

Recurrent Upper Cervical Spinal Intermediate Grade Melanocytoma: A Case Report and Literature Review

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Abstract: Melanocytoma is a rare primary pigmented neoplasm of the central nerve system, which commonly is associated with the leptomeninge. Typically, it is considered benign, malignant transformation of these lesions is even rarer. Spinal lesions often present as intradural extramedullary masses that are compressing adjacent neural structures to produce various neurological signs. Complete resection is the treatment of choice for spinal melanocytomas. Even with complete resection, recurrences are common and close followup is needed for long term outcomes. Radiation therapy should be reserved for those cases in which complete resection is not possible or there is recurrence.

Keywords: Melanocytoma; spine; surgical treatment.

INTRODUCTION

Melanocytoma is a benign lesion arising from leptomeningeal melanocytes, which composition of variably melanin producing cells. The lesion at times can mimic its malignant counterpart, melanoma, and recurrence is not unusual [1]. The neoplasm is rare which accounting for less than 1% of all nerve tumors. They are most commonly seen in supratentorial and thoracic spine extramedullary locations with spinal cord compression symptoms. These symptoms are similar with myelopathy or cerebral infarction, which makes the clinical diagnosis not always obvious [2].

The primary melanocytic neoplasms of the CNS are consisting of a lineage of tumors ranging from well-differentiated melanocytomas to the overtly malignant melanoma with aggressive histopathological features. However, there were a few melanocytic tumors whose characteristics cannot be defined into either variety. These were classified as intermediate-grade melanocytic tumors, with some cellular atypia and a considerable recurrence rate [3].

CASE REPORT

History

A 36-year-old man with history of 2 times of cervical melanocytoma resection surgery presented to our department for 4 weeks of muscle weakness. 4 years earlier, he complained numbness of both upper limbs for 2 months and received a C3-5 laminectomy surgery for resection of cervical neoplasm at the local hospital. Interestingly, his histopathological report was melanotic schwannoma. (Fig. 1) 3 years later, the numbness recurred with muscle weakness of both upper limbs. Pre-operative MRI revealed an extramedullary intra-dural tumor compressing the spinal cord dorsally at the axis level, which was hyper-intense on T1-weighted images and hypo-intense on T2-weighted images. (Fig. 2) Additional C2 laminectomy with internal fixation from C2-5 was employed. The second

histopathological report was melanocytoma. After the surgery, he regained his muscle strength and daily living. However, 1 year later, he was referred to our department for the second time of symptom recurrence. On hospital admittance, his chief complains was numbness and weakness of both upper and lower limbs. He denied any other systemic diseases. He was conscious and did not have any difficulty of communication.

Examinations

Physical examination revealed decreased motor and sensory function below neck of both side. The manual muscle strength test demonstrated 4/5 strength on the left and 3/5 on the right. Increase of deep tendon reflex was noted. Babinski sign was positive bilaterally. Muscle tonus of all extremities was increased. Fortunately the dysuria function was not impaired. The pre-operative MRI revealed an intradural extramedullary mass compressing the spinal cord at C2-3 levels (Fig. 3). The mass almost had the similar signal intensity as the second pre-operative MRI on T1 and T2 weighted images (Fig.2). Contrast enhancement revealed a heterogeneous enhancing. Pre-operative laboratory examinations consisted of complete blood count, electrolytes, liver, renal function tests, and urinalysis. They were all normal. With the presence of

typical MRI findings and histopathological reports, the diagnosis of melanocytoma was concluded, a rare neurologic neoplasm of the spine. Failure of intral-fixation was discovered by pre-operative X-ray pictures (Fig. 4).

Surgical treatment

The C2-3 levels of spinal compression were considered to be responsible of clinical symptoms. Surgical posterior decompression of axis lamina and re-fixation was performed. The dural was thin and expended. After opening the dura, immediately obvious was the very dark lesion, similar to charcoal. The frozen sections revealed a melanotic tumor. As the tumor moderately adherent to the surrounding spinal cord, a subtotal resection was performed. After resection, the dural was too thin and fragile which was not suitable for repair, and then a wide opening was left. (Fig. 5) The wound was closed in the usual multi-layered fashion. He was treated postoperatively with a rigid hard plastic collar that had to be worn constantly for 12 weeks.

