



## Epidural Melanocytic Schwannoma in Lumbar Vertebra: A Case Report

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### Abstract

Melanotic Schwannoma (MS) is a kind of potentially malignancy, which is rare in clinical practice, with a low incidence. We report a case with lumbar vertebra MS admitted to the Department of Neurosurgery in the First Affiliated Hospital of Nanjing Medical University in June 2022. This patient hospitalized at local hospital several times. After receiving medication, his symptoms had worsened. MRI examination shows the symptom was caused by the tumor compressing the nerve root. After surgical treatment, the patient's pain symptoms had improved significantly. The patient discharged 5 days after surgery. The patient has been living normally so far.

**Keywords:** Melanocytic Schwannoma; Lumbar vertebra; Epidural

### Introduction

MS is very rare, which was first reported by WG Millar in 1932, accounting for less than 1% of the total number of schwannomas [1]. A cytogenetic analysis related to MS showed that the karyotype of the tumor cell was diploid with obvious chromosome aberration, mainly showing trisomy of chromosome 6 and ring of chromosome 11. The long arm (6p) gain of chromosome 6 and the short arm (11q) gain of chromosome 11 are common genetic features of malignant melanoma [2]. Compared with MS, the common schwannoma is mainly the loss of genetic material on chromosome 22, which leads to the inactivation of NF2 allele [3], while the abnormality of chromosome 6 and 11 has not been found in cases with traditional schwannoma. Therefore, MS has also been considered as a separate melanoma in recent years, which is different from a schwannoma previously considered.

MS is common in young and middle-aged people, with a median age of 38 years old [4]. There is no significant difference in the incidence of MS among different genders. MS originated from neural crest, which is a kind of cells with differentiation potential of melanocytes and nerve sheath cells at the same time. It is common in spinal nerve roots and sympathetic ganglia, cervicothoracic segment, and other parts such as skin and soft tissue of head and neck, digestive tract, cervix and retroperitoneum have also been reported [5]. Its clinical manifestations are nonspecific, mainly manifested as shoulder, neck, waist and back pain. MS is mainly caused by tumor stimulating spinal nerve root, and sensory and functional disturbance of corresponding segments occurs when spinal cord is compressed, which is similar to compression symptoms caused by disc herniation or other space-occupying lesions in spinal canal.

### Case Presentation

The patient, a 56-year-old male, was admitted to the hospital with aggravation of right lower limb numbness and pain for more than 1 year and 15 days. The patient suffered from right lower limb numbness and pain without obvious inducement 1 year ago, which was aggravated after activities and could be relieved after rest. However, no attention was paid to the disease at that time. In the last half a month, the patient felt that her symptoms had worsened and her pain was unbearable at night (VAS score: 7 points). The lumbar enhanced MRI scan of local hospital hinted that: L5-S1 posterior intravertebral canal mass on the right side, considering neurogenic tumor. The patient came to our hospital for further treatment. The physical examination hinted that: Cranial nerve tests were negative, normal muscle force and muscle strength, pathological signs were negative, Achilles tendon reflexes were normal, right Straight leg lifting test was positive, with significant pain with slight numbness on the lateral side of the right calf and the dorsum of the foot. The lumbar enhanced MRI scan of our hospital demonstrated that: Hyperintense on T1 (Figure 1A, 1D) and hypointense on T2 (Figure 1B) on the right side of the spinal canal at the level of the L5 vertebra,

