Neuroradiology (2001) 43: 567–571

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Intramedullary schwannoma of the spinal cord: report of two cases

Received: 8 September 2000
Accepted: 9 November 2000

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Abstract We report magnetic resonance findings in the intramedullary schwannoma of the cervical spinal cord in two patients. In both cases, the solid portions of the tumours enhanced intensely after administration of Gd-DTPA and the enhanced border was sharply delineated from the adjacent spinal cord. Schwannomas should be considered in the differential diagnosis of intramedullary tumours when magnetic resonance images show a strongly enhancing mass with sharply delineated borders.

Keywords Schwannoma, intramedullary · Spinal cord · Magnetic resonance imaging

Introduction

Schwannomas are the most common primary spinal neoplasms. They are usually intradural-extramedullary and/or extradural in location, and the diagnosis of this tumour is usually not difficult with magnetic resonance(MR) imaging. Schwannomas rarely occur in an intramedullary location [1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11], and preoperative diagnosis of intramedullary schwannoma based on imaging is difficult [1, 2, 3, 4, 5, 6]. We report two cases of surgically proven intramedullary schwannoma of the spinal cord and discuss the MR findings.

Case report

Case 1

A 37-year-old woman had progressive numbness of the right arm over a period of 9 years. She underwent a C1 laminectomy and C2–7 laminoplasty for a spinal cord tumour 4 years prior to her visit to our hospital and her symptoms improved. She was referred to us because of progressive numbness and weakness of the right arm again for 1 year. She also noted weakness in the right leg. Examination revealed weakness in all limbs, especially in right arm, bilateral Babinski and Chaddock reflexes, spastic gait and sensory disturbance below C4.

MRI at 0.5 tesla before the first operation showed enlargement of the spinal cord from the medulla oblongata to C7. Sagittal contrast-enhanced T1-weighted images showed a well-delineated mass at C3–5 with intense, fairly homogeneous contrast enhancement. A kink between the mass and the dorsal aspect of the spinal cord at C5 suggested exophytic growth. The lesion gave mixed slightly low and isointense signal on T1-weighted images, and heterogeneously high signal with small isointense portions on T2-weighted images. Areas of markedly high signal on the T2-weighted images cephalad and caudal to the enhancing lesion suggested oedema (Fig. 1a–c). MRI, repeated at 1.5 tesla after the symptoms worsened, revealed progression of the enlargement of the spinal cord. The enhancing area had shifted down to the C4–6 level, probably because of increased swelling of the caudal spinal cord and the posterior decompression. The enhancing mass had increased in anteroposterior dimension, and the caudal kink was barely discernible (Fig. 1 d).

