Magnetic resonance imaging of intramedullary spinal cord schwannomas

Report of two cases and review of the literature

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Intramedullary spinal cord schwannomas are rare benign tumors for which resection is possible and safe. The purpose of this paper is to present the magnetic resonance (MR) imaging features in two cases of intramedullary spinal cord schwannoma to assist both neurosurgeons and pathologists in preventing misdiagnosis and resultant partial resection. The MR imaging evidence of a small- or medium-sized well-marginated intramedullary spinal cord tumor in a patient in whom no syringomyelia is present but in whom moderate edema with marked Gd enhancement can be seen should be considered in the differential diagnosis of intramedullary spinal cord schwannoma. In cases in which an associated thickened Gd-enhancing spinal nerve root is seen the diagnosis of schwannoma should be assumed.

KEY WORDS • intramedullary spinal cord tumor • schwannoma • magnetic resonance imaging • differential diagnosis

Case Reports

Case 1

History. This 59-year-old man was admitted to another institution with a 1-year history of slowly progressive weakness and numbness in both legs and upper limbs and neck stiffness. The patient underwent T1-weighted MR imaging of the cervical spine, which demonstrated spinal stenosis extending from C-2 to C-7 and a small intramedullary nodule of low signal intensity at the C-2 level. A clinical examination produced normal results, with no signs of NF. Neurological examination revealed spasticity and deep tendon reflex, which were exaggerated in all four limbs. The patient underwent a C2–7 laminectomy, but only minimal improvement in his symptoms was observed. One year later, he was admitted to our department because of worsening spasticity and walking difficulty.

Examination. Magnetic resonance imaging of the cervical spine revealed persistent stenosis, despite the C2–7 laminectomy, and confirmed the presence of a small (4–5 mm in maximum diameter) intramedullary round mass, with moderate central edema of the adjacent cord at C-2 (Fig. 1). The lesion appeared isointense on T1-weighted images and slightly hypointense on T2-weighted images; it enhanced homogeneously with administration of Gd.

Operation and Postoperative Course. Reoperation consisted of removal of scar tissue from the previous laminectomy, dural opening, and midline myelotomy facilitated by ultrasonic guidance. Complete microsurgical removal
of a small gray–white tumor was accomplished. Pathological examination revealed a typical benign schwannoma composed mainly of Antoni A tissue. The postoperative course was uneventful. At 1-year follow up, the patient’s spasticity and numbness had disappeared, and he was able to walk without difficulty. The most recent MR imaging control study, performed 4 years after surgery, confirmed the total removal of the tumor.

Case 2

History. This 47-year-old woman was admitted to our department with a 1-year history of weakness in the right leg and walking difficulty. The results of her clinical examination were normal, with no signs of NF. Neurological examination revealed paraparesis and clonus, more marked on the right side, with no sensory level.

Examination. Magnetic resonance imaging of the thoracic spine demonstrated a sharply marginated intramedullary mass (1.5 cm in maximum diameter) with only minimal peritumoral edema at T-8 (Fig. 2). The tumor appeared isointense on T₁-weighted images and inhomogeneously hyperintense on T₂-weighted images; it enhanced homogeneously with Gd administration. After Gd administration a thin enhancing subarachnoid band connecting the intramedullary mass to the posterolateral dural wall was demonstrated on the right side, which may have represented an enlarged dorsal nerve root. This latter finding led to a presumptive diagnosis of intramedullary spinal cord schwannoma.

Operation and Postoperative Course. A T-8 laminectomy and dural opening was performed, and a localized fusiform expansion of the spinal cord was seen. A midline dorsal myelotomy was performed and revealed a well-encapsulated intramedullary lesion, which was dissected from the surrounding neural tissue and completely removed. An abnormally enlarged right dorsal nerve root appeared in continuity with the tumor and was resected during surgery. Pathological examination revealed typical schwannoma composed of both Antoni A and Antoni B tissue. In the early postoperative course, a minimal worsening of paresis in the right inferior limb was noted; however, the patient was able to walk with a cane. Six months later, after rehabilitation therapy, she was able to walk without support. Control MR imaging obtained 3 years after surgery, revealed no residual or recurrent tumor. A small intramedullary area of high signal intensity on T₂-weighted images, representing gliosis, as well as a minimal reduction of the sagittal diameter of the spinal cord was seen.

