Cervical Intramedullary and Extramedullary Schwannoma

Servikal Bölgede Omurilik İçi ve Dışında Yerleşen Schwannoma

AHMET BEKAR, ŞEREF DOĞAN, TEOMAN CORDAN, ŞAHSİNE TOLUNAY

Uludağ University Medical Faculty, Departments of Neurosurgery (AB,ŞD,TC), and Pathology (ŞT), Bursa, Turkey

Abstract : A case of solitary cervical intra and extramedullary schwannoma diagnosed by magnetic resonance imaging and treated surgically is reported.

Key Words : Extramedullary, intramedullary, magnetic resonance imaging, schwannoma

Özet : Manyetik rezonans görüntüleme ile tanı konulan ve cerrahi olarak tedavi edilen servikal bölgede omurilik içinde ve dışında yerleşmiş schwannoma olgusu sunulmaktadır.

Anahtar Sözcükler : Ekstrameduller, intrameduller, manyetik rezonans görüntüleme, schwannoma

INTRODUCTION

It is rare for a schwannoma to occur in an intramedullary location (7,8,12). It has been reported that they comprise approximately 0.3% of primary intraspinal neoplasms (12). Because the schwann cell is normally not found within the parenchyma of the brain and the spinal cord it is not surprising that these lesions are rare in this location (7,14).

A case of cervical intramedullary and extramedullary schwannoma diagnosed by MRI and treated surgically is reported and the etiology, the role of the diagnostic procedures, and problems of surgical treatment are discussed.

CASE REPORT

A 40-year old man presented with a history of progressive spinal cord dysfunction. His complaints appeared 5 years ago, with a neck pain radiating to the left upper limb, associated with progressive numbness and weakness. Three years after his initial complaints, he experienced similar features in his right upper limb, and progressive weakness and spasticity of both lower limbs with difficulty in walking. Over 6 months before admission he noted constipation, urinary hesitancy and intermittent urinary incontinence accompanied by tenesmus.

There was no family history of Von Recklinghausen's disease.

Examination: His physical examination was normal. Neurological examination revealed spastic tetraparesis, prominent on the left extremities. All modalities of sensation were diminished below Th_2. Deep tendon reflexes were exaggerated in all four limbs. A positive Hoffman sign was present bilaterally, clonus and Babinsky signs were positive in the left lower limb. Gait was broad-based due to spasticity. Urine and stool retention were present. Marked wasting of the small muscles of the hands

was present in both upper extremities and there was disuse atrophy in the lower limbs.

Radiological Findings: Plain x-ray films of the cervical spine were normal. Magnetic resonance imaging (MRI) of the cervical cord revealed diffuse enlargement of the cervical cord extending from C2 to the level of Th1. The enlarged cord was of mixed signal intensity on T1 weighted image (Figure 1) and of increased signal intensity on T2 weighted image. Post gadolinium MRI revealed a high-density well delineated mass lesion which was occupying up to 90% of the spinal canal volume (Figure 1).The mass had cystic components. The preoperative diagnosis was therefore an intramedullary tumor with an exophytic component (Figure 2).

Operation: A C2-C7 laminectomy was performed. On opening the dura mater, a clearly defined tumor was seen. The tumor which was compressing the cord to right was grayish and encapsulated. It was easily dissected from the cord and was interpreted as an intradural extramedullary tumor. The tumor extended three segments superiorly in the extramedullary space, and penetrated the spinal cord at the midline. At this point the spinal cord was enlarged and the tumor extended in the intramedullary area. Because of the infiltrative nature of the tumor, the intramedullary part was resected subtotally. A dural patch graft was placed.

Histopathological examination revealed a connective tissue tumor composed predominantly of compact spindle-shaped cells, arranged in short bundles or interlacing fascicles. This pattern was consistent with Antoni A type schwannoma (Figure 3). With reticulin staining the tumor showed

