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Case Report

Post-resection complication of large dumbbell-shaped extradural schwannoma: a case report and literature review

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ABSTRACT

Primary spinal tumors are rare neoplasms with an incidence of 0.3 to 0.5 cases/1 lakh population. Schwannomas are the most common benign tumors of the spinal cord, arising from the nerve sheath cells accounting for 25% of all the spinal tumors. Complete surgical resection of the tumor is the treatment of choice in spinal schwannomas. Post resection complications are uncommon based on the factors affecting the tumor. In this case report, patient with extradural spinal schwannoma located at thoracic region was resected. She was complicated with paraplegia post operatively and successfully recovered following the standard protocol and rehabilitation.

Keywords: Schwannoma, Extradural, Paraplegia

INTRODUCTION

Primary spinal tumors are rare neoplasms with an incidence of 0.3 to 0.5 cases/1 lakh population.[1] These tumors can be seen in various locations including intramedullary or extramedullary, intradural or extradural locations, or even dumbbell (intradural, intraspinal, and extraspinal).[2] Schwannomas are the most common benign tumors of the spinal cord, arising from the nerve sheath cells accounting for 25% of all the spinal tumors.[3] Its prevalence is similar in males and females, and it is usually diagnosed during the fourth and fifth decades of life.[4] The appearance of symptoms is proportional to the size of the tumor and can be usually due to compression or infiltration of the spinal cord or nerve roots causing pain or neurological deficit.[3,5] Dumbbell-shaped tumors involve different compartments and thus can be surgically challenging.[6]

Complete surgical resection of the tumor is the treatment of choice in spinal schwannomas. Preservation of maximum neurological function based on the size, location, and extent of the tumor is the main goal of surgery. The known post-operative complications are new or worsening sensory and motor deficits, cerebrospinal fluid (CSF) leaks or pseudo meningocele, wound infections, spinal deformity, spinal hematomas, nonoperative cranial subdural hematoma, deep venous thrombosis, urinary retention, and recurrent laryngeal nerve injury.[2] The recurrence rate of schwannoma is found to be <5% which may be due to subtotal tumor removal which can manifest several years after the initial surgical resection.[7] We present a case of a fully recovered postoperative paraplegic patient after the resection of the extradural extraforaminal dumbbell shaped schwannoma.

CASE REPORT

A 55-year-old female presented to the orthopedic outpatient department with complaints of weakness in bilateral lower limbs which was progressive over 6 months and a history of difficulty in walking for one month. There was no previous history of trauma, back pain, radiculopathy, or any other systemic diseases. A detailed clinical examination was performed on the patient. Motor examination revealed bilateral lower limb spasticity, hypertonia, and a power grade of 4/5 bilaterally. Sensory examination revealed reduced sensation below the T8 level. Exaggerated deep tendon reflexes, bilateral babinski extensors, and ankle clonus were present.

A contrast magnetic resonance imaging (MRI) of the dorsolumbosacral spine was performed which revealed well defined lobulated heterogeneous lesion on the left side of the dorsal-spinal canal extending at the foraminal and extraforaminal paraspinal region with a dumb-bell shape (Figure 1). The mass measured approximately 3.7×2.0 cms intraspinally (extending from D5-D8 levels), and 3.5×3.2 cm extraforaminal and paraspinally (at D6-D7 levels). The mass was found to be causing significant compression and displacement of the adjacent cord on the right side, resulting in bilateral lower limb weakness which was diagnosed as an extramedullary extradural extraforaminal paraspinal nerve sheath tumor (Figures 2 and 3).

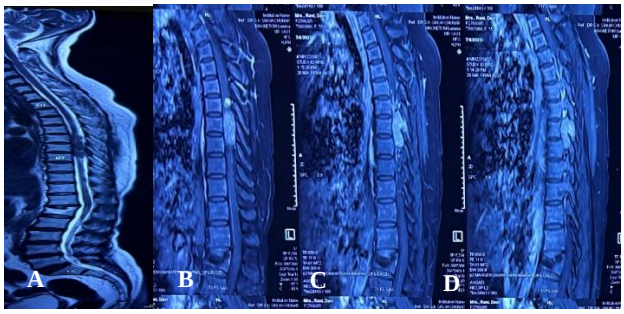

Figure 1 (A-D): T2W MRI sagittal section shows the hypodense area in the spinal canal at the level of D5-D7 vertebra.

