

A Rare Case of Hemorrhagic Melanotic Schwannoma in a 38-year-old Female

Abstract

Melanotic schwannoma is a rare form of nerve sheath tumor composed of melanin-producing neoplastic Schwann cells. Less than 200 cases have been reported worldwide. The entity has been associated with Carney complex, a rare genetic disorder characterized by multiple benign tumors. A 38-year-old female presented to our unit with sudden-onset lower back pain and radiculopathy triggered by a mechanical injury. Imaging demonstrated a lesion within the left L5/S1 neural exit foramen with remodeling of bony architecture typical of a chronic, benign process. She proceeded for resection and histology revealed a psammomatous melanotic schwannoma. The patient recovered well with improvement in symptomatology. Due to the aggressive nature of the disease, she remains under surveillance for local recurrence and distant metastasis. Clinicians should be aware of this malignant entity, despite its possible presentation with radiological features of a chronic, benign process. Unusual characteristics such as hemorrhage should be treated with a high index of suspicion.

Keywords: Carney complex, melanotic schwannoma, nerve sheath tumor, psammoma bodies

Introduction

Melanotic schwannoma is a rare form of nerve sheath tumor with <200 cases reported worldwide. It accounts for <1% of all peripheral nerve sheath tumors and has a predilection for spinal nerves.

Melanotic schwannomas can be divided into psammomatous or nonpsammomatous, a discordance that is important due to the former's association with Carney complex. This is a rare genetic disorder characterized by multiple benign tumors, most often affecting the endocrine system, heart, and skin.

In the past, melanotic schwannoma was thought to be an entity with a predominantly benign course, but in more recent literature, the condition has been demonstrated to be more aggressive in nature.

It is imperative that clinicians are aware of this malignant entity, despite its possible presentation with radiological features of a chronic, benign process. Unusual characteristics should be treated with a high index of suspicion.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

Case Report

A 38-year-old female presented to our neurosurgical department with a sudden-onset dull ache in her lower back region followed by sharp radicular pain radiating down from her left gluteal region to the foot while attempting to move a dresser. The patient reported a progression in both frequency and intensity of radicular pain over the following weeks with sleep disruption by the time of presentation. The pain was associated with intermittent paresthesia and numbness in the sole of her left foot, but she denied any weakness or other concerning symptoms.

The patient's general practitioner commenced her on nortriptyline, but this provided little symptom relief despite fine dosage titration.

On examination, gait was normal. Tandem walking was intact. The patient was able to walk on tiptoes and the ball of her heels. There was no evident muscle wasting in her lower limbs. Romberg's test was negative. She was able to straight leg raise up to 90° bilaterally, albeit with slight apprehension on the movement of her left leg. There was

How to cite this article: Biju RD, Duignan M, Maharaj M, Ziad F, Hussain Z. A rare case of hemorrhagic melanotic schwannoma in a 38-year-old female. *Asian J Neurosurg* 2020;15:709-12.

Submission: 07-Apr-2020 **Revised:** 01-Aug-2020
Accepted: 02-May-2020 **Published:** 28-Aug-2020

**Rakesh Danny Biju,
Maria Duignan,
Monish Maharaj,
Fouzia Ziad¹,
Zakier Hussain**

*Departments of Neurosurgery
and ¹Pathology, Waikato
Hospital, Hamilton,
New Zealand*

Address for correspondence:

*Dr. Rakesh Danny Biju,
Department of Neurosurgery,
Waikato Hospital, Pembroke
Street, Hamilton 3204,
New Zealand.*

E-mail: r.danny.b@hotmail.com

Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_142_20

Quick Response Code:



normal power in all muscles of her lower limbs, and the sensation was intact. Reflexes were preserved, and plantars were flexor bilaterally.

The patient had a lumbosacral X-ray, which showed scalloping of the left L5/S1 neural exit foramen typical of a chronic, benign process. A magnetic resonance imaging (MRI) of the spine confirmed this and demonstrated the presence of a well-circumscribed lesion within the foramen with some extraforaminal extension [Figures 1 and 2]. Some high T2 signal changes were noted within the lesion, suggesting recent intralesional hemorrhage likely precipitating her symptom onset and progression.