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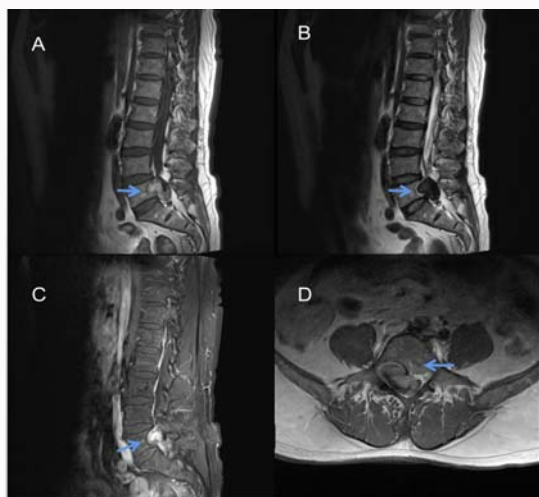
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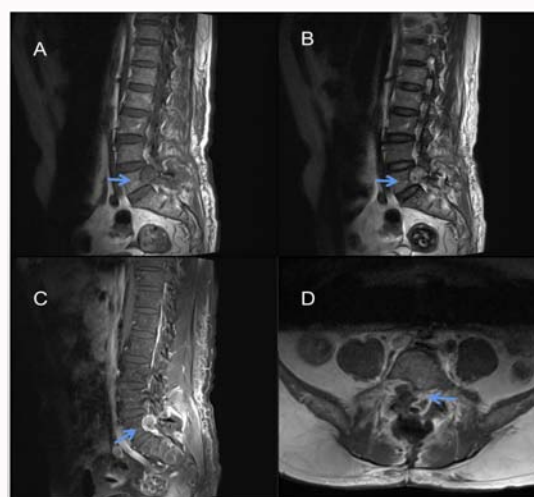
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**Figure 1:** Preoperative MRI plain scan + enhanced examination results. A) High T1 signal at the L5 vertebral body level (sagittal section); B) Low T2 signal at the L5 vertebral body level (sagittal section); C) Obvious T1 enhancement (sagittal section), the size was about 3.5 cm × 1.7 cm; D) The right intervertebral foramen at T1 (transverse section) was enlarged, which was closely related to the right L5 nerve root and showed dumbbell-shaped changes.



**Figure 3:** MRI plain scan + enhanced examination results 5 days after operation during hospitalization. A) High T1 signal at the L5 vertebral body level (sagittal section); B) Low T2 signal at the L5 vertebral body level (sagittal section); C) Obvious T1 enhancement (sagittal section), the size was about 3.5 cm × 1.7 cm; D) The right intervertebral foramen at T1 (transverse section) was enlarged, which was closely related to the right L5 nerve root and showed dumbbell-shaped changes.



**Figure 2:** Partial tumor specimens removed during operation with a black tarry appearance.

with significant enhancement (Figure 1C), the size of about 3.5 cm × 1.7 cm, enlargement intervertebral foramen, which is closely related to the right L5 nerve root, with dumbbell-like change. Lumbar spine CT scan showed that: strip-like shadow of soft tissue density on the right side of the vertebral canal at the level of L5 vertebral body, The vertebral body is deformed due to compression. X-ray of lumbar hyperextension and flexion showed that: No lumbar instability. Complete preoperative examination was improved; after eliminating surgical contraindications, surgery was performed under general anesthesia. During surgery, C-arm fluoroscopy located the L5 and S1 vertebra, 3 cm posterior median straight incision, paravertebral muscles were separated, L5 spinous process and vertebral body were exposed, L5 spinous process was removed. By grinding drill, milling cutter was used to remove L5 lamina and expose Dural sac. Exploration and investigation showed that the tumor was located outside the spinal dura mater, with clear boundary, complete capsule and black surface, extending to L5 and S1 intervertebral foramen. Under the surgical microscope, black viscous tissue was found in the capsule, which was tarry (Figure 2). Rapid pathology during the operation suggested melanoma. First of all, most of the tumor

in the capsule was removed, and then the adhesion with L5 nerve root was carefully separated along the capsule, and the tumor was completely removed. After complete hemostasis, the reduction and fixation of lamina was carried out, and muscles, subcutaneous and skin were sutured layer by layer. On the first day after operation, the pain in the original pain area was significantly reduced (VAS score for 3 points), the lower limbs were normal, and there was no new complaint of discomfort. Five days after operation, MRI plain scan and enhancement of lumbar spine showed that there were no residual tumors after operation (Figure 3). Postoperative routine pathology showed that melanin-rich lesions and pigmented schwannoma should be considered to be discharged; Immunohistochemistry showed that the tumor cells were S-100 (+), SOX10 (+), HMB45 (+), MelanA (+), Ki67 (focus +). The pathological diagnosis was malignant melanin nerve sheath tumor, formerly known as MS. The patient recovered well after operation and was discharged from hospital on the 5<sup>th</sup> day. The pain was obviously relieved compared with that before operation (VAS score 1). After operation, the patient sought medical treatment in the oncology department, and it was recommended to conduct reexamination regularly. The patients have lived normally since telephone follow-up.