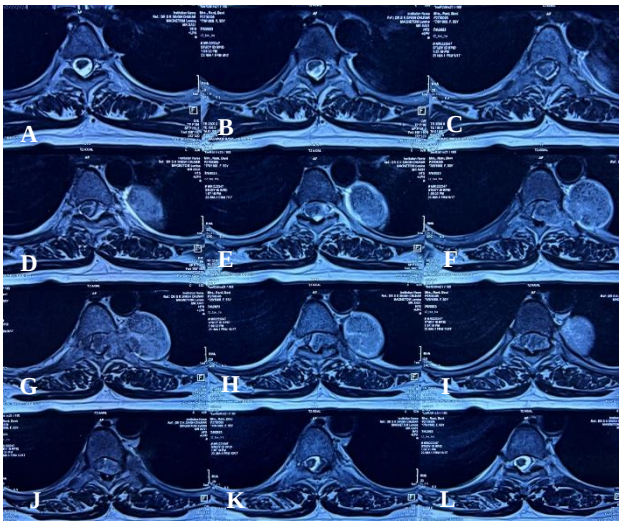

Figure 2 (A-L): T2W MRI Axial view shows the heterogenous extramedullary intradural extradural tumour.

The patient was counseled, admitted, and planned for resection of the tumor. Under general anesthesia, the patient was put in prone position ensuring adequate padding. Under all aseptic precautions, the thoracolumbar area was scrubbed, painted, and draped. A midline incision was made centering the D5-D8 vertebrae. Hemilaminectomy was done from D5-D8 vertebrae along with 6th and 7th rib excision, and costotransversectomy was performed (Figure 4). A large dumbbell-shaped tumor compressing the cord on the left side extending up to the posterior mediastinum was found. En-bloc mass resection of the tumor was performed (Figure 5).

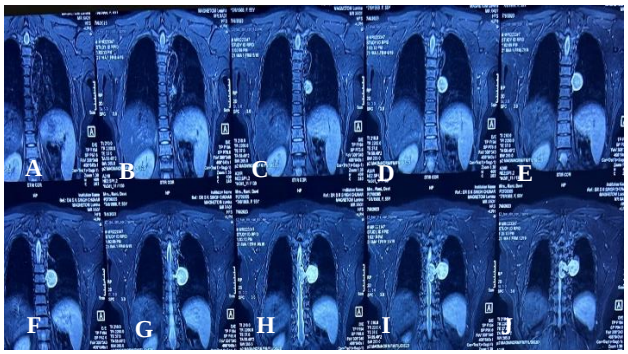

Figure 3 (A-J): MRI coronal view shows the mediastinum extension of the tumour towards the left side.

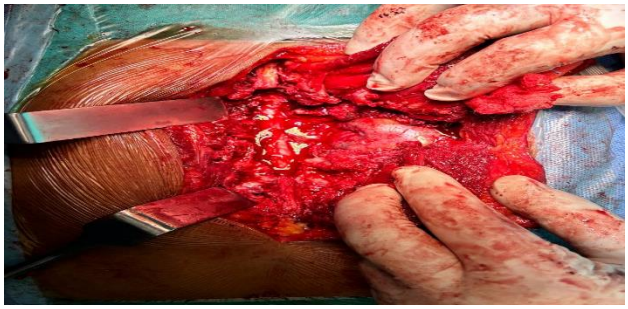

Figure 4: Hemilaminectomy done from D5-D8 along with the excision of 6th and 7th ribs costotransversectomy for the exposure of the mediastinal extension of the tumour.

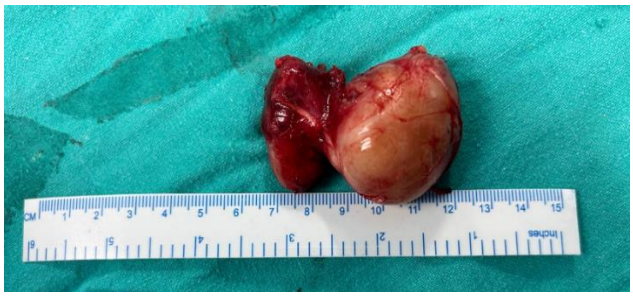

Figure 5: En bloc resected schwannoma.

Histopathological examination showed a well-circumscribed, well-encapsulated tumor comprising Verocay bodies composed of Antoni A and Antoni B areas which was diagnostic of Schwannoma.

Immediate postoperative examination revealed motor power of 0/5, absent sensation below T7 level, flaccid paraplegia, and LMN bladder. A repeat MRI was done to rule out hematoma collection or any other intraoperative injuries. Re-exploration was done which showed no

significant collection or injury and was closed with the insertion of a drain.

The patient was administered intravenous steroids for 5 days, followed by oral steroids for 1 week. Patient was later shifted to the rehabilitation ward where she stayed for 15 days. After 15 days, she showed signs of recovery. Initial flickering of movements in lower limbs, followed by bladder control and subsequent bowel control was achieved. At the end of 3 months follow up patient regained all the sensations and a motor power grade of 4/5 was achieved. The patient was able to walk with support.