Following a multidisciplinary discussion, the provisional diagnosis was a benign nerve sheath lesion arising from the left L5 nerve root, which gradually increased in size and became symptomatic post hemorrhage and rapid expansion. Surgical resection was recommended and carried out through a paramedian extraforaminal approach.

Intraoperatively, the lesion was noted to be encapsulated and hyperpigmented (grayish) with a hematoma present in its core. Internal debulking was performed, and the tumor was excised piecemeal. It appeared to be arising eccentrically from the nerve root, which was adequately decompressed by the end of the procedure.

Histopathology sections demonstrated tumor along with fragments of the adjacent dura and nerve root. There was a mixture of spindle and epithelioid cells arranged in fascicles, sheets, and nests. These cells contained abundant intracytoplasmic pigment and had prominent nucleoli [Figures 3 and 4]. A vaguely lobular and palisading pattern of arrangement was noted [Figure 4]. There were also psammoma bodies which coalesced in some foci forming areas of calcification [Figure 3]. Tumor was seen infiltrating dura with ganglion cells adjacent to it. Scattered mitotic activity was seen, including focal areas where up to five mitotic figures were identified in 10 high-power fields. No areas of necrosis were identified. While there were overlapping histologic features between a melanoma and schwannoma, the entity appeared to be biologically distinct from both. Immunohistochemistry confirmed the presence of both SOX10 and HMB45 positive tumor cells [Figures 5 and 6] and loss of PRKAR1A staining. The morphology and immunophenotype were consistent with a psammomatous melanotic schwannoma. Differentials included malignant melanoma and meningeal melanocytoma, both of which were ruled out due to the presence of psammoma bodies.^[1]

Postoperatively, the patient recovered well with improvement in both her back and radicular pain. A postoperative MRI of the spine showed satisfactory resection of the lesion, and a staging computed tomography showed no evidence of extraspinal lesions.

As part of investigations for Carney complex, she had blood tests which demonstrated no endocrinopathy.



Figure 1: Sagittal and axial T2-weighted magnetic resonance imaging of the lumbosacral spine demonstrating the lesion in the left L5/S1 neural foramen with some extraforaminal extension. It appears to be a well-circumscribed solid lesion with a central liquid core. Remodeling of the neural exit foramen is noted, suggesting a chronic, benign process



Figure 2: Coronal T2-weighted magnetic resonance imaging of the lumbosacral spine demonstrating the lesion

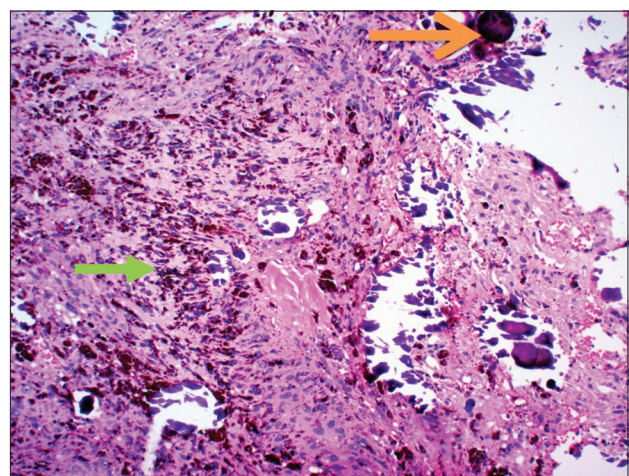


Figure 3: Psammoma body (orange arrow). Pigmented spindle cells (green arrow) demonstrating nuclear enlargement with vesicular chromatin and distinct nucleoli

The patient was discharged home 3 days later and remains under close surveillance for local and distant metastasis

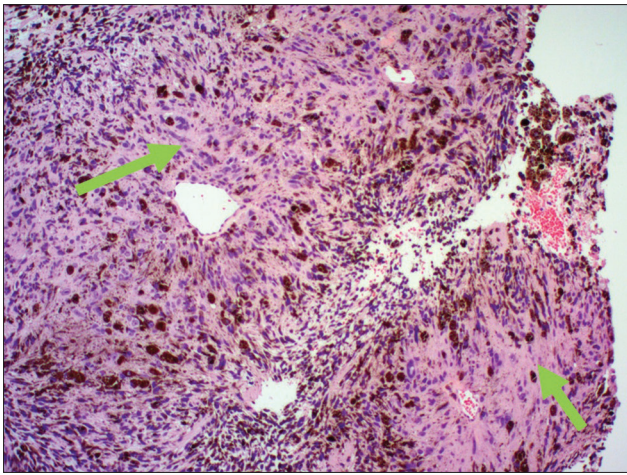


Figure 4: Spindle cells demonstrating palisading arrangement (green arrows). Both spindle and epithelioid cells show abundant intracytoplasmic pigment deposition

