Case Report

Intramedullary Spinal Schwannoma: Case Report and Review of Preoperative Magnetic Resonance Imaging Features

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Intramedullary schwannomas are rare spinal cord tumours. Correct preoperative diagnosis is essential for proper surgical planning and complete resection. We present a case of cervical intramedullary schwannoma followed by discussion on its preoperative magnetic resonance imaging features and review of the literature. [*Asian J Surg* 2006;29(4):306–8]

Key Words: intramedullary schwannoma, magnetic resonance imaging, spinal cord tumour

Introduction

Nerve sheath tumours (schwannomas and neurofibromas) are the most common primary tumours of the spine, accounting for 30% of all intraspinal masses. They are usually intradural extramedullary or, less often, extradural in location. Intramedullary schwannomas are rare. Since the earliest report in 1931, only about 60 cases of intramedullary spinal cord schwannoma have been reported so far in the literature.[1] It represents about 0.3–1.5% of all primary intraspinal tumours.[2]

Since the risk involved in removal and the surgical strategy are very different for an intramedullary lesion and an extramedullary schwannoma, awareness of this possible diagnosis on imaging studies will be important to prevent misdiagnosis during surgical exploration or on frozen section histological examination.

We report a patient with an intramedullary schwannoma with histological confirmation. A brief review of the literature focused on preoperative imaging findings and clinical presentation are presented.

Case report

A 45-year-old man had a past history of nasopharyngeal carcinoma 13 years previously that was treated by radiotherapy. Follow-up magnetic resonance imaging (MRI) of the nasopharynx and neck in March 2004 incidentally showed a small ($11 \times 7 \times 12$ mm) intraspinal lesion at C5–6 level of the spinal cord. This lesion was well defined and located at the posterolateral aspect of the cervical cord on the right side. It was isointense on T1-weighted (T1W) images (Figure 1) and mildly hyperintense on T2W images (Figure 2). Marked homogeneous contrast enhancement was noted, no dural tail or cervical syrinx was detected, and a thickened and slightly enhancing nerve root was noted at the periphery of the lesion (Figure 3). No oedema was seen in the spinal cord. The lesion had both intramedullary and extramedullary components. The absence of syrinx formation and cord oedema, the absence of cord expansion and neurological symptoms and the presence of an enhancing nerve root and significant extramedullary component made it unlikely to be an

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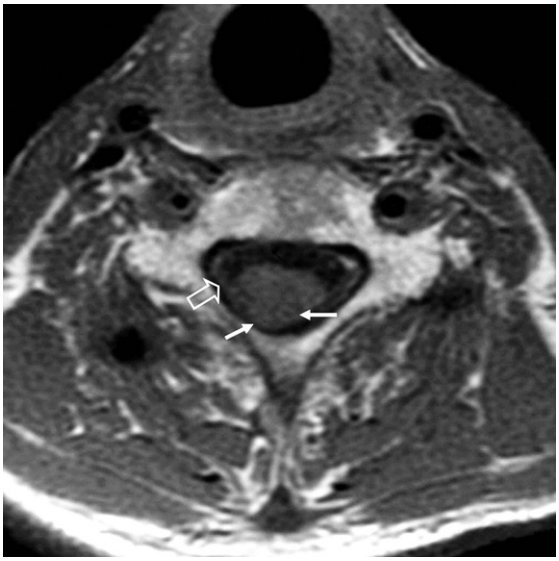

Figure 1. Axial T1-weighted magnetic resonance imaging of the cervical spine shows a well-defined isointense intraspinal lesion at the C5/6 level of the spinal cord (arrows). It has both intramedullary and extramedullary components. A thickened nerve root is noted at the right side of the tumour (open arrow).

