Journal of Cancer Diagnosis

Case Report Open Access

Melanotic Schwannoma: A Case Report and Literature Review

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Abstract

Melanotic Schwannoma (MS) is a variant of Schwannoma considered to be rare, characterized by a structure of variably melanin-producing Schwann cells, usually arising from posterior spinal nerve roots. Even though it's a relatively benign tumor, malignancy and metastases have been reported. They occur as isolated tumors or take part in the Carney Complex (CC) syndrome. Imaging by the MRI is essential, finding a lesion that is hyperintense in T1 and hypointense in T2. The therapeutic care is mainly surgical by total resection and the adjuvant therapy has yet to prove its efficiency.

Here we report our case of Melanotic Schwannoma located next to L5-S1, and how we did manage it, after a review of the literature.

Keywords: Spine; Melanotic Schwannoma; Posterior spinal nerve; Malignancy

Introduction

Melanotic Schwannoma (MS) was initially described in 1932 by Millar and proposed as a form of schwannoma in 1961 by Hodson [1]. Afterward, they termed the pathology Melanotic Schwannoma in 1975 [2, 3]. Around 200 cases have been reported in the literature [4]. This tumor is rare and uncommon but seen worldwide, representing less than 1% of all nerve sheath tumors [5]. The MS is usually a benign neoplasm but malignancy and aggressive behavior can be seen, involving in most cases the posterior nerve roots, about 30.5%. [4]. Still it has been revealed that the gastrointestinal tract, cranial nerve roots, sympathetic chain, peripheral nerves, and the spinal cord can be affected [6, 7]. The clinical phenotype of this pathology is defined to be caused by a Mendelian dominant hereditary pattern [8]. This type of schwannoma is described to have a structure of variably melanin-producing Schwann cells and metastatic potential [9, 10], It is hypothesized that the Schwann cells have the potential to synthesize melanin because both the Schwann cells and melanocytes originate from neural crest cells [11]. Which gives a tumor that seems black to brown or dark blue macroscopically [12, 13] and highlights under a microscopic view a heavy melanin deposition, spindle morphology, nuclear pleomorphism, and low mitotic rate with or without psammoma bodies [14, 15].

The lesion location involving spinal or other nerves, dictates the different symptoms related to the site and imaging is helpful in the management and the diagnosis of Melanotic schwannoma [16, 17], the modality of choice is magnetic resonance imaging and it is described as hyperintense in T1 and hypointense in T2 as originally reported by Bendszus [18].

The therapeutic care for this disease is challenging, and the evolution may be difficult to predict. We review our case and the status of MS in the literature, discussing the different components related to this pathology.

Report Case

A 21-year-old female without specific medical history expects a 3-year-old back pain with sciatica whose symptomatic treatment was not effective, even worsening her symptoms after an episode of childbirth followed by the occurrence of bilateral L5 lumbar sciatica and the appearance of genito-sphincteric disorder such as urinary and anal incontinence.

Additionally, the clinical examination found bilateral L5 and S1 low back sciatica with a partial left distal motor deficit of L5 and S1 as well as hypoaesthesia of the same path along with peri-anal hypoesthesia and hypotonia. This fits in the cauda equine syndrome.

The exploration by MRI showed a lesion next to L5-S1 exiting through the left foramen, displaying a slightly hyper intense signal in T1 taking the contrast in a hemogenic way, hypo to iso-intense signal in T2. Meanwhile computed tomography exploration discovered a bone erosion of S1 (Figures 1, 2 and 3).

The therapeutic attitude was a surgical decompression by an L5-S1 laminectomy and left-facing for aminotomy with an arthrodesis.

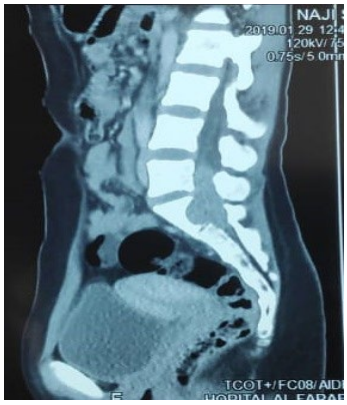

Figure 1: Sagittal, computed tomography image showing a bone erosion and deformation of S1 with a mass at the level L5-S1.