DISCUSSION

Schwannomas are benign slow-growing tumors originating from the Schwann cells of peripheral nerves.[5] Apart from the rare intramedullary schwannoma, it has been classified as intradural, intradural-extradural, and purely extradural. The most common clinical symptoms are radiating pain and radiculopathy, with rare presentation of sensory and motor deficits. Whereas in our case, the prime complaint was paraparesis along with pyramidal tract signs similar to the study done by Paulo et al.[8] Schwannomas usually originate from the sensory nerves with sensory symptoms. In contrast, the present case has weakness as the symptom which represents motor involvement. Nerve sheath tumors are more often seen in the cervical and lumbar, although schwannomas can occur in the thoracic region, it is an uncommon location which is the site of involvement in our case.[2] Solitary schwannomas are often encountered with good neurological results whereas multiple schwannomas in a patient are referred to as schwannomatosis. Schwannomatosis is usually indicative of an underlying tumor predisposition syndrome, such as neurofibromatosis which decides the prognosis of the patient.[9]

Many classifications are proposed for nerve sheath tumors based on the extent, location, and size of the tumors.[10] Sridhar et al and modified Shridhar by Park et al, are the routinely used classifications in clinical practice.[11,12] According to the Sridhar et al classification, our case belongs to type 4 b which is an intraspinal tumor extending into extraspinal space with the tumor being >2 cm.[11]

Spinal schwannoma has two histopathological patterns, Antoni A (hypercellular) and Antoni B patterns (hypocellular). In our case, histopathological examination showed spindle cells with areas of predominant hypercellular and focal hypocellular patterns, without evidence of malignancy.[13]

Surgical resection of the tumor is the most accepted treatment for benign nerve sheath tumors with maximum preservation of the neurological outcomes.[1] In our study, T5–T8 hemilaminectomy was done for the exposure of the tumor and 6th, 7th ribs were removed for the decompression of the posterior mediastinal extension of the tumor.

In our study, we have used spinal-schwannoma postoperative neurological deterioration scoring system (SPNDSS) to predict postoperative neurological deterioration. According to this system, scoring is done based on 5 variables: including a duration of preoperative symptoms of more than 3 months, preoperative radiating pain, tumor size larger than 2cm, tumor occurring in the lumbar spine, and dumbbell shape. In this scoring system, a scale ranging from 0 to 7 based on these scores is designed which is used to categorize the tumor as low risk (0-2 point – 8.7% prediction risk of postoperative neurological deterioration), moderate risk (3–5 point - 36% prediction risk of postoperative neurological deterioration) and high risk (6–7 point - 87.5% prediction risk of postoperative neurological deterioration). Our case falls under the moderate risk stratification category with 36% prediction risk, as per this SPNDSS scoring system.[14]

The use of intraoperative neuromonitoring was associated with the determination of the extent of deficit thus enhancing the extent of resection and minimizing postoperative neurological deficits. Although in our case neuromonitoring was not done, it is the safe and effective method to be practiced but few studies showed no difference in the outcome with or without the neuromonitoring.

Many studies have been reported to witness postoperative complications to be 32% following the resection of the spinal tumor.[2] Common complications include hematomas, new or worsening sensory symptoms followed by new or worsening weakness, CSF leak or pseudo meningocele, wound infection, and spinal deformity. The prediction of such postoperative complications is always debatable because the neurological and surgical outcomes are influenced by many factors such as tumor size, intradural location and its extension, number and location of the tumor, preoperative involvement of the sensory, motor, or bladder, and bowel involvement. Kim et al reported a neurologic deficit rate of 23% in a study done on 31 cases that involved functionally important nerve roots, but the observed deficit was not functionally debilitating in any of the cases.[15] In our case, patient with preoperative paraparesis of 4\5 with extradural dumbbell-shaped schwannoma following resection had postoperative complication of complete paraplegia below the level of T7. A similar complication of transient and later permanent root palsy post resection of extradural schwannoma was seen in a study done by Paulo et al.[8] Even though our patient had a complication of complete paraplegia, she was able to achieve complete recovery of all the deficits with appropriate treatment and rehabilitation protocol.

CONCLUSION

Schwannoma is a rare benign tumor that can be effectively treated with surgical removal. Although postoperative complications are rare, their occurrence cannot be predicted even with the use of risk assessment scoring

systems. Therefore, operating surgeon needs to have a comprehensive understanding and practice of appropriate management protocols to ensure the complete recovery of patients with spinal tumors.

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