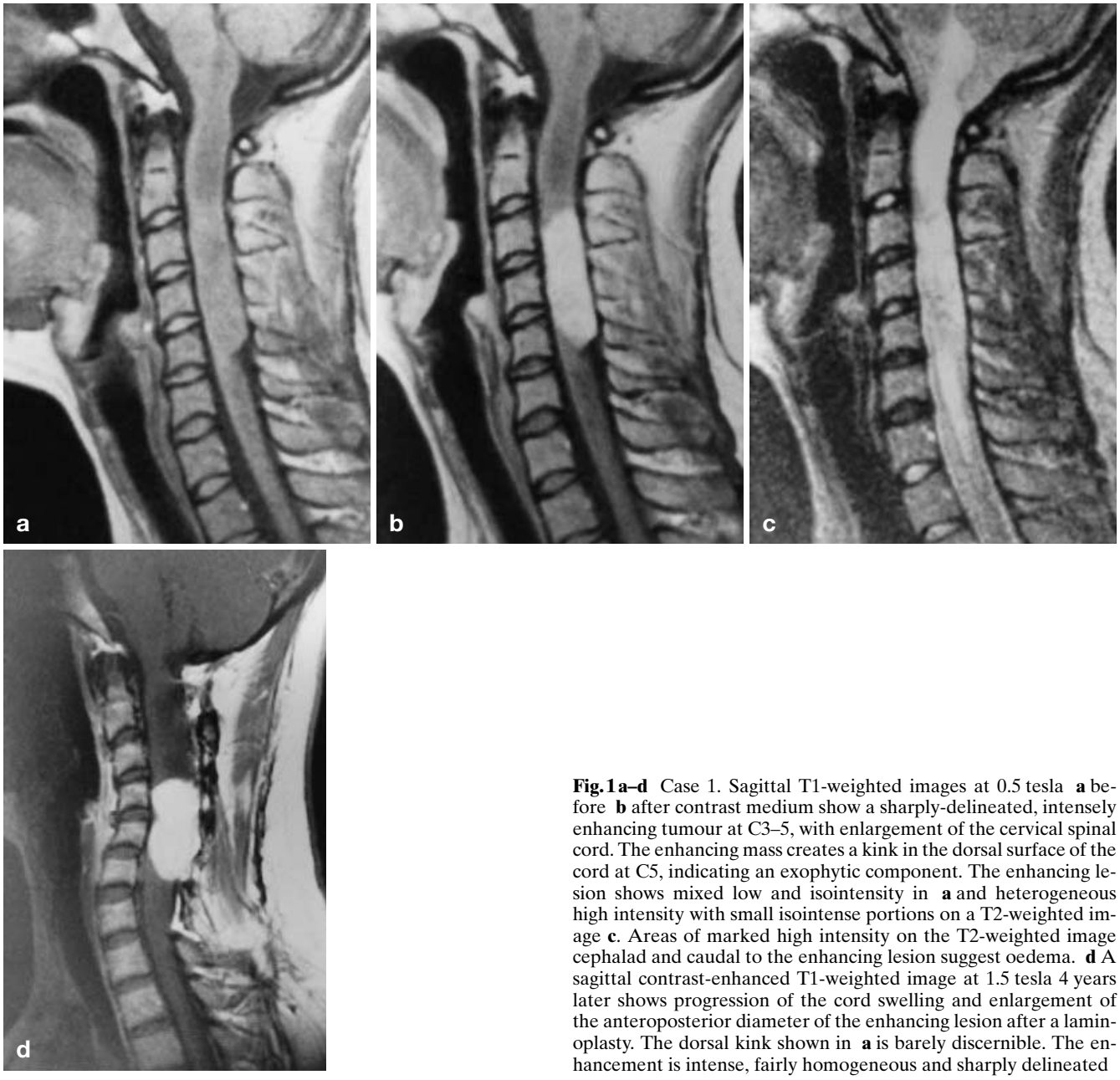

Fig. 1 a–d Case 1. Sagittal T1-weighted images at 0.5 tesla **a** before **b** after contrast medium show a sharply-delineated, intensely enhancing tumour at C3–5, with enlargement of the cervical spinal cord. The enhancing mass creates a kink in the dorsal surface of the cord at C5, indicating an exophytic component. The enhancing lesion shows mixed low and isointensity in **a** and heterogeneous high intensity with small isointense portions on a T2-weighted image **c**. Areas of marked high intensity on the T2-weighted image cephalad and caudal to the enhancing lesion suggest oedema. **d** A sagittal contrast-enhanced T1-weighted image at 1.5 tesla 4 years later shows progression of the cord swelling and enlargement of the anteroposterior diameter of the enhancing lesion after a laminoplasty. The dorsal kink shown in **a** is barely discernible. The enhancement is intense, fairly homogeneous and sharply delineated

With a presumptive diagnosis of spinal cord ependymoma, the patient underwent C3–6 laminectomy. After opening the dura mater, a hard, yellowish tumour was found just under the pia mater. A longitudinal incision of the latter revealed a subpial tumour. Most of its margins were well-demarcated from the spinal cord, but the middle of the tumour was densely adherent to the cord. Upon removal of the tumour, this fragment was left behind. The tumour was thought to originate from the medial dorsal subpial space. Histological examination revealed a typical schwannoma consisting of Antoni A and B tissue. After surgery the patient's symptoms and signs regressed.

Case 2

A 17-year-old girl had progressive gait disturbance, bilateral arm weakness and sensory disturbance of the left leg. She had mild gait disturbance 1 year prior to her first visit to our institution, which worsened to tetraparesis. Examination revealed weakness of the limbs, more marked on the left, and bilateral Babinski and Chaddock reflexes. Hypoalgesia and hypoaesthesia was detected in the left leg, along with disturbance of vibration and position sense. The patient could not stand still and her gait was spastic.

MRI at 1.5 tesla revealed diffuse enlargement of the spinal cord extending from the medulla oblongata to C6. Sagittal and axial contrast-enhanced T1-weighted images showed intense, sharply

