Intramedullary melanotic schwannoma: Case report and review of the literature

Szu-Yen Pan, Yu-Ching Cheng¹, Ting-Hsien Kao

Departments of Neurosurgery, and ¹Radiology, Taichung Veterans General Hospital, Taichung, Taiwan, R.O.C.

E-mail: *Tinghsien Kao - joshuakth@gmail.com; Szuyn Pan - psyen@sghc.gov.tw; Yuching Cheng - cat.hello@yahoo.com.tw
*Corresponding author:

Received: 11 March 14  Accepted: 23 May 14  Published: 16 July 14

Abstract

**Background:** Intramedullary melanotic schwannomas are very rare. Only seven cases have been previously reported. This is the first reported case of such a tumor presenting with intratumoral hemorrhage closely mimicking a hemorrhagic ependymoma.

**Case Description:** A 23-year-old female patient presented with progressive right lower extremity pain and numbness for 3 months and this was followed by acute paraparesis and urinary incontinence. Physical examination revealed sensory and motor dysfunction below T4 level. Magnetic resonance imaging of her spine showed a heterogeneous signal lesion occupying the intramedullary space with small foci of hyperintensity on T1WI and hypointensity on T2WI at the T4-5 level. She underwent emergent surgical decompression and microsurgical excision of the tumor. A histopathological diagnosis of melanotic schwannoma was made. Postoperative neurological recovery revealed progressive improvement in her examination findings.

**Conclusion:** Among spinal cord neoplasms, melanotic schwannomas are extremely rare tumors. The similar source of Schwann cells and melanocytes, the neuroectoderm, has been suggested to be the origin of melanotic schwannomas. Intramedullary melanotic schwannomas presenting with hemorrhage tend to be mistaken with ependymoma, which by far is the most common spinal tumor to present with hemorrhage. Total surgical excision if safely possible is currently considered the ideal treatment of choice.

**Key Words:** Intramedullary schwannomas, intramedullary tumor, melanotic schwannomas, schwannomas imaging features

INTRODUCTION

Melanotic schwannoma is a nerve sheath tumor composed of melanin-producing cells with ultrastructural features of Schwann cells. The tumors occurred predominately in an intradural extramedullary location present at a younger age than conventional schwannomas. Intramedullary melanotic schwannomas are very rare. Only seven cases have been reported in literature.[3] This is the first reported patient to be of Asian ethnicity.

CASE REPORT

A 23-year-old female patient without any significant medical or genetic history had been suffering from progressive right lower limb numbness and soreness
with pain for 3 months. She acutely deteriorated developing progressive paraparesis with urinary incontinence days before being brought to our hospital. Her physical examination revealed sensory impairment with pathological reflexes below the T4 level. Muscle power of bilateral lower extremities was Grade 2. A magnetic resonance imaging (MRI) study was undertaken immediately after examination and revealed an intramedullary heterogeneous enhanced lesion at T4-5 level [Figure 1]. A small cystic section located at the upper portion of the mass lesion with hyperintensity on T1WI, hypointensity on T2WI, and a long segment of syringomyelia from T4 to T10 level was also noted. We thought of hemorrhagic ependymoma, astrocytoma with high grade component, hemangioblastoma, and schwannoma as differential diagnosis.

In view of her acute and progressive neurological decline and the MRI findings suggesting a focal well defined intramedullary tumor, we decided to operate on her emergently. We performed laminectomies from lower T3 to upper T6 level. The dura was opened in midline and kept by tenting suture. The tumor was intradural intramedullary and was visualized on the surface of the dorsal cord. The tumor appeared to be dark black in color and appeared to have a plane demarcating the tumor from adjacent normal cord parenchyma. En bloc tumor resection was performed after devascularization carefully. Different stages of hemorrhage were visualized and resected. Intraoperative histopathology revealed a preliminary report of a melanin-rich low-grade neoplasm with Ki-67 of 5%. A surgical gross total resection was confirmed by postoperative MRI scans [Figure 2]. The patient neurologically improved postoperatively, gradually beginning to walk after a week with assistance and subsequent near total neurological recovery in 3 months. Follow-up MRIs performed 3 years after surgery showed no recurrence and the patient remained stable being able to perform normal living activities with a residual mild paresthesia of lower extremities.