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Received: 05-Oct-2022, Manuscript No: JCD-22-73478, **Editor assigned:** 07-Oct-2022, PreQC No: JCD-22-73478(PQ), **Reviewed:** 21-Oct-2022, QC No: JCD-22-73478, **Revised:** 26-Oct-2022, Manuscript No: JCD-22-73478(R), **Published:** 02-Nov-2022, DOI: 10.4172/2476-2253.1000161

Citation: Elfarissi MA, Dahamou M, Dehneh Y, Mohamed K, Haloui A, et al. (2022) Melanotic Schwannoma: A Case Report and Literature Review. J Cancer Diagn 6: 161.

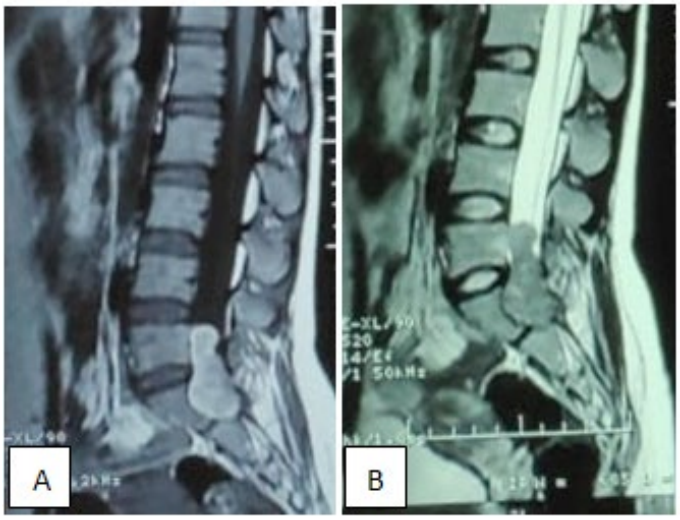

Figure 2: Sagittal, MRI image of the lumbar-sacral spine. Showing a mass at the L5–S1 level occupying the spinal canal hyperintense on T1 and hypointense on T2.

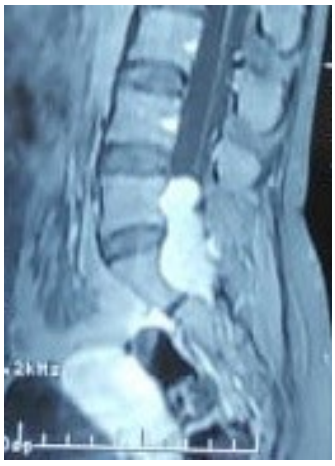

Figure 3: Sagittal, T1 post-contrast MRI image displaying enhancement of the mass.

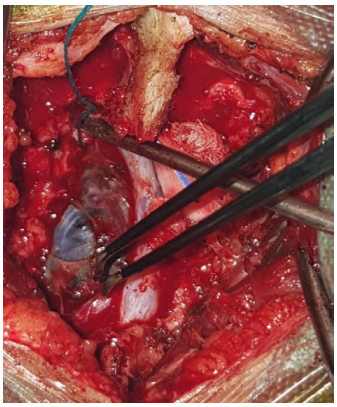

Figure 4: Photograph showing the tumor located at L5-S1. the lesion appeared to be connected to the nerve, covered by a thin fibrous membrane.

During the operation, we discovered a lesion that seems encapsulated in which the content was dark brown. Total resection was then made (Figures 4, 5 and 6).

The postoperative evolution was very favourable by the total recovery of the motor and sensory deficit, as well as the genito-sphincter disorders and the disappearance of the lumbosciatic pain. The result of the pathology study concerning the operative biopsy came back as melanocytic Schwannoma (Figures 7, 8 and 9).

After the result of the pathology study and the therapeutic attitude taken by the total resection of the lesion and a thorough review of the literature, the decision was to make the patient undergo a radiological assessment showing no other secondary lesion, which led us to simple monitoring by imaging and the result was satisfying by the non-recurrence of the lesion after a two years follow-up (Figure 10).