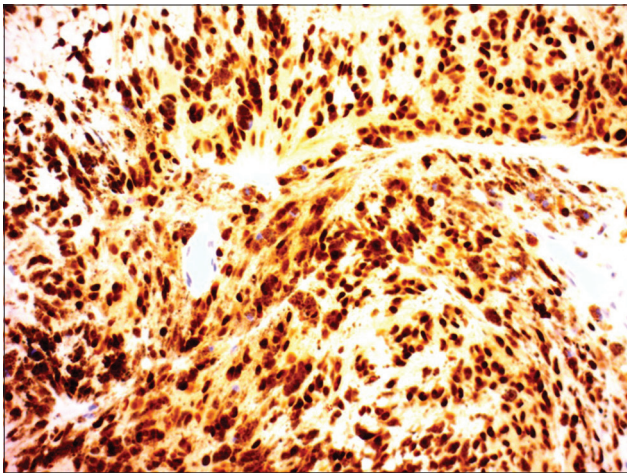


Figure 5: Immunohistochemistry confirming SOX10-positive tumor cells. SOX10 is a sensitive and specific marker of malignant melanoma

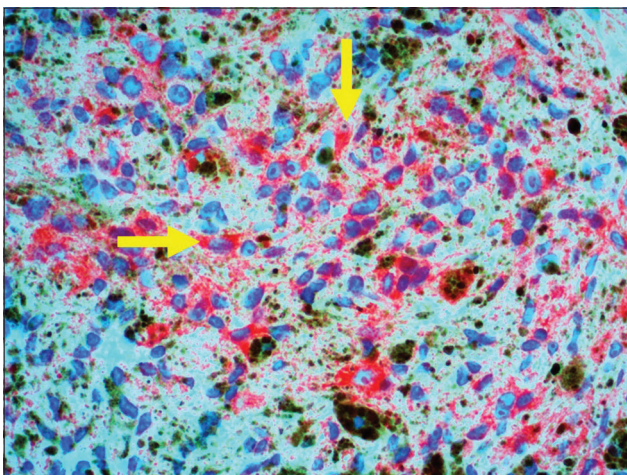


Figure 6: Immunohistochemistry confirming HMB45-positive tumor cells (yellow arrows). HMB 45 is a monoclonal antibody used as a common marker to confirm melanoma

with an echocardiogram and genetic testing for Carney complex pending at the time of this report.

Discussion

Melanotic schwannoma is a rare form of nerve sheath tumor. Less than 200 cases have been reported worldwide. It accounts for <1% of all peripheral nerve sheath tumors and has a predilection for spinal nerves.^[2] It has no sex predisposition with a mean age of presentation of the 38-year-old female.^[3]

On a cellular level, lesions are composed of cells possibly derived from the multipotent neural crest cells of origin that also differentiate to form melanocytes. It often exhibits hybrid microscopic and ultrastructural features of both melanocytes and Schwann cells. The classic morphological features of melanotic schwannomas were demonstrated in our patient's histology, including spindle and epithelioid cells with lightly eosinophilic, somewhat fibrillar-appearing cytoplasm growing in fascicles, sheets, and nests with variably abundant melanin pigmentation. In most cases reported in the literature, spindle cells dominated the composition of tumor.^[4] Melanotic schwannomas can be divided into psammomatous or nonpsammomatous, a discordance that is important due to the former's association with Carney complex. Psammoma bodies are round collections of calcium deposits appreciated under the microscope.^[5] In one of the largest case series to date, Torres-Mora *et al.* noted the occasional hemorrhagic core in some melanotic schwannomas, but only one out of the 40 cases reported an encapsulated tumor like ours.^[4]

