

Extramedullary melanotic schwannoma recurrence in the cervical vertebral arch: a case report and review of the literature

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Abstract

Melanotic schwannoma (MS), a slowly growing nerve sheath tumor, is not a purely benign tumor. MS accounts for less than 1% of all nerve sheath tumors. We herein describe a rare case of MS and present a literature review focusing on the treatment of this disease. Twelve years before presentation at our hospital, a 41-year-old woman was examined because of an 8-month history of neck pain and 6-month history of upper extremity numbness and weakness. She underwent surgery to remove a tumor, and the pathological examination confirmed a diagnosis of MS. Twelve years later, at 53 years of age, the patient presented to our hospital with a 2-year history of neck pain and upper extremity numbness and weakness. Posterior cervical tumor resection was performed along with posterior cervical laminectomy, decompression and intraspinal space-occupying internal fixation, and radiotherapy. MS recurrence was confirmed. No tumor recurrence or metastasis was found after 7 months of follow-up. Recurrence of MS is rare, and its diagnosis depends on pathological features. Radical excision is the primary treatment for MS. Incomplete resection of MS is a risk factor for postoperative recurrence and metastasis. Furthermore, postoperative adjuvant radiotherapy should be performed to prevent recurrence and metastasis of MS.

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Keywords

Melanotic schwannoma, tumor, recurrence, radiotherapy, pathological diagnosis, surgical resection

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Introduction

Melanotic schwannoma (MS) is a tumor of intermediate dignity. A rare neoplasm, MS accounts for less than 1% of all nerve sheath tumors and has a propensity for spinal nerve involvement.^{1,2} Fewer than 200 cases of MS have been reported to date. MS affects patients aged 10 to 92 years, but the peak age at onset is 30 to 40 years, and no sex predilection exists.³ We herein describe a rare case of cervical epidural MS that required surgical resection for local recurrence.

Case presentation

Twelve years before presentation to our hospital, a 41-year-old woman was examined because of an 8-month history of neck pain and 6-month history of numbness and weakness of the upper extremities. Neurological examination revealed diminished skin sensation of the upper extremities. Positive pathological reflexes were confirmed using the Spurling test and Eaten test. Muscle strength was assessed using a muscle strength grading scale. Grade 4/5 musculus biceps brachii strength, grade 3/5 musculus triceps brachii strength, and grade 3/5 grip were found upon physical examination. The positive pathological reflexes that could be elicited were patellar tendon hyperreflexia and the bilateral Babinski sign. Magnetic resonance imaging (MRI) of the cervical spine showed hypointense masses in the spinal canal at the C2 to C4 vertebral bodies on T1- and

T2-weighted imaging. A tumor was located in the epidural space. The mass was about 4.0×1.1 cm in size, and the local spinal cord was compressed. A benign tumor was considered; however, the nature of the tumor required pathological confirmation. The patient had already experienced symptoms of central compression, and medical treatment was ineffective. Surgical treatment was required to remove the tumor. Posterior cervical tumor resection was performed. Intraoperative examination revealed that the tumor, which had a cystic substance of about $3.0 \times 2.0 \times 1.5$ cm, was located at C2 to C3 in the right epidural space. The neoplasm was adherent to the epidural membrane, and the capsule, which was filled with black sticky masses, was still intact and dark black. After exposure, the dura mater was found to be free of tumor invasion. Histological examination showed clusters of plump, spindled, and heavily pigmented tumor cells (Figure 1). Thus, the diagnosis of MS was made. The patient was able to work normally after the first operation. However, she refused to undergo another MRI examination of the cervical spine after the first operation.