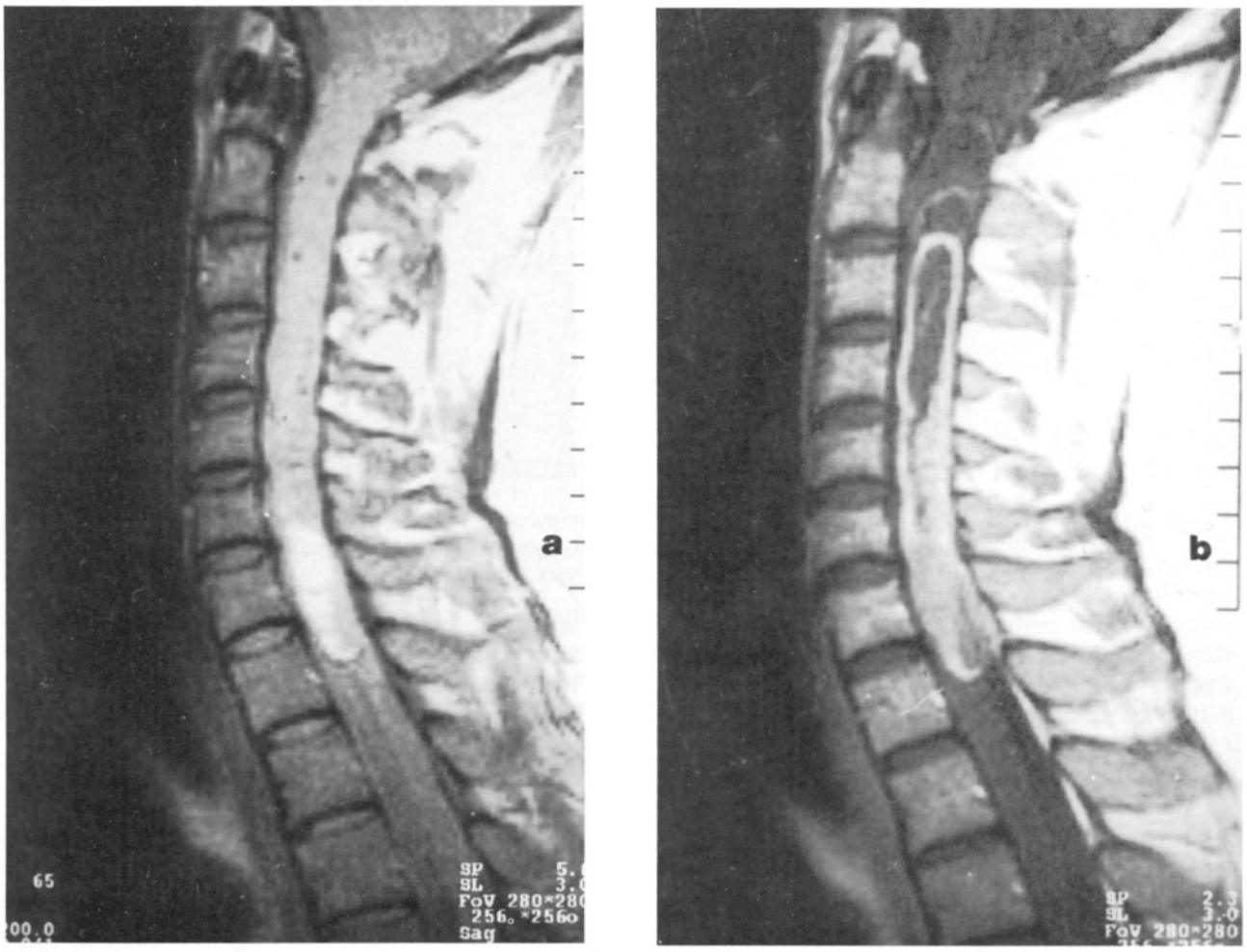

Figure 1, a) Pre contrast T1 weighted MRI revealing a diffuse enlargement of spinal cord showing mixed signal intensity between C_2-Th_1, b) sagittal post gadolinium T1 weighted image showing a high density well delineated mass lesion.