Fifty percent of patients with psammomatous melanotic schwannoma have a Carney complex. This is a rare genetic disorder characterized by multiple benign tumors, most often affecting the endocrine system, heart, and skin. Abnormalities in skin pigmentation result in a spotty appearance of affected areas, which is most commonly caused (70%) by a mutation in the *PRKAR1A* gene within chromosome 17 (Locus 17q23-24).^[6]

A patient's presenting complaint is often dependent on the location of the lesion. Our patient developed radicular pain as a result of the L5 nerve root involvement. She had remained asymptomatic till a mechanical injury provoked a small volume hemorrhage likely, causing a rapid expansion in the size of the lesion and contributing to the symptoms that lead to its discovery. Posterior spinal nerve roots are one of the more common sites for these lesions, along with cranial nerve roots and the sympathetic chain. Less common primary sites include the peripheral nerves and the gastrointestinal tract.^[7,8]

MRI remains the diagnostic imaging of choice in neurological and spinal disease. To date, there have been no large reviews of imaging characteristics for melanotic schwannomas, but it is widely acknowledged to exhibit signal hyperintensity on T1-weighted sequences and hypointensity/isointensity on T2-weighted sequences due to the presence of melanin within.^[8] These characteristics

were demonstrated in our patient's imaging along with a T2 signal hyperintensity representing a central liquid core, which was the hematoma noted intra-operatively.

Previously, melanotic schwannoma was thought to be an entity with a predominantly benign course. The metastatic potential was only demonstrated in up to 26% of cases.^[9] In more recent literature, the condition has been demonstrated to be more aggressive in nature, with local recurrence noted in 35% and evidence of distant metastases in 42% of cases.^[4] Of note, only three cases within the review documented a positive Carney complex status, and of these, only one (33%) had metastatic disease.

Given these figures, aggressive treatment is recommended. The aim of surgical intervention should be to achieve gross total resection. Postoperatively, the patient should be kept under close surveillance with interval imaging looking for potential recurrence of the disease. There are currently no formal recommendations with regard to the duration of surveillance.

Conclusion

Clinicians should be aware of this malignant entity, despite its possible presentation with radiological features of a chronic, benign process. Unusual characteristics such as hemorrhage should be treated with a high index of suspicion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal

their identity, but anonymity cannot bechrological order guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Louis D. WHO Classification of Tumours of the Central Nervous System. 4th ed. France: LARC; 2006. p. 173.
2. Khoo M, Pressney I, Hargunani R, Tirabosco R. Melanotic schwannoma: An 11-year case series. *Skeletal Radiol* 2016;45:29-34.
3. Gulati HK, Joshi AR, Anand M, Deshmukh SD. Non psammomatous melanocytic schwannoma presenting as a subcutaneous nodule: A rare presentation of a rare lesion. *Asian J Neurosurg* 2016;11:317-8.
4. Torres-Mora J, Dry S, Li X, Binder S, Amin M, Folpe AL. Malignant melanotic schwannian tumor: A clinicopathologic, immunohistochemical, and gene expression profiling study of 40 cases, with a proposal for the reclassification of "melanotic schwannoma". *Am J Surg Pathol* 2014;38:94-105.
5. Bernd WS, James MW, Robert AE. Atlas of Tumor Pathology: Tumors of the Peripheral Nervous System. 3rd ed. Washington DC: American Registry of Pathology; 1999.
6. Carney Complex. Available from: <https://rarediseases.org/rare-diseases/carney-complex/>. [Last accessed on 2019 Jul 22].
7. Carney JA. Psammomatous melanotic schwannoma. A distinctive, heritable tumor with special associations, including cardiac myxoma and the Cushing syndrome. *Am J Surg Pathol* 1990;14:206-22.
8. Liessi G, Barbazza R, Sartori F, Sabbadin P, Scapinello A. CT and MR imaging of melanocytic schwannomas; report of three cases. *Eur J Radiol* 1990;11:138-42.
9. Vallat-Decouvelaere AV, Wassef M, Lot G, Catala M, Moussalam M, Caruel N, *et al.* Spinal melanotic schwannoma: A tumour with poor prognosis. *Histopathology* 1999;35:558-66.