Pathological findings and Postoperative course

Histopathological examination confirmed the lesion to be intermediate grade melanocytoma. Pathology specimens were immunohistochemically stained with Melan-A, HMB-45, S-100, and CK

polynestic, Vimentin, CD34, EMA, collagen IV, GFAP, and Ki-67. Melan-A, HMB-45, S-100, Vimentin, CK polynestic were positive, Ki-67 was 5-10% (+), Vimentin, CD34, EMA, collagen IV, GFAP was negative (Fig. 6).

As there was obvious leakage of cerebrospinal fluid, the drainage was removed 1 week later, with a suture of the drainage site. Fortunately, all of the wounds were healed. Two weeks later, his muscle tonus was deceased and muscle strength was improved. The manual muscle test demonstrated 5/5 strength on the left limbs and 4/5 on the right limbs. After rehabilitation, the patient could stand up depended on himself. After an uncomplicated hospital course, he was discharged home 2 weeks later. After follow-up for 3 months, there was no recurrence of neck pain, neurologic examination was normal.

The design and performance of this study conformed to ethical standards of Helsinki Declaration and our national legislation. It was approved by Medical Ethical Committee of our institution. The patient was enquired whether or not willing to take part in a scientific research and informed consent forms were signed by him.



Fig-1: Post-operative X-ray pictures of the first time



Fig-2: pre-operative X-ray pictures of the second time surgery

