Cervical Intramedullary and Extramedullary Schwannoma

Servikal Bölgede Omurilik İç ve Dışında Yerleşen Schwannoma

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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

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 Th2. 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 fasciciles. 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 C2-Th1, b) sagittal post gadolinium T1 weighted image showing a high density well delineated mass lesion.

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Figure 2. Axial post gadolinium T1 weighted images, a) intramedullary location of the tumor at C2 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).

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 T1-weighted images and hyperintense in T2-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
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). Hypotheses 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 necessitate 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.

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REFERENCES