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LETTER TO THE EDITOR

# Intramedullary malignant peripheral nerve sheath tumor

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## To the Editor

Primary spinal cord tumors represent 2–4% of central nervous system (CNS) tumors. Intramedullary tumors predominantly include diffuse astrocytomas and ependymomas [1]. Intramedullary schwannomas are rare and account for 0.3–1.5% of all spinal schwannomas [2]. Thus far, no primary intramedullary malignant peripheral nerve sheath tumor (MPNST) has been described. Only a post-radiation intramedullary MPNST was described, in 2006, with metachronous laryngeal cancer [3].

We report the case of a 56-year-old man who developed a sensory impairment in his left leg and in his right arm over a 6-month period. Cervical magnetic resonance imaging (MRI) revealed a C2–C3 intramedullary, heterogeneous, contrast-enhancing lesion (Fig. 1a, b). Neurological examination revealed sensory loss of the lower extremities and of the right arm. No significant motor defects were detected. Patellar reflexes were increased.

### Surgical treatment and postoperative course

Surgery was carried out with continuous sensory and motor function monitoring. The lesion was located in the posterior aspect of the spinal cord, just underneath the dura, and appeared firm and white. Posterior myelotomy was performed, and tumor removal was apparently complete. No clear margins were discernible between the tumor and the spinal cord. During the final surgical maneuvers, intraoperative motor function monitoring revealed a sudden, severe decrease of muscle motor evoked potentials (MEPs) in all extremities, especially on the right side, while D-wave showed a decrease of up to 50%. Surgery was immediately stopped before performing further samplings of the surgical field walls, because of the high risk of permanent impairment. However, resection was complete, as confirmed by postoperative MRI showing gross total resection of the lesion, and a very thin, decompressed spinal cord (Fig. 1c–e). After surgery, the patient presented with tetraparesis that gradually improved during the following weeks. Two months after surgery he could walk with cane. MRI confirmed satisfactory tumor removal. Iphosphamide was given as adjuvant chemotherapy. No recurrences were detected on cerebrospinal MRI 9 months after surgery (Fig. 1f).

### Histological findings and diagnosis

Histopathological examination showed a malignant spindle cell neoplasm featuring extensive hypercellular areas with limited collagenous areas. No necrosis was found. Mitotic count revealed up to 6 mitoses/10 high-power fields (HPF). S100 protein was focally positive in tumor cells, whereas CKCAM 5.2, epithelial membrane antigen (EMA), collagen IV, desmin, myogenin, and human melanoma black (HMB)-45 were completely negative. Glial fibrillary acidic protein (GFAP) and neurofilament stains were limited to the underlying cord parenchyma infiltrated by the tumor (Fig. 2). Molecular immunology borstel (MIB)-1 labeling was moderate. Fluorescence in situ hybridization analysis (probe LSI SS18 Break Apart FISH probe kit, Vysis) did not reveal rearrangements in the *SYT* gene.