Twelve years later, at 53 years of age, the patient presented to our hospital with a 2-year history of neck pain with numbness and weakness of the extremities. These symptoms had worsened during the most recent 6 months. Neurological examination revealed that the skin sensation of the upper extremities was significantly reduced. The skin sensation diminished below the

umbilical plane of the trunk, and the diminishment continued to the lower extremities. Muscle strength was assessed using a muscle strength grading scale. Grade 4/5 muscle strength of the extremities was observed during the physical examination. The positive pathological reflexes were the bilateral Hoffmann sign, bilateral Babinski sign, bilateral biceps hyperreflexia, and patellar tendon hyperreflexia. On MRI of

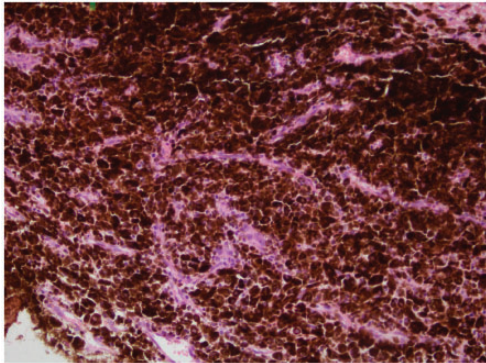


Figure 1. Surgical specimen collected during the first operation was examined by hematoxylin and eosin staining (200× magnification). Clusters of plump, spindled, and heavily pigmented tumor cells were found.

the cervical spine, the C1 and C2 vertebral bodies in the right spinal canal exhibited a semicircular mixture composed of a slightly long T1 hypointensity and slightly longer T2 signal shadow (dominated by T2 hypointensity signal). The border of the spinal canal was still clear and adjacent to the dural sac, and the spinal cord was compressed. The possibility of MS recurrence was considered (Figure 2).

The patient underwent posterior cervical laminectomy for decompression and intra-spinal space-occupying internal fixation with the spinal cord compressed. During this operation, scar tissue was found at the distal end of the C2 lamina. The right side of the interlaminar space of C1 and C2 was occupied by a 1-cm × 2-cm mass of soft tissue, which was black and lobulated and had a clear boundary and deep base into the vertebral canal. The upper boundary of the tumor reached the level of the C1 posterior arch, and the lower boundary of the tumor reached below the C2 lamina and adhered to the surrounding scar tissue. The medial side of the tumor was closely associated with the epidural space, and the border in the right posterolateral part of the dura was

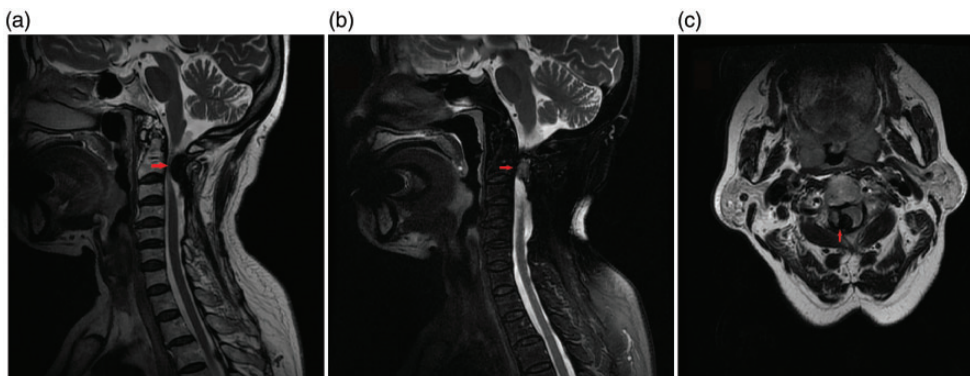


Figure 2. Magnetic resonance imaging examination of cervical spine before the second operation showed that the C1 and C2 vertebral bodies in the right spinal canal exhibited a semicircular mixture containing (a) slightly long T1 and (b) slightly longer T2 signal shadow (based on short T2 signal). (c) The border was still clear and adjacent to the dural sac, the spinal cord was compressed, and the accessory parts on the right side of C3 and C4 were absent. No abnormal signal was observed in the medullary cavity.

not clear. With the assistance of bipolar coagulation, the lesions in the spinal canal were carefully separated. The C2 nerve root was not resected intraoperatively. After tumor resection, no evidence of rupture was observed in the dura. Given that the border of the tumor in the right posterolateral part of the dura was not clear, bipolar electrocoagulation was performed around the operative field on the edge of the tumor bed to secure the interruption of the blood supply during the second operation. The last part of the tumor was totally removed (Figures 3 and 4).