Grossly, the tumor was heavily pigmented and almost entirely grayish-black tissue. The surgical specimen showed demarcated borders and ovoid shaped with rubbery consistency. Hematoxylin and eosin staining revealed that the tumor had been composed of both bland-looking epithelioid and spindle tumor cells containing melanin pigment and focal grooved nucleus. The tumor appeared to infiltrate nerve fascicles, which was accompanied by a large sized blood vessel. Immunohistochemistry was positive for diffuse Human Melanoma Black-45 (HMB-45), melan-A, vimentin, and S-100 [Figure 3].

**DISCUSSION**

Schwannomas constitute 30% of primary spinal neoplasms and occur predominately in intradural extramedullary locations associated with ventral or dorsal nerve roots. Melanotic schwannomas are uncommon tumors. Since its first reported appearance in 1932, only about 100 cases have been reported in the related literature. The occurrence of these tumors is referred to as melanocytic or pigmented schwannomas in an intradural extramedullary location. Patients can have various presentations depending upon the tumor location. Melanotic schwannomas of the peripheral nervous system are rare tumors that present at a younger age than conventional schwannomas. They are grossly pigmented and composed of Schwann cells that contain multiple melanosomes. Ten percent of these tumors are malignant, suggesting that the presence of...
melanotic Schwann cells may predispose to malignant transformation in some patients. Different theories exist as to the histogenesis of these tumors. Culhaci et al.\(^1\) attributed the abnormal differentiation of neural crest cells into Schwann cells with melanogenetic properties as a potential cause. Because progenitor neural crest cells are precursors of Schwann cells and melanocytes, this error in differentiation seems plausible. Mandybur suggested a melanomatous transformation of neoplastic Schwann cells according to the electron microscopic findings of cells with melanosomes in an intraspinal tumor.\(^5\) He explained the histogenesis of melanotic nerve sheath tumors as originating from aberrant or ectopic melanotic cells. To date, the actual diagnosis of melanotic schwannoma remains difficult and melanotic schwannoma has been described as a part of the Carney complex.\(^7\) Distinguishing melanotic schwannoma from malignant melanoma is of paramount importance in planning the management, but these two tumors are most likely to share the same origin.\(^4\)

Melanotic schwannoma is less common in the spinal canal. Most of the spinal melanotic schwannomas are located in the thoracic region. These tumors can grow extradurally or intramedullary. Only seven cases of intramedullary melanotic schwannoma have been previously reported.\(^3\) Our case is the eighth report of intramedullary melanotic schwannoma and the only case presenting with tumor bleeding. Various causes to explain this hemorrhage include ectatic and hyalinized tumor vessels undergoing spontaneous thrombosis and distal tumor necrosis, or rapid tumor growth, or malignant transformation. However, there is no definite relevance. For some cases, paramagnetic effect due to free radicals in melanin can aid to differentiate melanotic lesion from nonmelanotic lesion. According to the MRI of this case, the T1 high signal intensity was not as obvious as the other seven cases previously presented. There was also evidence of hemorrhaging episodes and syrinx formation that was not reported in the prior cases. The typical melanin signal not being significant in this case may be due to a previous hemorrhage or the amount of melanin within tumor. The melanotic schwannoma picture of our