Discussion

Schwannomas (neuromas or neurilemomas) are the most common primary tumors of the spine, accounting for approximately 30%. They most commonly arise from nerve sheaths in the spinal canal; thus, usually their location is extramedullary intradural and/or extradural. Since the earliest report in 1931,9 approximately 60 intramedullary spinal cord schwannomas have been described in the literature.3,8,10,12,13,18,19 The lack of Schwann cells present in the normal spinal cord has been cited to explain the tumor’s rarity; however, small perivascular bundles of peripheral nerves normally occur within the spinal cord and are considered the logical source of intramedullary schwannomas.2,11,20 Other theories suggest the proliferation of Schwann cells ensheathing aberrant intramedullary nerve fibers7 or resulting from medial displacement of neural crest cells during embryogenesis17 from differentiation of multipotential mesenchymal elements such as pial cells of neuroectodermal origin,6,15 and finally from dorsal nerve root entry zone and centripetal growth.21 The latter hypothesis may explain the growth pattern observed in MR imaging in the patient in our Case 2, which was confirmed at surgery.
Intramedullary spinal cord schwannoma is usually a small well-encapsulated solid mass, and cystic and/or necrotic components are uncommon. At histological examination dense cellular Antoni A tissue is usually found, hypocellular Antoni B tissue is seldom seen, and melanotic features are rare.\textsuperscript{1,18}

Intramedullary spinal cord schwannomas develop most frequently in the cervical cord (62\% of cases), followed by the thoracic cord (22\% of cases), and lumbar cord, including the conus medullaris.\textsuperscript{13,18} There is no significant sex and age predilection. Despite the frequent association between spinal schwannomas and NF, only 12\% of patients with intramedullary spinal cord schwannomas are affected by NF.\textsuperscript{19} Neither of our patients was affected by NF.

Symptoms vary depending on tumor location. In both our patients, progressive motor and sensory disturbance were the symptoms appearing at onset.

At surgery, complete dissection of well-encapsulated schwannomas is reported in most cases.\textsuperscript{3,8,10,12,13,16,18,19} The use of intraoperative ultrasonography provides exact tumor location and guides the extent of myelotomy, and limits surgical trauma; thus, complete microsurgical tumor removal can be achieved with minimal risks and good clinical outcome. In both our patients, radical resection of the tumor resulted in clear improvement of symptoms at long-term follow up.

Because surgical removal is not feasible for infiltrating gliomas, even if it is attempted, preoperative neuroradiological and/or intraoperative biopsy–based differential diagnosis is crucial. At present, MR imaging is definitely the method of choice for the diagnosis of spinal cord tumors; however, a definitive histological diagnosis based on morphology or signal intensity remains difficult\textsuperscript{4,14} and differentiation from other intramedullary tumors (such as astrocytomias, ependymomas, hemangioblastomas, metas-tases) may be impossible.

Intramedullary spinal cord schwannomas are most commonly reported to be iso- or hypointense on T\textsubscript{1}-weighted images, and moderately hyperintense on T\textsubscript{2}-weighted images; however, MR imaging signal intensity depends on different macroscopic features (solid or cystic components, degenerative processes) or histological composition (pre-
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valence of Antoni A or Antoni B tissue, melanotic variant. Generally, tumors are well marginated, and perilesional edema is moderate. Administration of Gd allows better definition of the tumor from associated edema. Patchy enhancement has been reported in tumors with cystic and/or necrotic components. Associated syringomyelia is uncommon. A confident preoperative diagnosis is possible when a predominant extramedullary component is present or when the intramedullary spinal cord tumor is in continuity with a thickened spinal nerve root such as in the patient in Case 2.

The two cases presented here represent good examples of the variability of MR imaging findings of intramedullary spinal cord schwannomas. In Case 1, the MR imaging findings were completely nonspecific. In Case 2, the preoperative detection of a presumed thickened enhancing dorsal nerve root in continuity with an inhomogeneous intramedullary tumor led to the correct diagnosis.

In conclusion, MR imaging evidence obtained in a patient with a small- to medium-sized intramedullary spinal cord tumor, with central cord edema and without syringomyelia, revealing marked Gd-enhancement and sharp margins should raise suspicion of a schwannoma in differential diagnosis of intramedullary spinal cord tumors. Moreover, the diagnosis of intramedullary spinal cord schwannoma should be practically granted when a thickened Gd-enhancing spinal nerve root is seen in continuity with the tumor.

References


Manuscript received September 24, 3002. Accepted in final form March 28, 2003.

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