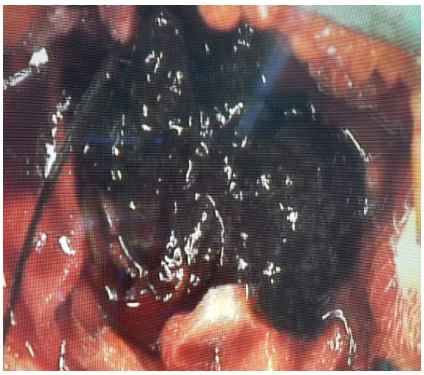

Figure 5: The tumor after opening the membrane shows a lesion that seems black to brown.

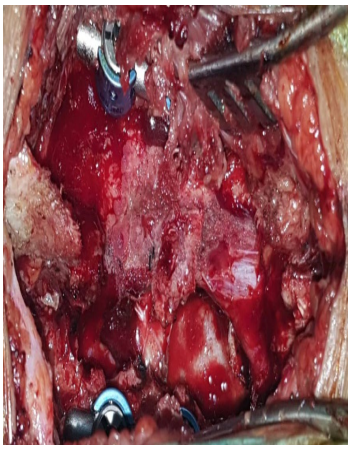

Figure 6: The surgical site after total resection of the lesion from the lumbar-sacral canal, before closure.

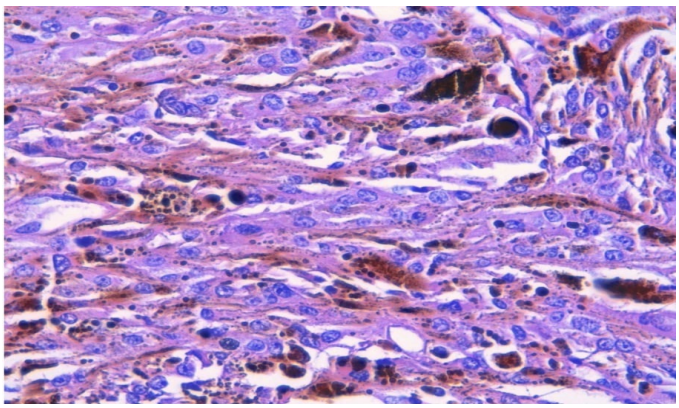

Figure 7: A tumor proliferation arranged in swirling and intertwined bundles, made up mostly of spindle-shaped cells, with fewer oval ones, provided with a nucleus with fine chromatin and an abundant eosinophilic cytoplasm containing important deposits of melanin pigments.

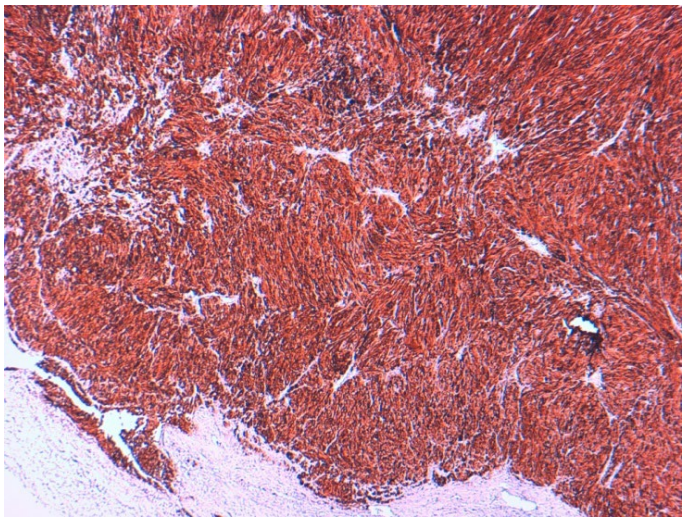

Figure 8: Low power view showing intersecting fascicles of spindle cells displaying a diffuse expression of Melan-A.

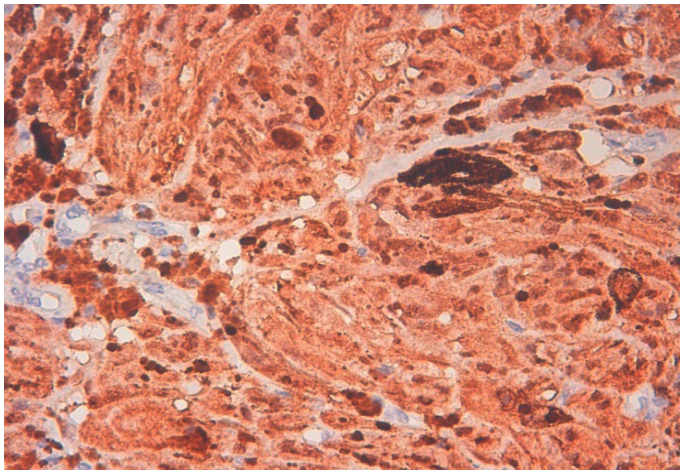

Figure 9: High power view of tumor cells showing cytoplasmic expression of PS100.