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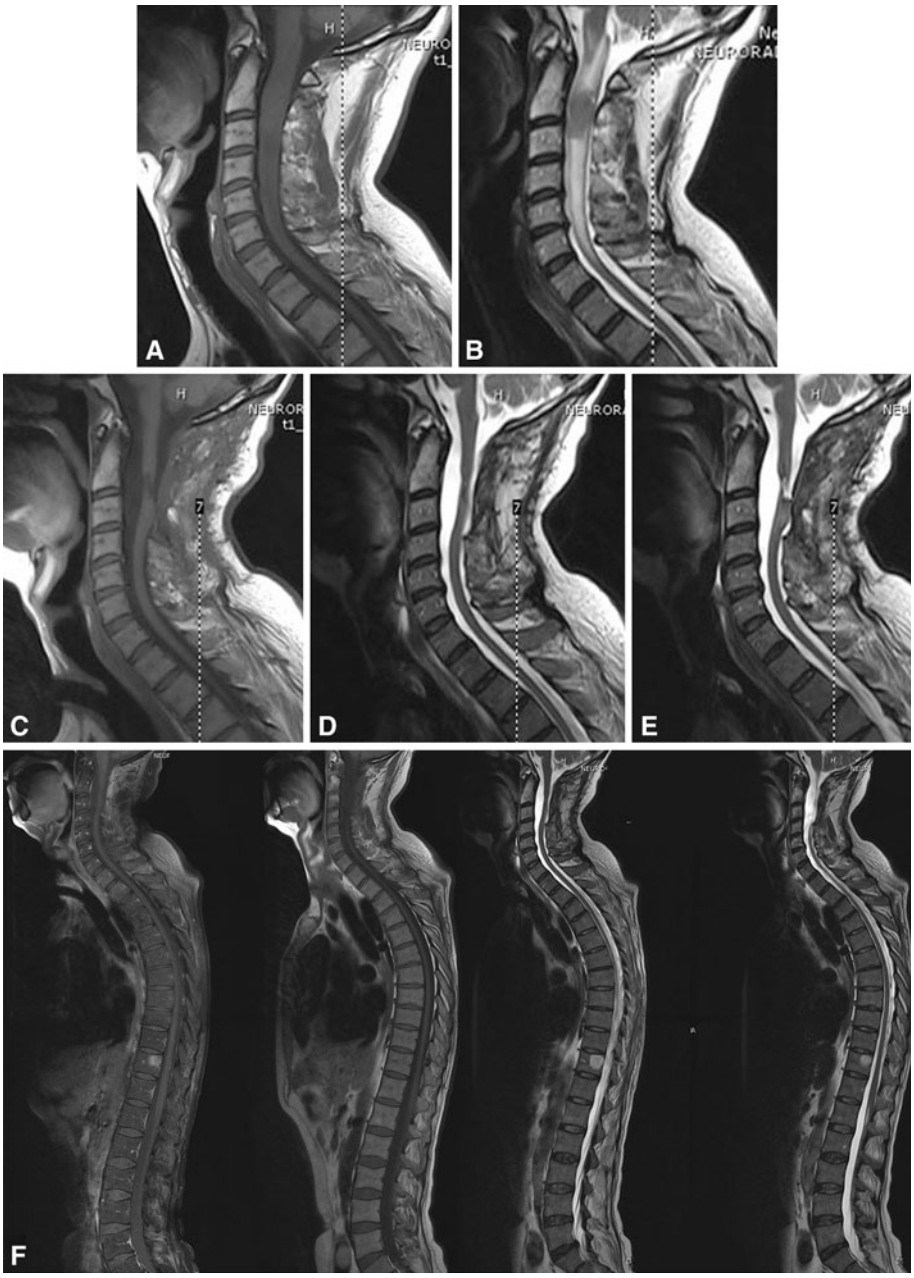


**Fig. 1** Preoperative **a** T1- and **b** T2-weighted sagittal magnetic resonance imaging (MRI), showing a C2–C3 intramedullary heterogeneous lesion. Postoperative **c** T1- and **d**, **e** T2-weighted sagittal MRI, confirming resection of the lesion. **f** T1 and T2 sagittal spinal MRI performed 9 months after surgery: no relapses were detected

Intramedullary tumors are predominantly represented by diffuse astrocytomas and ependymomas [1]. Intramedullary schwannomas probably arise from clusters of Schwann cells embedded in the medullary parenchyma [2]. They are slow-growing benign tumors that generally cause back pain without specific symptoms. Neuroimaging is also nonspecific, and the diagnosis is often attained only after pathological examination.

Primitive intramedullary MPNSTs have not been described in literature, with the sole exception of one postradiation case [3]. MPNSTs are rare soft-tissue sarcomas with a tendency for recurrence and metastasis. They are resistant to conventional therapies, and their deep-seated position and locally invasive growth hinder complete surgical resection. MPNSTs respond poorly to conventional chemo- and radiotherapies, and effective alternative therapies are not yet available.

Patients with neurofibromatosis type 1 (NF1) develop more neurofibromas, at younger age, and have increased risk of MPNSTs [4]. Our patient did not show any of the typical cutaneous hallmarks of NF1 disease, and was therefore considered sporadic.