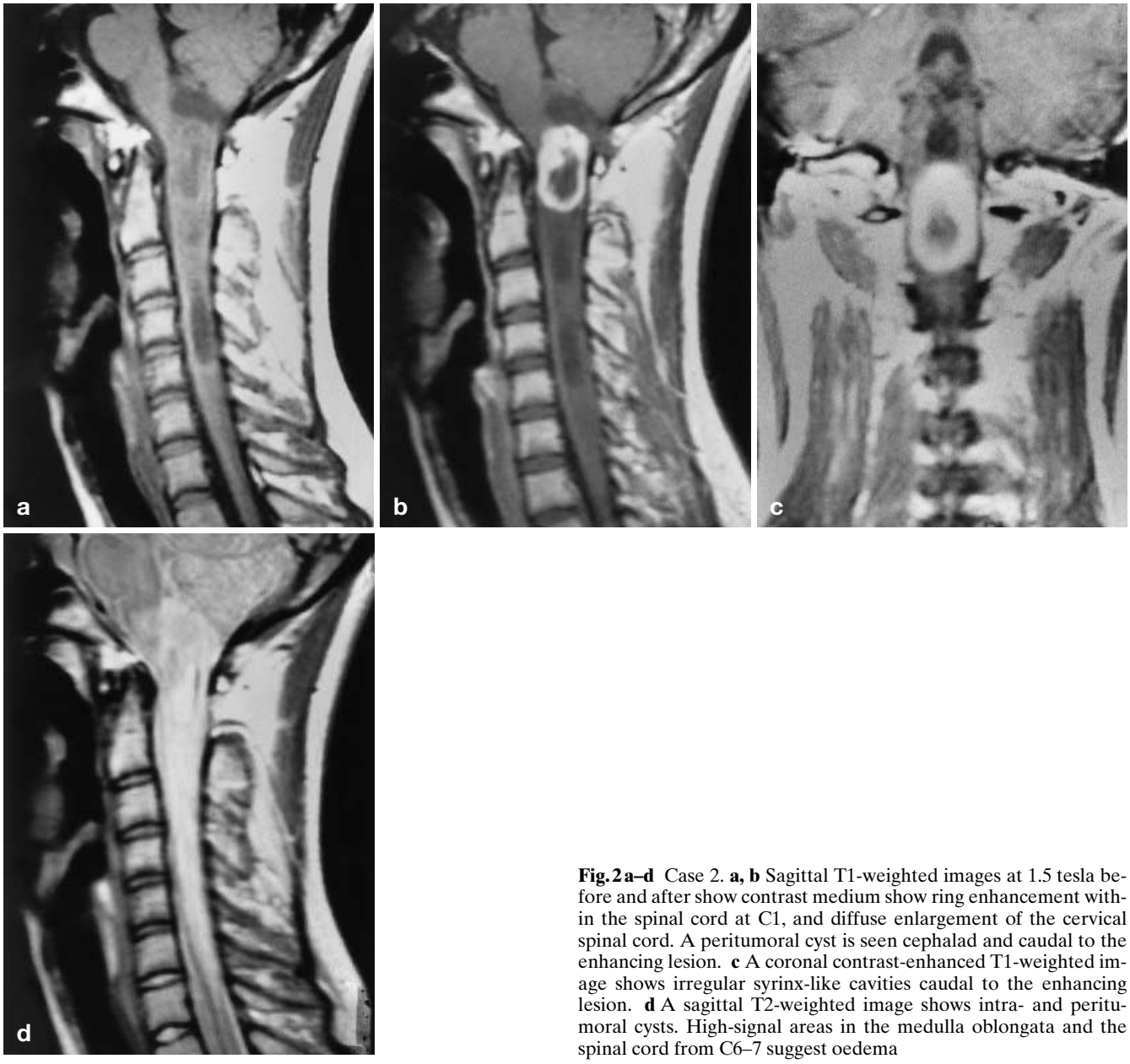

Fig. 2 a–d Case 2. **a, b** Sagittal T1-weighted images at 1.5 tesla before and after show contrast medium show ring enhancement within the spinal cord at C1, and diffuse enlargement of the cervical spinal cord. A peritumoral cyst is seen cephalad and caudal to the enhancing lesion. **c** A coronal contrast-enhanced T1-weighted image shows irregular syrinx-like cavities caudal to the enhancing lesion. **d** A sagittal T2-weighted image shows intra- and peritumoral cysts. High-signal areas in the medulla oblongata and the spinal cord from C6–7 suggest oedema

delineated intramedullary ring enhancement at C1 and nonenhancing cystic areas both cephalad and caudal to it. On T2-weighted images there were high-signal areas in the medulla oblongata and from C1 to C6 Fig. 2a–d). These findings suggested an intramedullary tumour with peritumoral cysts (or a syrinx) and oedema. The presumptive diagnosis was intramedullary schwannoma or ependymoma.