After the operation, the patient's symptoms of limb numbness and weakness were alleviated. The muscle grade in the limbs was 5/5. Postoperative radiographic examination of the cervical spine showed stable internal fixation (Figure 5). The tissue was removed for pathological examination, which showed clusters of plump, spindled, and heavily pigmented tumor cells (Figure 6). The schwannian nature of the cells was confirmed by electron microscopy. Thus, the diagnosis of MS was confirmed.

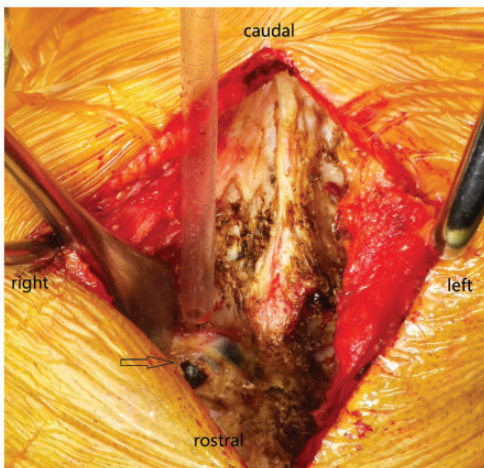


Figure 3. Operative view during the second operation. A mass lesion was found in the right posterolateral epidural space near the vertebral bodies of C1 and C2.



Figure 4. The mass measured about $1.0 \times 1.5 \times 3.0$ cm and was black.



Figure 5. The spinous processes of C2 and C3 were mostly absent, and both sides of the vertebral arches showed a longitudinal series of metal rods and screw shadows, indicating stable internal fixation.

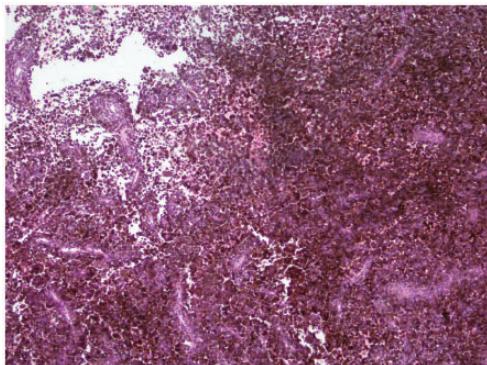


Figure 6. Surgical specimen collected during the second operation was examined by hematoxylin and eosin staining (200× magnification). Clusters of plump, spindled, and heavily pigmented tumor cells were found.

Considering the recurrence of MS and the fact that the dura of C2 was not resected, radiotherapy was performed to prevent further recurrence. No tumor recurrence or metastasis was found after 7 months of follow-up.

This report was approved by Yantai Yuhuangding Hospital (Approval No. [2019]161). The patient provided written informed consent.

Discussion

MS occurs in the paraspinal nerve roots (especially at the level of the cervical and thoracic spinal nerve roots), lumbar vertebrae, and sacrococcygeal region.^{4–6} In 1932, Millar⁷ first described this finding as a malignant melanotic tumor of the ganglion cell, which was subsequently named MS by Fu et al.⁸ in 1975.

MRI is the preferred method for assessing spinal cord injury. The boundaries of the tumor are usually clear on MRI. Given the paramagnetic effect of melanin, MS exhibits hypointensity on T2-weighted images and hyperintensity on T1-weighted images. The hyperintensity on T2-weighted images is a feature of

melanotic tumors but only occurs in about 25% of all melanotic cases. The uneven distribution of melanin in the tumor and the presence of a hematoma can lead to uneven signals in the lesion, making MRI interpretation more complex.^{9–11} Schwannomas are easily confused with intraspinal melanotic tumors. The imaging findings of a primary intraspinal melanocytoma are characterized using short T1 and T2 signals (72%).^{12,13} MRI in our patient showed T1 hypointensity and a short T2 signal in the vertebral bodies of C2 to C4. These results were consistent with an MRI diagnosis of MS. However, early diagnosis remains difficult, and postoperative pathological examination is still required.