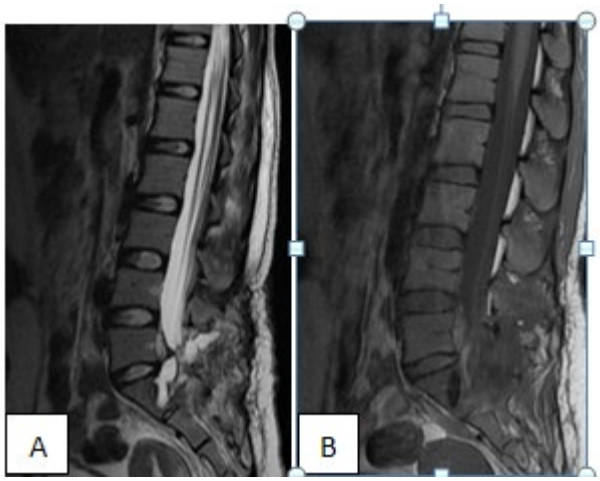

Figure 10: MRI imaging showing the total resection of the lesion.

Discussion

The MS is described as melanocytic or pigmented schwannoma, evolving in intradural extra medullary location [19]. Emerging mostly in posterior spinal nerve root [5, 20] although other locations have been reported including oral cavity, oesophagus and stomach wall, bronchus, uterine cervix, retro peritoneum, parotid, the sympathetic chain, acoustic nerve, cerebellum, orbit, choroid, soft tissues, and the heart [7, 21]. This anomaly is mostly solitary, however, multifocal tumors can be found in some cases. Macroscopically this type of schwannoma is usually solid, but cysts can be found on the surface of the tumor. The indicated lesion takes a round or ovoid shape, appearing to be connected to the nerve, well-circumscribed, and covered only by a thin fibrous membrane. The diameter of the MS is frequently around 5 cm or even larger and the consistency when cutting the surface is tar-like with the color varying from gray to pitch black [9, 13, 14, 22 and 23]. Microscopically, the MS is circumscribed but not encapsulated, arranged in interlacing fascicles or nests of plump spindle and epitheliod cells and it appears to be accumulating melanin in neoplastic cells with associated melanophages, demonstrating a great variation in the cytoplasmic pigmentation containing nuclei that can be round, ovoid or elongated with delicate and evenly distributed chromatin, containing small distinct nucleoli alongside some areas having large and prominent one, With rarely seen mitosis [7, 9, 13, 16, 24]. This pathology can be divided into two types, psammomatous and non-psammomatous. As matter of fact, about 10% of the MS do not express psammoma bodies, are regarded as sporadic type, and may demonstrate a malignant advance [25]. On the other hand, the psammomatous one can be related to the Carney complex (CC) and is characterized by the existence of myxomas (the heart, breast, uterus, skin lentigines), endocrinopathy (Cushing's syndrome, pituitary adenoma producing growth hormone), epithelioid blue nevi, Sertoli cell tumors of the testis, tumors of the thyroid and ductal adenomas of the breast [26-28]. When CC is associated, sheets of adipose-like cells can be found [9, 13]. And if the diagnosis of CC is set, the following annual exams are recommended: echocardiogram, measurement of urinary, free-cortisol levels, and testing of serum insulin-like growth factor-1 levels. A testicular and thyroid ultrasonography is in the same way needed at the initial evaluation [16]. So early observation and management of cardiovascular disorders are fundamental being the most common cause of morbidity and mortality [16].

Meanwhile, the MS in the immune-histochemical staining for S100, SOX10, HMB-45, Melan-A, p16, and vimentin, demonstrate positive results [9, 13] as well as detecting both laminin and collagen IV in all cases on the linear and peri-cellular immunoreactions study [29].