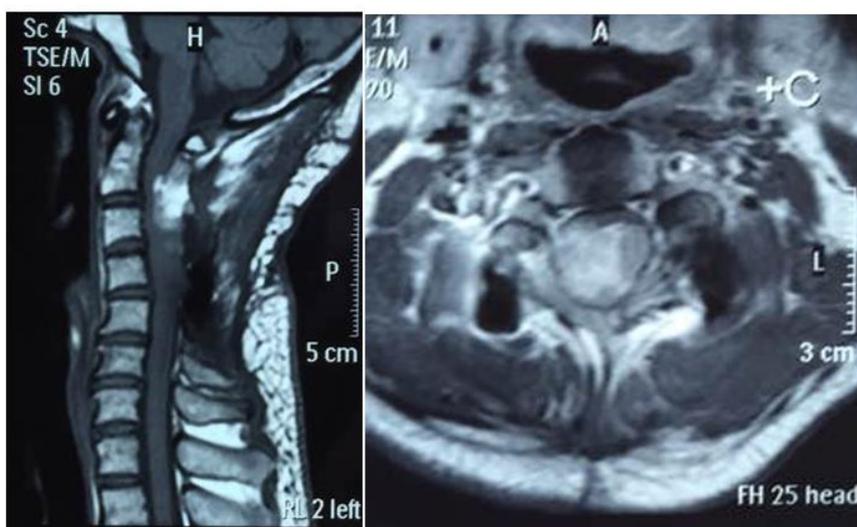


Fig-3: pre-operative T1 image of the third time

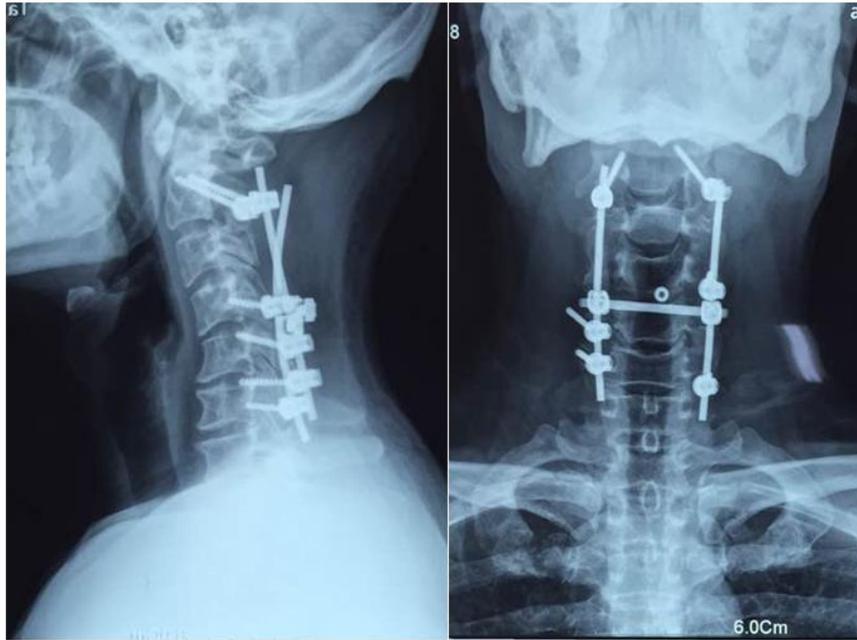


Fig-4: pre-operative T1 image of the third time

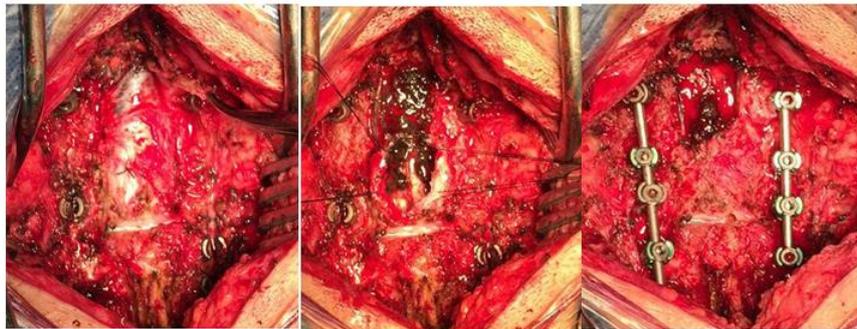


Fig-5 Intra-operative images of the third time

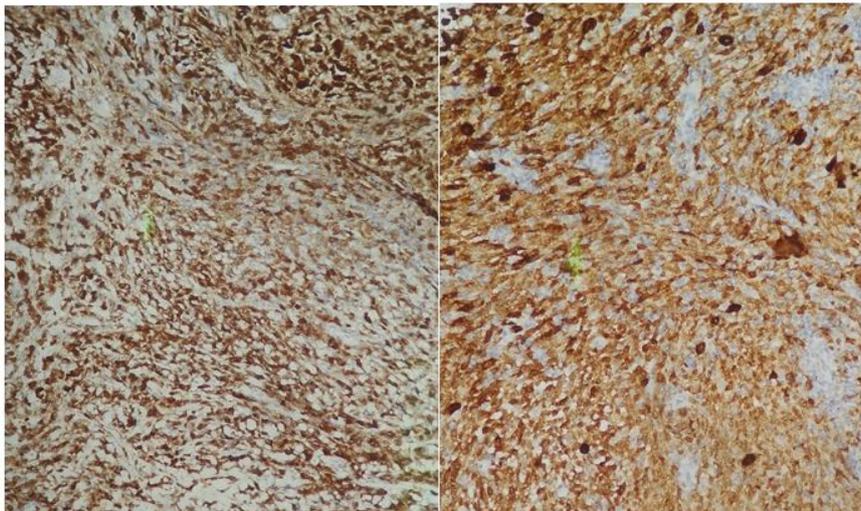


Fig-6: Photomicrograph of an S-100 (A) and Melan-A (B) stained section of the spinal melanocytoma resected in this patient

DISCUSSION

Neural crest is the originate site of melanocytes, therefore, a lot of neuromelanin producing cells are located in the central never system, such as pars compacta of substantia nigra and locus ceruleus. However, Melanocytic neoplasms of the CNS are rare and generally represent malignant and metastasis. These neoplasms are composed by a heterogeneous group of tumors which include benign melanocytomas, melanotic schwannomas, melanotic meningiomas, melanotic ependymomas and highly aggressive metastatic malignant melanomas [3, 4]. Melanocytomas are typically benign solid tumors that come from leptomeningeal melanocytes in the central neuraxis [5]. These lesions are exceedingly rare, with no more than 100 cases reported in the literature. Approximately 50% of the cases are found intracranially and the other half in the spinal canal. Although the majority of lesions are associated with the dura, a small percentage is either intramedullary or adherent to the outer surface of the spinal cord [6, 7].

