

Case Report

Thoracic Intramedullary Schwannoma Accompanying by Extramedullary Beads-Like Daughter Schwannomas

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Thoracic intramedullary schwannomas are rare spinal cord tumors. Most of these tumors have been reported as a single lesion in the spinal cord. The authors report the first case of intramedullary schwannoma accompanying by extramedullary beads-like daughter masses of the thoracic spine. A 68-year-old male presented with walking disturbance and decreased sensation below T10 dermatome. Imaging workup revealed an intramedullary mass at T6 and T7. T6 and T7 laminectomy and mass removal were performed. Intraoperatively, extramedullary beads-like daughter masses along the nerve roots adjacent to intramedullary mass were identified. Total removal of intramedullary lesion and partial resection of extramedullary masses were done. Histological analysis confirmed the diagnosis of schwannoma. The patient could ambulate independently at postoperative 1 month without any neurological sequelae. The authors experienced a surgical case of intramedullary schwannoma accompanying by extramedullary beads-like same pathologies in the thoracic spine.

Key Words : Neurilemmoma · Intramedullary lesion · Spinal cord neoplasm.

INTRODUCTION

Spinal schwannomas are ones of the most common primary tumors of the spine, but intramedullary schwannomas in non-neurofibromatosis patients have been reported rarely, accounting for less than 1.1% of all schwannomas^{2-6,9,10,15}. As previously stated, these lesions have appear as intramedullary single lesions or dumbbell intra- and extramedullary ones.

The authors report a case of an intramedullary schwannoma accompanied by extramedullary bead-like daughter masses in the thoracic spine.

CASE REPORT

A 68-year-old male was admitted with a 17-month history of walking disturbance and decreased sensation below the T10 dermatome. He had no traumatic accident and no other previous disorders such as neurofibromatosis, or specific lesions on his skin. Neurological examination revealed that the motor function of both legs was grade 4/5. The sensation of all modalities below the T10 dermatomes was reduced. His bilateral knee and

ankle jerks were hyperactive and continuous ankle clonus was observed. Gadolinium-enhanced magnetic resonance imaging (MRI) demonstrated an ill-demarcated, highly enhanced intramedullary ellipsoidal mass at the level from T6-7. A moderate spinal cord edema was also noted at the T3/4-9 level, and syrinx formation was noted above the mass. No abnormal enhancing lesion was present along the nerve root near the mass. The mass (6×7×30 mm) was located at the center of the spinal cord and was preoperatively considered to be intradural and intramedullary (Fig. 1, 2).

The operation was done with a standard posterior midline approach. Following skin incision and muscle dissection, T6

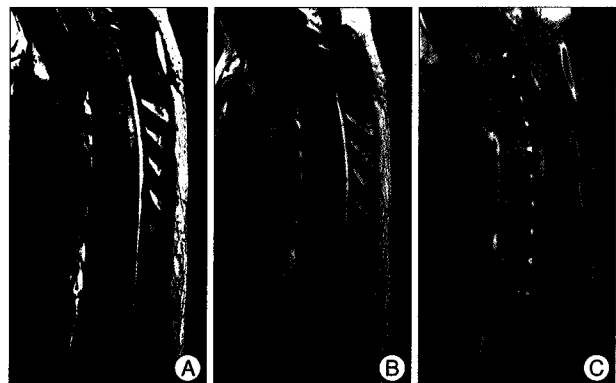


Fig. 1. Sagittal T2 (A), T1 (B) and T1 postgadolinium (C) spinal magnetic resonance imaging demonstrating an ill demarcating and heterogeneously enhancing intramedullary mass at level of T5-T6.

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and T7 laminectomies were performed. When the dura and arachnoid were opened, an expanded thoracic cord and extramedullary bead-like masses along the adjacent nerve roots were identified (Fig. 3). The bead-like masses were firmly attached to the nerve roots and partially resected. The frozen section findings were consistent with a schwannoma. After the midline myelotomy, a dark brown intradural mass was seen. Frozen section findings of the intramedullary lesion were also defined as a schwannoma. The lesion was dissected off the spinal cord tissue and totally removed. The extramedullary and intramedullary masses were not connected. Intraoperative monitoring using motor-evoked potential was uneventful. After the operation, the patient's sensory and motor deficits gradually improved. A month after the operation, the patient could ambulate with no assistance.

Histological analysis confirmed the diagnosis of schwannoma. Immunocytochemical staining for S100 protein and silver was strongly positive, and staining for glial fibrillary acidic protein and epithelial membrane antigen was negative (Fig. 4).

DISCUSSION

Conti et al.⁴⁾ analyzed the literature from 1931 to 2002 and found only about 50 reported cases of non-neurofibroma related intramedullary schwannomas. Most of these lesions have been reported as single intramedullary lesions, and more rarely as dumbbell intra- and extramedullary ones. The present case of an intramedullary schwannoma is accompanied by bead-shaped multiple small schwannomas arising on the adjacent nerve roots, which are separated from each other.