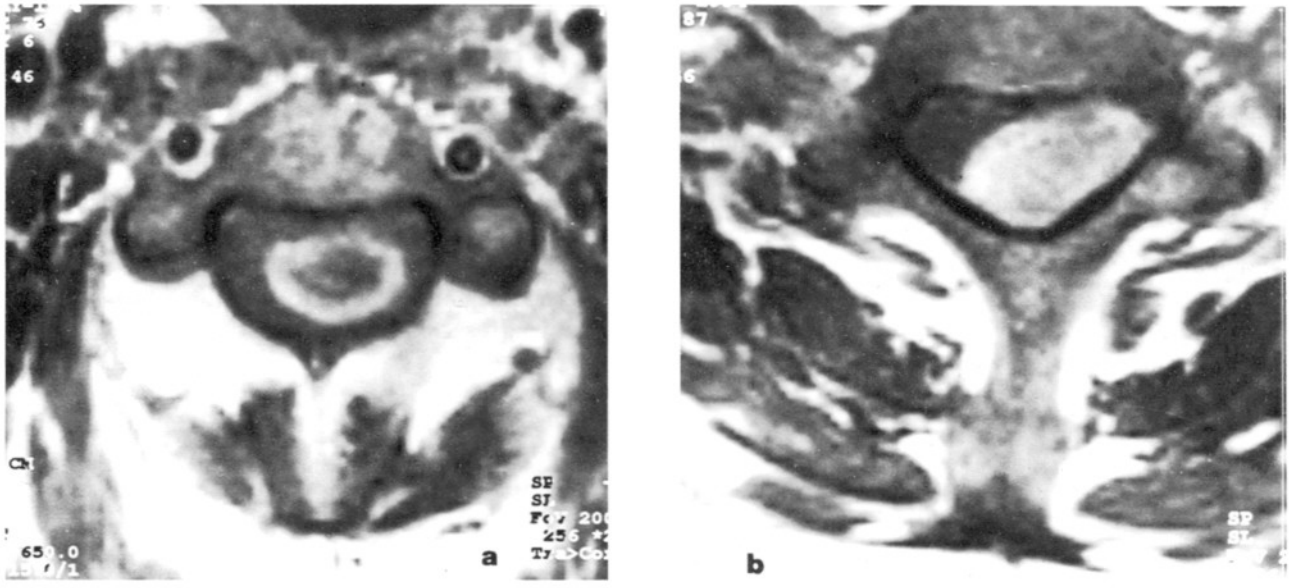

Figure 2, Axial post gadolinium T1 weighted images, a) intramedullary location of the tumor at C_2 level, and b) extramedullary component of the tumor at C6 level..

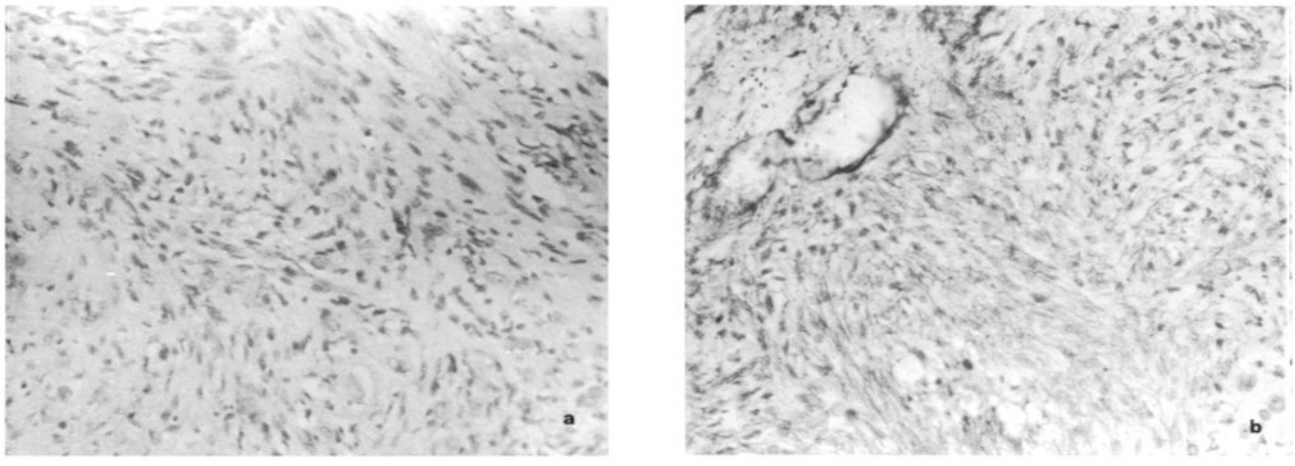

Figure 3, a) Photomicrograph showing a connective-tissue tumor composed of spindle-shaped cells, arranged in short bundles or interlacing fascicles consistent with an Antoni A area of a typical schwannoma, (H&E X100), b) reticulin stain showing reticulogenetic perivascular and pericellular pattern, (X100).

reticulogenetic perivascular and pericellular pattern (Figure 3). Immunocytochemical staining for glial fibrillary acidic protein and EMA was negative and S-100 was positive.

Postoperative course: After the operation spastic tetraparesis increased and urine retention required catheter drainage. During the postoperative two months the patient showed a steady improvement and was able to void spontaneously. After 5 months he was able to walk with support and one year later he could again work as a watchman with only one cane. Control MRI revealed a residual mass of 2 cm and a syrinx at the superior end of the intramedullary mass at the level of C4-5 which was isointense in T_1-weighted images and hyperintense in T_2-weighted images and enhanced by gadolinium injection (Figure 4).