The diagnosis of MS in this case was based on histopathologic examination. Microscopically, most MSs are solitary lesions that are round or ovoid, multiple, and multicentric. Notably, Carney complex is occasionally observed in patients with MS. The cut surface of the sectioned tumor has the consistency of tar and is usually pigmented, ranging from brown to uniformly black.¹⁴ The pigment of the cut tumor surface is uniformly black during the second operation. Histopathologically, MS is cellular and non-encapsulated, and the tumor is lined externally by a thin fibrous capsule. MS is composed of closely packed pigmented, plump spindle and epithelioid cells arranged in interlacing fascicles or nests. The diagnosis of MS is suggested by a predominantly spindled morphology, vacuolated (adipocyte-like) cells, psammoma bodies, heavy melanin pigmentation, and striking nuclear pleomorphism with a relatively low mitotic rate.¹⁵ Following surgery, the pathological findings are used to confirm the diagnosis of MS. The tumor cells and macrophages are positive on Fontana–Masson staining as evidenced by brown to black granules and negative on Prussian blue and periodic acid–Schiff staining.^{1,16,17} Half of patients

with MS have Carney complex, whereas the other half of patients have MS exhibiting psammoma bodies (psammomatous MS). Multiple studies have failed to identify any distinguishing morphological features. Elevated mitotic activity may predict malignant behavior.¹⁵ Comparison of the pathologic findings in the present case with those in previously reported cases showed that the resected tissue was consistent with the manifestation of MS. However, melanin is abundant in MS, and the immunohistochemical effect is poor. Thus, immunohistochemical examinations cannot be performed.

Radical excision, which is the primary treatment for MS, is important in preventing tumor recurrence or metastasis.¹⁸ The tumor has a tendency to recur locally and metastasize.^{19,20} Recurrence of MS is only found in partially excised tumors.^{4,21} The blood supply of the tumor should be divided before the tumor is completely removed. Adhesion of the tumor to the dura mater, pia mater, and spinal cord should be carefully separated during the operation.¹¹ If the tumor is closely adhered to the spinal cord, a residual tumor capsule should be left to prevent spinal cord injury caused by forced stripping, and postoperative radiotherapy should be performed.^{4,22} Stereotactic spine radiosurgery may also be performed to treat spinal MS; this involves the use of highly focused radiation beams to target the spinal tumor.^{1,23} In the present case, the locally recurrent MS was grossly resected again, but clear margins could not be achieved. MS treated with adjuvant radiotherapy has a low rate of recurrence and metastasis. Radiotherapy should be considered in cases of tumor recurrence, metastasis, histologic criteria of malignancy, or incomplete surgical resection.^{3,12} Thus, even when adjuvant radiotherapy is administered to avoid relapse, close observation should be continued. Following the operation and adjuvant radiotherapy in the

present case, the patient's limb numbness and weakness were alleviated. The muscle grade of the limbs was 5/5, and the patient was able to work normally. No tumor recurrence was found during the 7-month follow-up period.

Conclusions

Recurrence of extramedullary MS in the spinal canal is rare, and its diagnosis depends on the pathological features. This condition should be distinguished from melanoma. The present case illustrates that incomplete resection of the tumor is a risk factor for postoperative recurrence and metastasis. Therefore, the intraoperative blood supply should be disconnected from the tumor, and the tumor should be completely removed. For patients with severe adhesion and incomplete resection, postoperative adjuvant radiotherapy should be performed to prevent recurrence and metastasis of MS.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

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