Schwannomas originate from the Schwann cells. Schwann cells are absent in the central nervous system, thus it can explain the rarity of intramedullary schwannomas. The pathogenesis of intramedullary schwannoma is still not clear, and various hypotheses have been proposed. The postulated pathogeneses of these tumors' origin include Schwann cells along the intramedullary perivascular nervous plexus^{7,14,16)}, focal intramedullary proliferation of Schwann cells in reaction to chronic disease or trauma^{1,11)}, ectopic Schwann cells originating from migrating neural crest cells^{8,12,17,18)}, and Schwann cells related to aberrant intramedullary myelin fibers¹¹⁾. Also, Schwann cells in the posterior nerve root at the root entry zone are assumed to be one of pathogeneses of intramedullary tumors^{13,19)}. A tumor arising from Schwann cells in this "critical area,"

where the nerve root loses its sheath, could enter the subpial area in the spinal cord and appear as an intramedullary mass. The schwannomas in the present case consisted of an intramedullary large one and extramedullary bead-like small ones on the

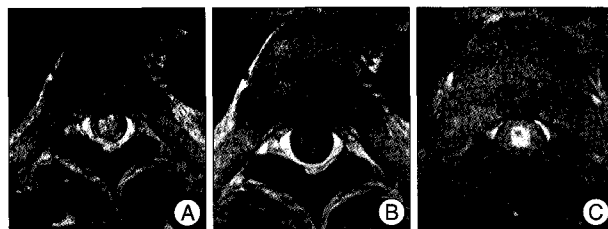


Fig. 2. A: Axial T2 (A), T1 (B) and T1 postgadolinium (C) spinal magnetic resonance imaging showing that the mass is located in the center of spinal cord.

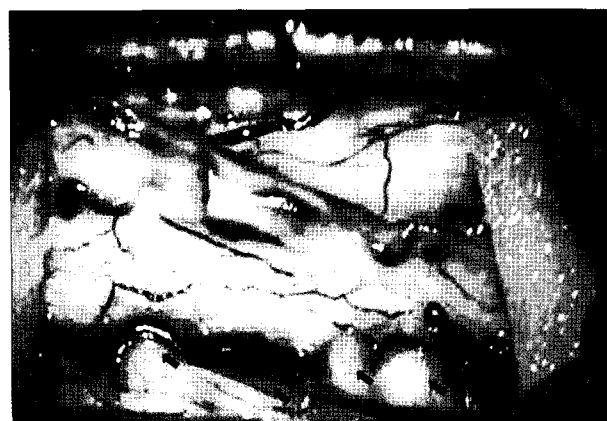


Fig. 3. Intraoperative photograph showing extramedullary multiple beads-like masses along the nerve roots (black arrows).

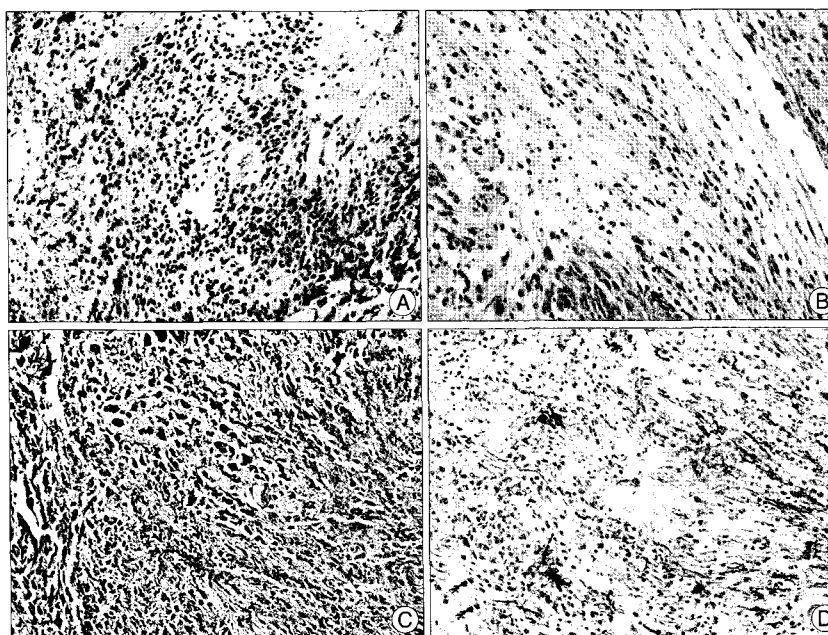


Fig. 4. Histopathological findings. The pathological diagnosis is schwannoma. A : Hematoxylin and eosin staining. Elongated bipolar spindle cells with arrangement of nuclei in palisades pattern, typical of Antoni A tissue ($\times 200$). B : Hematoxylin and eosin staining. The field is contrasted with the loosely organized hypocellular Antoni B region ($\times 200$). C : Photomicrograph showing positive reactions in immunohistochemical staining for S100 protein. D : Negative staining for glial fibrillary acidic protein.

adjacent nerve roots. Although the masses were separated from each other, the pathogenesis of all masses would be the same in a single patient. These findings could support the hypothesis that intramedullary schwannomas originate from Schwann cells in the posterior nerve root at the root entry zone.

Complete surgical excision is the treatment of choice for spinal schwannomas, including intramedullary ones. In the present case, however, the authors could not completely remove all tumors. Although it was a simple matter to totally excise the intramedullary mass, there was no option but to partially resect the extramedullary bead-like masses because of their tight attachment to the nerve roots. The authors plan to follow this patient in the short term and treat him with radiotherapy if his tumors increase in size or recur.

CONCLUSION

The authors experienced a surgical case of an intramedullary schwannoma accompanied by extramedullary bead-like masses of the same pathology in the thoracic spine. The present case could be helpful for understanding intramedullary schwannomas' pathogenesis.

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