## Discussion

Total surgical resection is recognized as an effective means to treat MS at present, and radiotherapy and chemotherapy can be supplemented after total resection. Currently, it is difficult to predict the prognosis of this disease. By retrospectively analyzing 40 cases, a literature pointed out that the recurrence rate in situ is 35% and the metastasis rate is 44% [6]. According to the reports of a few recurrent patients at home and abroad, it is speculated that the recurrence, metastasis and malignant transformation of the disease may be related to tumor residue, morphological heterogeneity and infiltration [7]. Therefore, during the operation, the surgeon should try his best to cut the tumor completely and decompress it fully on the premise of protecting the peripheral blood vessels, spinal cord and

nerves. There is no unified conclusion on adjuvant treatment except surgery. It has been reported that radiotherapy has certain effects on tumor treatment and prevention of recurrence [8], which provides some basis for the follow-up treatment of patients after operation. Italiano reported a case without radical surgery in 2011. The patient found tumor recurrence two months after operation, and after chemotherapy, the tumor progression was delayed to a certain extent [2].

Plain and enhanced MRI scan plays an important role in preoperative diagnosis; CT examination can understand the invasion of tumor to vertebral body and whether the intervertebral foramen area is enlarged, which is helpful for the formulation of surgical plan. The MRI features of MS are complicated, which is related to the different location and content of melanocytes in the tumor. The paramagnetic radical of melanin in cells can reduce the relaxation time of T1 and T2 [9], namely short T1 and short T2. According to the melanocyte content in tumor and T1 and T2 manifestations of MRI, foreign scholars divided MS into four types: Melanin type, non-melanin type, mixed type and hemorrhagic type, most of which are melanin type in clinical practice [10]. The histological features of common melanin type are >10% melanocytes in lesions, and MRI features are short T1, short T2, that is, high signal intensity in T1 and low signal intensity in T2, which are mostly round, oval or dumbbell-shaped. Enhanced examination can indicate mild, moderate or obvious enhancement.

The diagnosis of MS mainly depends on microscope, electron microscope observation and immunohistochemistry. Microscopically, MS showed epithelioid, spindle-shaped cells arranged in bundle, wheel or interweaving, which were similar to Antoni A area of schwannoma. The nucleus was round or oval, with obvious nucleoli and unusual mitosis. If obvious atypia, more division and necrosis infiltrated surrounding tissues were found, malignancy should be considered [5]. Fontana-Masson melanin silver staining was positive and PAS staining was negative in tumor cells. Under electron microscope, macrophages rich in mature melanin bodies were scattered, and a large number of glycogen granules were deposited in the nucleus. In ultrastructure, continuous basement membrane, long-spaced collagen fibers and other Schwann cells were found, and some of them contained melanin bodies [11]. In immunohistochemistry, most patients showed positive S-100, HMB-45, Melan-A, Leu-7 and NSE detection results. Tumors may be accompanied by hemorrhage, cystic degeneration, necrosis, calcification and sandy bodies. MS can be distinguished from primary or metastatic melanoma, melanoma, pigmented neurofibroma and cellular blue nevus according to their microstructure and immunohistochemistry.

In conclusion, MS is a potentially malignant tumor, and its etiology, treatment and prognosis are unclear, which remains to be further studied. At present, the only recognized effective treatment

is total surgical resection. Radiotherapy can play a certain role in preventing recurrence after operation and patients without total tumor resection during operation. Due to the high recurrence rate and metastasis rate of this disease, regular follow-up and reexamination play an important role in understanding the disease condition, determining the treatment plan and evaluating the prognosis.

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Haosong Xu collected the data and wrote the report. Jia Xu, revised and polished the article. Xiaoming Lu gave professional opinion on the content of the article. All authors involved in managing patients.

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