DISCUSSION

Intramedullary schwannomas are rare tumors (2,9,11,13,15,16). We found 47 cases in the literature, in addition to our case (3,8,20). Of these cases only three have been reported as having both intramedullary and extramedullary component (5,7,10,16). Gorman et al., have reported the extramedullary component to be an exophytic

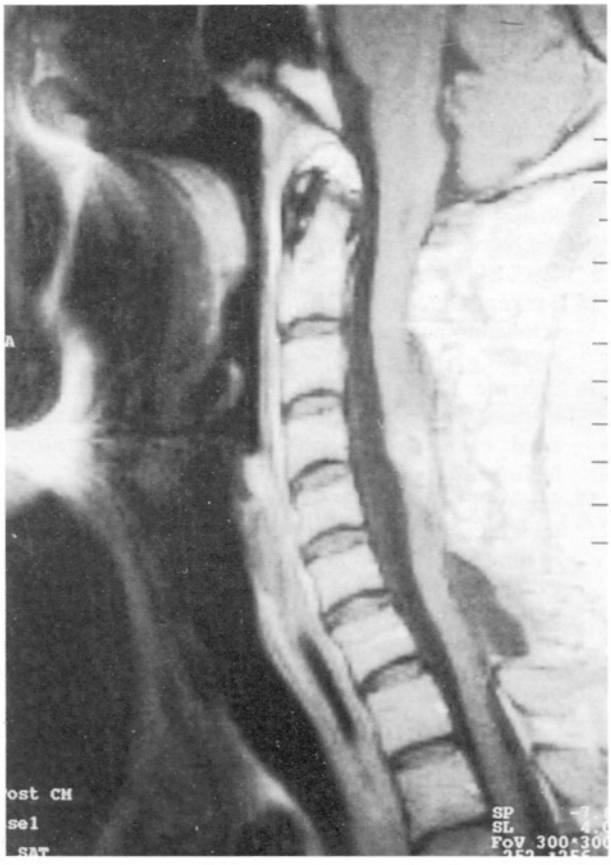

Figure 4. Post gadolinium T1 weighted sagittal MRI revealed enhancement at the level of C_4-C_5 one year after the operation.

extension of the intramedullary tumor from the enlarged spinal cord (7).

It is evident from the literature that males and females are affected equally. The mean age is 40, and the most affected part is the cervical cord. The time lag between the initial symptoms and the treatment ranges from 6 weeks to 12 years with a mean interval of 2.7-2.8 years (3,8). It was approximately 4 years in our case.

As Herregodts et al. have mentioned major clinical signs are motor deficiency (50%), diminished sensitivity (30%), and loss of genitourinary or rectal control (16%) (8). The clinical presentation in our case was that of progressive spinal cord dysfunction and radicular pain. It is also reported that when the syndrome is fully developed there are no pathognomonic clinical signs that allow differentiation between an intramedullary and extramedullary tumor (7).

According to recent reports, MRI and high resolution CT techniques facilitate the diagnosis and localization of intramedullary spinal cord tumors (1,5,6,17,20). In our case the use of gadolinium enhanced MRI contributed to the diagnosis of an intramedullary tumor with an extramedullary component and differentiating if from cord edema (1).

The absence of schwann cells within the brain and spinal cord in normal individuals has raised speculation as to the pathogenesis of these tumors (2,8,13,18). Hypoteses have discussed the possibility that these tumors arise from proliferation of schwann cells in the perivascular plexuses within the central nervous system (2,9,11,18). Alternatively, the disordered migration of the neural crest elements at the moment of neural tube closure during the fourth week of embryogenesis may result in these tumors (9,12,13,18).

As the tumor in our case was dorsally placed this theory seems plausible. Since all reported spinal cord schwannomas have been located within the dorsal columns a pathological relationship to a midline structure is probable(7,8).

Schwannomas are usually benign, well-delineated and posteriorly located tumors,and are eminently suited for surgical excision (9,11,13). In the literature it is reported that the use of ultrasonic aspirator and surgical microscope facilitate the removal of intramedullary tumors with minimal damage to adjacent cord substance (4,8,19).