In the reported case, neuroimaging suggested the possibility of intramedullary ependymoma (hypointense T2 signal, isointense T1 signal, with diffuse and homogeneous enhancement after gadolinium injection). Intraoperative

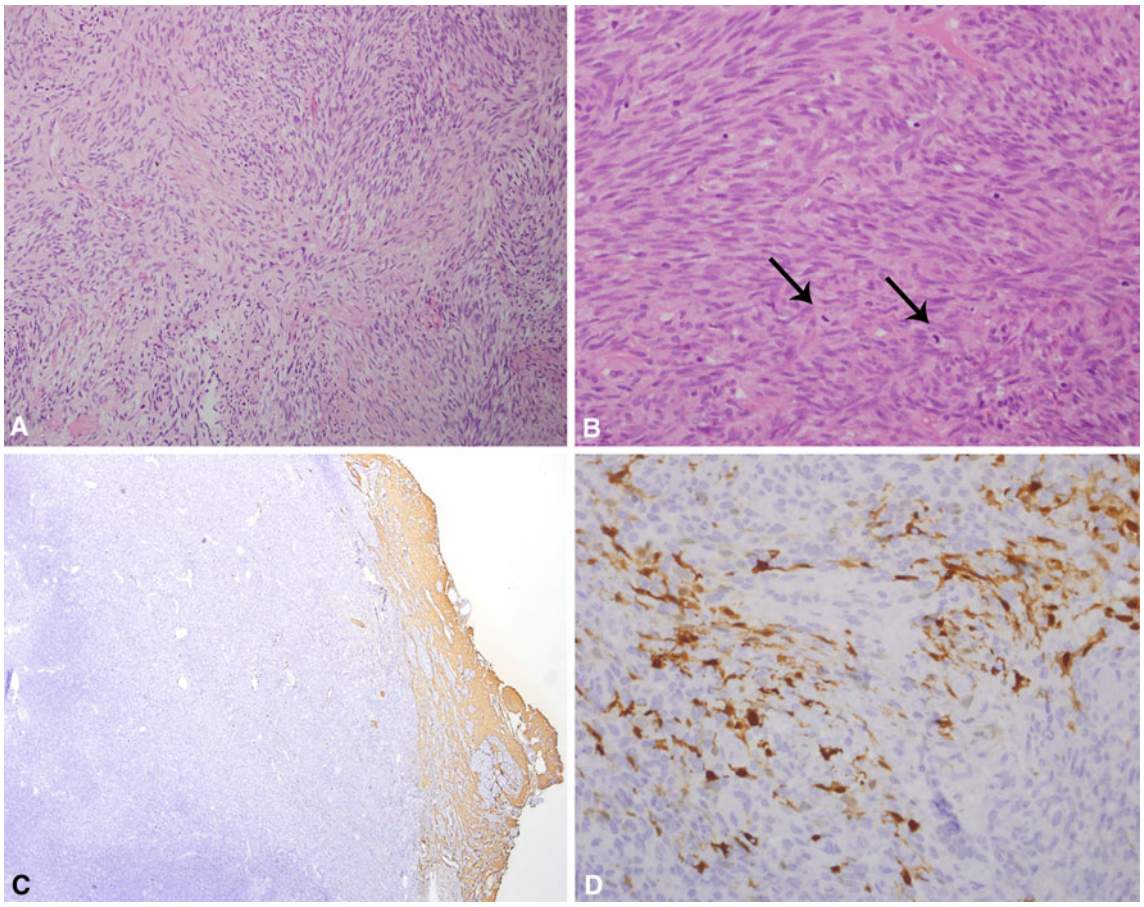


**Fig. 2** The tumor was composed of spindle cells and featured more extensive cellular areas associated with focal collagenous areas (**a**, 20×). At high power, brisk mitoses were seen (**b**, *arrows*, 40×). The neoplasm was intramedullary, as shown by GFAP, which stained the normal tissue surrounding the tumor (**c**, 5×). S100 was focally expressed (**d**, 40×)

macroscopic findings were not specific. However, the histological features of the neoplasm, as well as the focal expression of S100 protein, were consistent with the diagnosis of MPNST. Furthermore, the absence of expression of epithelial markers such as EMA and CKCAM 5.2, along with the absence of *SYT* rearrangement, ruled out the possibility of synovial sarcoma, very rarely described in the spinal cord [5]. Moreover, the absence of collagen IV, as well as the focal staining pattern of S100 protein, excluded the possibility of cellular schwannoma, which could also enter into the differential diagnosis.

To date, this is the first reported case of intramedullary MPNST. Similarly to other rare tumor types, immunohistochemistry proved to be of utmost value for objective diagnosis of this unusual spinal cord tumor.

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