. The anteroposterior extent and consistency of the tumor should be evaluated on MR imaging to assess the suitability for a posterolateral surgical approach.

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