| Series                | Age (year) | Sex | Presentation                                      | Localization          | Duration of symptoms | Treatment                                      | Follow-up                                |
|-----------------------|------------|-----|--------------------------------------------------|-----------------------|----------------------|------------------------------------------------|------------------------------------------|
| Solomon et al., 1987  | 69         | M   | Right Brown-Sequard syndrome                     | Caudal medulla-C3     | 4 years              | Gross total resection                           | Sensory impairment deterioration after operation without long-term follow up data |
| Marchese et al., 1990 | 72         | F   | Quadriaparesis                                   | C4-C6                 | 20 years             | Gross total resection                           | Partial recovery                         |
| Sola-Perez et al., 1994 | 63        | F   | Right cervical dorsal radicular pain              | C7-T1                 | -                    | Fine needle aspiration                          | -                                        |
| Acciarri et al., 1999 | 44         | F   | Spastic quadriaparesis                           | T2-T3                 | 10 years             | Gross total resection                           | 4 months improved                        |
| Santaguida et al., 2006 | 35        | M   | Right hemiparesis and Progressive weakness and lower leg spasticity | C4-C6                 | 10 months            | Gross total resection; radiation therapy at recurrence; chemotherapy | Recurrence at 10-12 months, Radiation therapy, Re-operation at 4 years, Postoperative improvement |
| Mouchaty et al., 2008 | 56         | F   | Incomplete flaccid paraplegia                    | T12-L1                | >6 months            | Gross total resection                           | 12 months partial recovery                |
| Hoover et al., 2012   | 62         | F   | Progressive lower limbs weakness with urinary incontinence | T11                   | <6 months            | Gross total resection                           | 10 months, obvious recovery except sensory deficits |
| Pan et al., 2014 (present report) | 23 | F   | Incomplete flaccid paraplegia with urinary incontinence | T4-5                  | <6 months            | Gross total resection                           | 3 years, near total neurological recovery except sensory deficits |

Figure 3: Morphological appearance along with immunohistochemical profile of melanotic schwannoma. Photomicrograph showing tumor composed of both bland-looking epithelioid and spindle tumor cells containing melanin pigment and focal grooved nucleus, proliferation of astrocytes with eosinophilic body and Rosenthal fibers formation (a,b). Immunohistochemistry was shown to have diffuse immunocoloration for melan-A (c) and S-100 (d)
case is mimic hemorrhagic ependymoma or high grade glioma and more difficult for differential diagnosis based on the MRI alone.

A detailed review of the related literature, including intramedullary melanotic schwannoma presentation, duration of symptoms, location, treatment, and outcome, are illustrated in Table 1. All reported cases of intramedullary schwannoma involve the cervical and/or thoracic region, except for case 6, which suggests that the embryonic closure of the more rostral components of the neural tube and crest in these regions may be involved in the cause of these tumors. The tumor has been predominant in female patients.

Surgical excision is treatment of choice in most cases. In seven operative cases, six did not show recurrence, whereas one showed aggressiveness and contiguous invasion requiring radiotherapy, re-operation, and carboplatin chemotherapy. Our patient presented acute deterioration of neurologic deficit and recovered well after early surgical decompression of the spinal cord compromise. She showed a nearly complete recovery and has been recurrence free over the last 3 years of follow-up. Our report is the first Asian case and the youngest one, also the only case with tumor hemorrhage. It remains unknown at this time if there is any relative factor between race and tumor bleeding risk.

CONCLUSION

Intramedullary melanotic schwannomas are extremely rare. Intramedullary melanotic schwannomas presenting with hemorrhage tend to be mistaken with ependymoma, which by far is the most common spinal tumor to present with hemorrhage. The theory that the tumor may originate from the rostral components of the neural tube is still compatible with our patient. Total surgical resection is the best treatment. Further genetics study and follow up are expected.

REFERENCES

1. Culhaci N, Dikicioglu E, Meteoglu I, Boylu S. Multiple melanotic schwannoma. Ann Diagn Pathol 2003;7:254-8.
2. Font RL, Truong LD. Melanotic schwannoma of soft tissues. Electron-microscopic observations and review of literature. Am J Surg Pathol 1984;8:129-38.
3. Hoover JM, Bledsoe JM, Giannini C, Krauss WE. Intramedullary melanotic schwannoma. Rare Tumors 2012;4:e3.
4. Killeen RM, Davy CL, Bauserman SC. Melanocytic schwannoma. Cancer 1988;62:174-83.
5. Mandybur TI. Melanotic nerve sheath tumors. J Neurosurg 1974;41:187-92.
6. Murali R, Field AS, McKenzie PR, McLeod DJ, Stretch JR, Thompson JF, et al. Melanotic schwannoma mimicking metastatic pigmented melanoma: A pitfall in cytological diagnosis. Pathology 2010;42:287-9.
7. Vezzosi D, Vignaux O, Dupin N, Bertherat J. Carney complex: Clinical and genetic 2010 update. Ann Endocrinol (Paris) 2010;71:486-93.