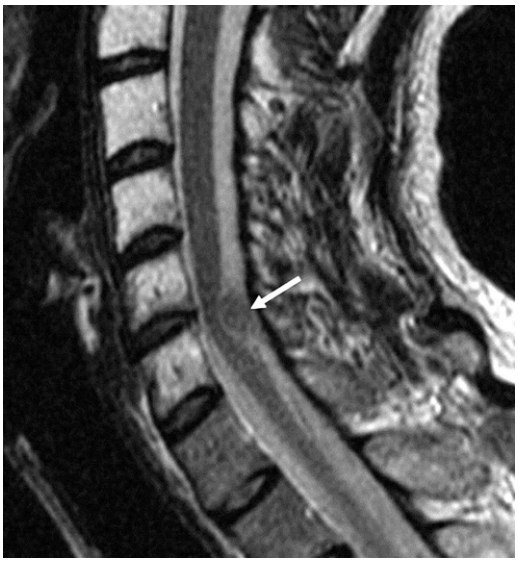

Figure 2. Sagittal T2-weighted magnetic resonance imaging of the cervical cord demonstrates the slightly hyperintense tumour at the posterolateral aspect of the cervical cord with significant extramedullary component (arrow). No cord oedema or syrinx formation is seen.

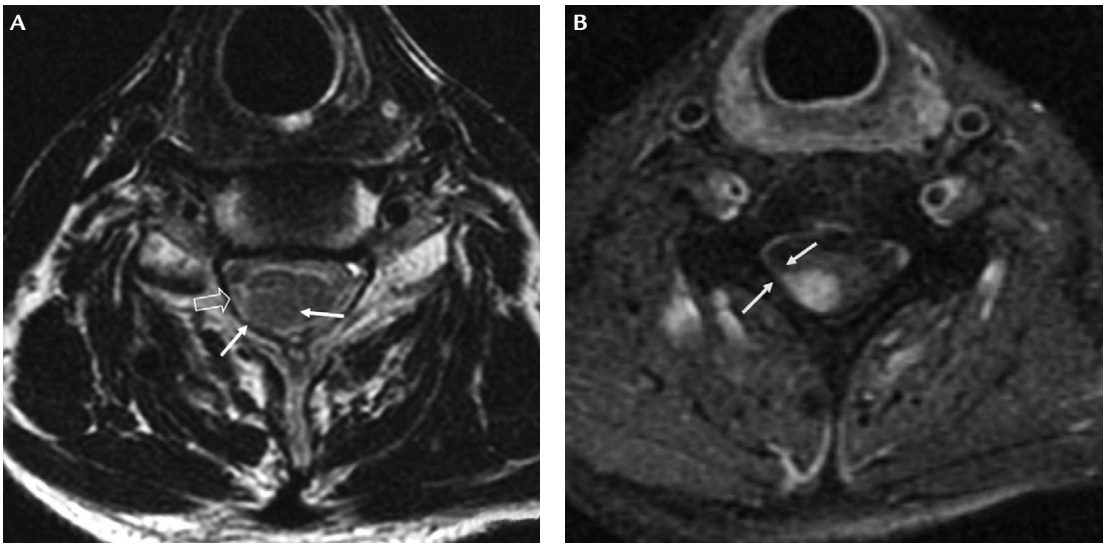

Figure 3. (A) Axial T2-weighted and (B) postcontrast fat saturated T1-weighted images of the cervical cord show the slightly thickened and enhancing nerve root at the right side of the tumour (open arrow). The tumour is well defined with homogeneous contrast enhancement (arrows).

intramedullary tumour. Therefore, the preoperative imaging diagnosis suggested an extramedullary tumour with possible intramedullary extension.

He showed no neurological symptoms or focal signs at the time of diagnosis. He underwent operation 4 days after the MRI examination. Intraoperatively, a yellowish intradural tumour was seen on the right posterior surface of the spinal cord extending into both C6 and C7 sensory rootlets (Figure 4). There was a significant intramedullary element. Frozen section findings were consistent with schwannoma. The lesion was dissected off the spinal cord tissue and total removal was achieved. Intraoperative monitoring included somatic sensory evoked potential through the median nerve and electromyography. The procedure was uneventful and the patient recovered well. At the 4-month postoperative follow-up, the patient only complained of occasional hyperaesthesia of the upper limbs and no other neurological deficits.