Since schwannomas are sometimes difficult to differentiate from spinal gliomas on MRI or during surgical exploration, a biopsy specimen should be obtained to obtain a correct diagnosis on frozen sections (2). Although the diagnosis of the frozen section was schwannoma in our case resection was incomplete because of high cervical localization.

Schwannomas of the spinal cord are rare tumors and necessisate high quality investigative techniques, particularly MRI. Since they are usually benign, complete surgical resection with histological confirmation should be considered with ultrasonic aspirator and surgical microscope.

Correspondence: Ahmet Bekar MD
Department of Neurosurgery,
Uludağ University School of Medicine,
Bursa, Turkey
Phone: (224) 4428081
Fax : (224) 4428034

REFERENCES

1. Aichner F, Poewe W, Rogalsky W, Wallnšfer K, Willeit J, Gerstenbrand F: Magnetic resonance imaging in the diagnosis of spinal cord diseases. J Neurol Neurosurg Psychiatr 48:1220-1229, 1985
2. Aryanpur J, Long DM: Schwannoma of the medulla oblongata : case report. J Neurosurg 69: 446-449, 1988
3. Drapkin AJ: Intramedullary schwannoma (letter). J Neurosurg 75:834, 1991
4. Epstein FJ, Farmer JP, Freed D: Adult intramedullary spinal cord ependymomas: the result of surgery in patients. J Neurosurg 79:204-209, 1993
5. Gelabert GM, Garcia AA, Conde AC, Bollar ZA, Martinez RR, Reyer OF: Intramedullary spinal neurofibromas diagnosed with computed tomography: report of a case. Neurosurgery 16:543-545, 1985
6. Gonzalez MG, Allut AG, Alonso CC, Zabala AB, Rumbo RM, Oliveros FR: Intramedullary spinal neurofibroma diagnosed with computed tomography: report of a case. Neurosurgery 16:543-545, 1985
7. Gorman PH, Rigamonti D, Joslyn JN: Intramedullary and extramedullary schwannoma of the cervical spinal cord: case report. Surg Neurol 32:459-462, 1989
8. Herregodts P, Vloebergus M, Schmedding E, Goossens A, Stadnic T, D'Haens J: Solitary dorsal intramedullary schwannoma:case report. J Neurosurg 74:816-820,1991
9. Marchese MJ, McDonald JV: Intramedullary melanotic schwannoma of the cervical spinal cord: report of a case. Surg Neurol 33:353-355, 1990
10. Nicoletti GF, Passanisi M, Castana L, Albanese V: Intramedullary spinal neurinoma:case report and review of 46 cases. J Neurosurg Sci 38 :187-191, 1994
11. Prakash B, Roy S, Tandon PN: Schwannoma of the brainstem. J Neurosurg 53:121-123,1980
12. Ross DA, Edwards MS, Wilson CB: Intramedullary neurilemmomas of the spinal cord: report of two cases and review of the literature. Neurosurgery 19: 458-464, 1986
13. Rout D, Pillai SM, RadhakrishnanVV: Cervical intramedullary schwannoma. J Neurosurg 58:962-964,1983
14. Russel DS, Rubinstein LS : Pathology of Tumours of the Nervous System. 2nd edition, London: E Arnold, 1963, 244 p.
15. Sarkar C, Mehta VS, Roy S: Intracerebellar schwannoma: case report. J Neurosurg 67: 120-123, 1987
16. Sharma R, Tandon SC, Mohanty S, Gupta S: Intramedullary neurofibroma of the cervical spinal cord: case report with review of the literature. Neurosurgery 15: 546-548, 1984
17. Slasky BS, Bydder GM, Niendorf HP, Young IR: MR imaging with gadolinium-DTPA in the differentiation of tumor, syrinx, and cyst of the spinal cord. J Comput A Tomogr 11 :845-850, 1987
18. Solomon RA, Handler MS, Sedelli RV, Stein BM: Intramedullary melanotic schwannoma of the cervicomedullary junction. Neurosurgery 20:36-38,1987
19. Şekerci Z, İyigün Ö, Kandemir B, Çelik F: Intramedullary schwannoma of thoracic spinal cord: case report. Turk Neurosurg 3:28-30, 1993
20. Takemoto K, Matsumura Y, Hashimoto H, Inoue Y, Fukuda T, Shakudo M, Nemoto Y, Onoyama Y, Yasui T, Hakuba A, Nishimura S, Ban S: MR imaging of intraspinal tumors-capability in histological differentiation and compartmentalization of extramedullary tumors. Neuroradiology 30:303-309, 1988