The patient underwent occipital craniectomy, C1 laminectomy, and C2 laminoplasty. Upon opening the dura mater, the spinal cord was found to be swollen. At the level of foramen magnum, there was a peritumoral cyst under the pia mater. A midline myelotomy revealed a hard, grey tumour containing a cyst. Its margin was slightly blurred because of gliosis. After the tumour was totally removed, the ventral pia mater was observed on the left. It was thought that the tumour arose in the left ventral subpial space. Histological examination revealed a typical schwannoma consisting of Antoni A and B tissue. After the surgery the patient's symptoms and signs improved.

Discussion

Intramedullary nerve-sheath tumours are rare, because the fibres in the central nervous system do not have Schwann cells. According to Ross et al. [1]., intramedullary schwannomas constitute 1.1 % of spinal schwanno-

mas. Drapkin [10], reviewing reports up to 1990, found 37 cases of nerve-sheath tumours within the spinal cord and brain stem. Other than neurofibromas, 31 cases had been reported up to 1990. We found six cases after that date, including our own [2, 3, 4]. Of these 37 cases, 15 occurred in males and 21 in females (data were not available in one case), and the age at presentation ranged from 12 to 75 years (mean 41.7 years). Intramedullary schwannomas were most frequent (51 %) in the cervical spinal cord, followed by the thoracic region (22 %).

The pathogenesis of intramedullary schwannoma is a matter of controversy because Schwann cells are absent in the brain and spinal cord in normal individuals. Hypotheses regarding the origin of intramedullary schwannomas include: 1. late neoplastic development of ectopic Schwann cells originating from the embryonic neural ridge during development; 2. Schwann cells ensheathing aberrant intramedullary nerve fibres; 3. Schwann cells extending along the branches of the anterior spinal artery; 4. neoplastic growth from dorsal-root Schwann cells in a "critical area" where posterior roots lose their sheaths on entering the pia mater; and 5. transformation of pial cells of neuroectodermal origin into Schwann cells [1, 2, 3]. Our surgical findings suggested that the origin of the tumour was in the dorsal (in case 1) or ventral (case 2) subpial space, and in case 2 the origin was probably around the exit zone of the anterior root.

MRI is the most useful method for diagnosing spinal intramedullary tumours [4, 11]. The findings in intramedullary schwannoma have been described in 10 cases. The tumour was mainly cervical in eight and mainly thoracic in two. Sagittal images showed enlargement of the spinal cord. The tumour was usually accompanied by peripheral oedema and/or peritumoral cystic cavities so that the enlargement of the spinal cord was usually diffuse. An exophytic component was present in some cases. On T1-weighted images, the tumour usually gave lower signal than the spinal cord, with or without isointense areas within it. Of the 10 cases two were melanotic and gave predominantly high signal on T1-weighted images. On T2-weighted images, the tumour usually showed inhomogeneous high signal, with occasional isointense or low-signal areas. The margins varied from poorly to well delineated on both T1- and T2-weighted images, depending on the signal intensities of the tumour and the adjacent spinal cord. The tumour showed marked enhancement in all six cases in which contrast medium was given; the degree of the enhancement was not detailed in two cases, but the illustrations indicated that it was intense. The enhancement was homogenous or patchy; the latter was attributed to a cyst within the tumour at least in one case. The margin of the enhancing mass was well delineated in three cases; in the remaining three cases report did not describe the margin of the tumour, but once again the illustrations indicate that the enhancing mass was also sharply delineated.

These signal intensity and enhancement patterns were thus identical to those of intradural extramedullary schwannomas. Pathologically, the latter are usually heterogeneous, having both densely cellular Antoni type A and hypocellular Antoni type B areas. Cystic degeneration or necrosis is common and small haemorrhages and fatty degeneration may occur [11, 12, 13]. This heterogeneity explains the variable appearances on MRI [7, 8, 11, 12, 13, 14, 15]. Schwannomas usually show intense contrast enhancement, probably because the open-gap junctions are short, straight, and patent, freely communicating with a relatively large extracellular space [16, 17].

Although the number of reported cases is small, we believe that the characteristic findings of intramedullary schwannomas are the intense enhancement and well-delineated margins of the tumour on contrast-enhanced T1-weighted images. As we had already seen our first case, our presumptive diagnosis in case 2 included intramedullary schwannoma. Due to the rarity of the intramedullary schwannomas, they may be difficult to differentiate from gliomas during surgical exploration and even on frozen section [3]; they have been misdiagnosed as pilocytic astrocytomas [3, 9]. Since complete surgical removal is not attempted in the case of infiltrating gliomas, correct diagnosis from biopsy specimen is crucial, and alerting the neurosurgeons and pathologists to the possibility of an intramedullary schwannoma preoperatively is therefore important to avoid misdiagnosis from the frozen sections.

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