The MS is described on the molecular level by a complex karyotype with recurrent monosomy of band 22q, and fluctuating full chromosomal gains and recurrent losses generally involving chromosome 1 and 21 and chromosome arm 17q [28] and have the possibility to arise from two genetic disorders that have mutations on chromosome 17 [7]. The first one is the CC as an autosomal-dominant genetic disease by the mutation within chromosome 17 affecting the protein kinase cAMP-dependent type l regulatory subunit alpha gen [PRKAR1A] [9, 24, 30]. The second one is the neurofibromatosis type l which is characterized by neuro fibromas and cafe au lait spots, having the mutation on the 17q11 band [31].

Clinically the MS concerns adults with an average age of 38 years old; with no sex predilection [29], depends on the location and the involvement of spinal or other nerves, and is related to the impairment of the somatic and autonomic parts as well as sympathetic ganglia [9]. As the tumor is growing, the spinal nerves may be compressed leading to both motor and sensory symptoms, such as tingling sensations, numbness, weakness, and pain in the lower limbs. Adjacent structures

may suffer from the mass effect and reveal mechanical dysfunction as described in 13% of cases [16]. The MS has other manifestations on the skin, described as resembling melanomas lesions, while subcutaneous involvement is revealed as a slowly growing soft tissue mass [32], in affected bones, the patient can be complaining of a pain bony mass [16, 33]. However, 29% of cases are asymptomatic.

The radiological assessment of MS by standard Radiography and computed tomography shows a widening of the intervertebral foramina, sclerosis, and even bone erosion [16, 17]. The myelograms demonstrate obstruction of the contrast flow without displacement of the spinal cord [16].

But the investigation of choice is the MRI, which it is assuming a dumbbell appearance [28], described as hyperintense on T1-weighted and hypointense on T2-weighted sequences due to paramagnetic free radicals in melanin [27, 34, 35].

The MS has some differential diagnoses including conventional schwannoma, pigmented lesions such as pigmented neuro-fibroma, meningeal melanocytoma, metastatic melanoma, and clear cell sarcoma [36].

The therapeutic care as suggested in the literature is mainly surgical by the complete resection of the tumor whenever possible, knowing that it's difficult and even impossible in some cases, because of the local infiltration. This leads to adjuvant postoperative therapy considered to reduce the high risk of local recurrences, malignant transformation, metastases, and subarachnoid space seeding [5, 27, 28, 37]. Because the prognosis can be poor if other systemic manifestations occur [16].

It was reported that the chance of recurrence following resection is 18.2% and metastasis is 9.1% [7], making radiotherapy recommended following subtotal resection [38]. The employment of fractional radiation therapy for difficult MS tumors, counting those near vital structures like the spinal cord, has yet to show some definite life expectancy benefit [16]. Even chemotherapy has displayed a low feedback rate with no life span benefit [39]. The complications can always emerge and it depends on where the tumors are located or even the complications related to CC [40]. The behavior of MS is challenging to predict, and the patients need a longer follow-up period due to its malignant potential [38]. Adequate guidelines for adjuvant therapies have yet to be officially implemented.

Conclusion

MS is a variant of schwannoma, considered to be rare, composed of Schwann cells and melanin pigment. It's usually a benign tumor, even though malignity and metastasis have been reported. The diagnosis of the lesion can be suspected by the MRI and the confirmation is histological. More examination should be done to find other systemic manifestations, especially when discovering a CC syndrome. The therapeutic care is mainly surgical by total excision when possible, knowing that the benefit of adjuvant treatment has yet to be proven. A close follow-up of the patients is highly needed due to the malignant potential of the tumor. Further studies are required to clarify guidelines concerning all the aspects of this pathology.

Conflict of Interest

The authors have no conflicts of interest to declare.

This article is done with the consent of the patient.

All persons who meet authorship criteria are listed as authors, and all authors certify that they have participated sufficiently in the work to take public responsibility for the content, including participation in the concept, design, analysis, writing, or revision of the manuscript. Furthermore, each author certifies that this material or similar material has not been and will not be submitted to or published in any other publication before. And there are no conflicts of interest and ethical Adherence, and financial disclosure.

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