Melanocytomas are slowly growing tumors that usually present with myelopathy. In rare cases, however, they may have small hemorrhages and present with signs of superficial siderosis [8, 9]. These neoplasms are most commonly found as solitary lesions, but can present as multifocal lesions or be disseminated along the arachnoid and dura mater[10-12]. Less commonly, a single nerve root can be involved, which can confuse the diagnosis with melanotic schwannoma [13, 14].

The MRI features of meningeal melanocytoma are uniformly hyperintense on T1-weighted and moderately hyperintense on T2-weighted images. Enhancement was not appreciable as the tumor is already hyperintense on T1-weighted images. The MRI features of melanocytoma are similar with melanotic schwannomas and melanotic meningioma's which should be taken into differential diagnosis consideration.

The diagnosis of meningeal melanocytoma was described by Limas and Tio in 1972, who were the first to recognize that the cells of origin of this tumor type are melanocytes in the leptomeninges as opposed to meningotheial cells [5]. Thus the term meningeal melanocytoma has become the standard nomenclature used to describe this tumor type. The pathological characteristics of this tumor are that of a well demarcated lesion with a gray to black appearance. Histologically, these tumors contain spindle or epithelioid cells with little or no cellular pleomorphism and rare mitoses. Histopathological analysis also reveals immunoreactivity for HBM-45 and S-100 protein but not for epithelial membrane antigen [3, 15]. MIB-1 count is useful in deterring proliferation of tumor cells and evaluates the possibility of local recurrence, despite the histology.

In 1999, Brat *et al.* [3] introduced a new pathological diagnosis: intermediate-grade melanocytic tumors which distinguished itself from well-differentiated melanocytomas and the malignant melanomas. In his spectrum, one end of the spectrum was melanocytoma with benign histopathological characteristics and low MIB-1 proliferation index; the other end of the spectrum was the malignant melanoma with aggressive histopathological features and elevated MIB-1 count. The intermediate-grade was defined as some cellular atypia and intermediate MIB counts. In our report, the multi-recurrence biological behavior of the tumors correlated well with the histopathological differentiation of the lesions [16].

Surgery is the primary mode of treatment for spinal melanocytomas. Local control rates have been shown to be four times higher if complete resection is achieved. Several elegant compilations of the existing case reports by Rades have clearly shown that GTR offers a greater survival benefit and a lower recurrence rate compared with subtotal resection [17, 18]. Without question, a complete resection provides a greater disease-free survival duration than an incomplete resection, but this is just not possible in cases in which the tumor is either adherent to the surface of the spinal cord or is completely intramedullary. Morbidity and mortality rates for incomplete resection now mandate the use of adjuvant radiation therapy. In addition, this benefit is more favorable with doses of 45 Gy[19].

In our own cases of intramedullary spinal melanocytoma, the patient suffered a significant recurrence for two times. Radiation treatment was not used initially in any of these cases because there was no radiotherapical equipment at local hospital. After subtotal resection and follow-up for 3 months, there was no recurrence of neck pain, neurologic examination was normal. Very close post-operative monitoring and diligent follow-up is crucial, which allows for early adjuvant therapy or re-resection

CONCLUSIONS

Melanocytomas are benign tumors, which originate from leptomeningeal melanocytes and clinically present with myelopathy. HBM-45 and S-100 protein are positive in immunoreactivity tests, but not for epithelial membrane antigen. MIB-1 count is useful in deterring proliferation of tumor cells and evaluates the possibility of local recurrence. Intermediate MIB counts with some cellular atypia are the indicator of intermediate-grade melanocytoma. Complete resection offers a greater survival benefit; however, recurrence is not unusual. Close monitoring and diligent follow-up is critical.

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