Histological analysis of the tumour confirmed the diagnosis of schwannoma with predominantly Antoni A

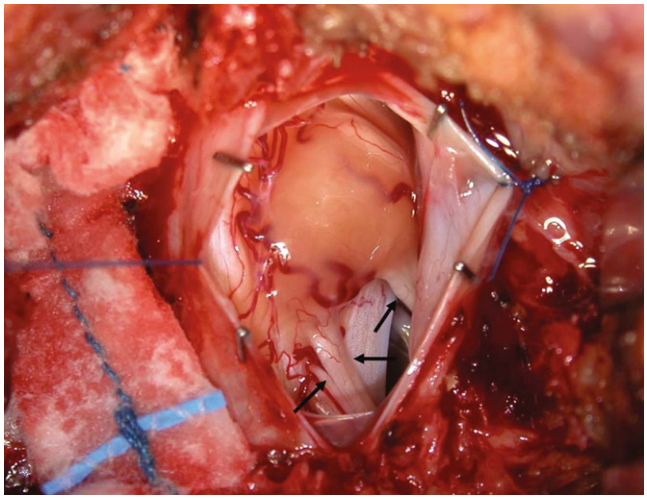

Figure 4. Intraoperative photo showing a yellowish tumour at the right posterior surface of the spinal cord, extending into both C6 and C7 sensory rootlets (arrows).

and focally Antoni B areas. A nerve bundle was also noted at the periphery of one tumour fragment.

Discussion

Schwannomas are well-encapsulated tumours, typically attached to nerve roots. Sites of the lesion are most frequently intradural, less frequently extradural or dumbbell-shaped.[4] Intramedullary schwannomas are rare in non-neurofibroma patients, accounting for 0.3% of intraspinal schwannomas and 1.1% of spinal schwannomas. About 12% of patients with intramedullary schwannomas are affected by neurofibroma.[1] Conti et al analysed the literature from 1931 to 2002 and found that there are only about 50 reported cases of non-neurofibroma related intramedullary schwannomas.[4]

The pathogenesis of intramedullary schwannomas is still controversial. Its rarity is explained by the absence of Schwann cells in the central nervous system. Postulated pathogeneses include central inclusion of Schwann cells during embryological development; Schwann cells vesting aberrant intramedullary myelin fibres; and extension of Schwann cells along the intramedullary perivascular nervous plexus.[1,4]

The most common site of spinal intramedullary schwannoma is in the cervical cord (62%), followed by the thoracic cord (22%) and then the lumbar cord and conus medullaris. No sex predilection is found. Mean age at presentation was 42.9 years.[2]

Symptoms vary depending on tumour location. Duration between first symptoms and diagnosis was usually long, with a mean period of 28.2 months (6 weeks–12 years). Complete dissection and tumour removal is reported in most cases.

MRI examination is the modality of choice for the diagnosis of spinal cord tumour. Typically, intramedullary tumours are centred and located in the spinal cord, causing expansion of the cord. They are commonly ill defined and associated with cord oedema and syringomyelia. This is contrary to extramedullary tumours, which are usually well defined, occasionally dumbbell-shaped and, most importantly, they displace the spinal cord rather than expand it. Also, a plane of cerebrospinal fluid cleavage is usually visible between an extramedullary tumour and the spinal cord. On MRI, intramedullary schwannomas are most commonly iso- or hypointense on T1W images, and moderately hyperintense on T2W images. Melanotic type of schwannomas may demonstrate high signals on T1W images.[2] The tumours are usually well marginated, with moderate perilesional oedema. Homogeneous or nodular contrast enhancement is seen in nearly all cases.[2] Associated syrinogomyelia is uncommon.[4]

A confident preoperative diagnosis is possible only if a predominant extramedullary component is present or when intramedullary spinal cord tumour is in continuity with a thickened and enhancing spinal nerve root.[4] Retrospectively, these findings were also present in the preoperative MRI of our patient.

In conclusion, in patients with intramedullary spinal cord tumour, MRI findings of absence of syringomyelia, marked contrast enhancement and sharp margins should raise the possibility of an intramedullary schwannoma. The presence of an extramedullary component and thickened enhancing spinal nerve roots in continuity with the tumour are further supportive evidence.

References

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