



L3 Rootlet Recurrent Melanocytic Schwannoma – Case Report and Literature Review

Georgi K. Georgiev¹, Ivan V. Todorov¹, Todor P. Shamov¹, Ivan V. Krushovenski¹, Albena D. Fakirova², Boryana N. Ilcheva², Iliyan T. Georgiev¹, Nikolai D. Georgiev¹, Vladimir S. Prandjev¹, Tihomir N. Eftimov¹

¹ Clinic of Neurosurgery, Military Medical Academy, Sofia, Bulgaria

² Department of Pathology, Military Medical Academy, Sofia, Bulgaria

Corresponding author: Georgi K. Georgiev, Military Medical Academy, 3 St Georgi Sofiyski St., 1606 Sofia Center, Bulgaria; E-mail: georgi_krasimirov91@abv.bg; Tel.: +359 879 171 561

Received: 23 June 2020 ♦ **Accepted:** 30 July 2020 ♦ **Published:** 30 June 2021

Citation: Georgiev GK, Todorov IV, Shamov TP, Krushovenski IV, Fakirova AD, Ilcheva BN, Georgiev IT, Georgiev ND, Prandjev VS, Eftimov TN. L3 rootlet recurrent melanocytic schwannoma – case report and literature review. *Folia Med (Plovdiv)* 2021;63(3):448-56. doi: 10.3897/folmed.63.e55295.

Abstract

First described by Miller in 1932, melanocytic schwannoma (MS) (melanotic schwannoma, pigmented schwannoma) is a rare variation of peripheral nerve sheath tumours with ectodermal origin occurring predominantly in somatic, but also in the autonomic peripheral system with around two hundred cases in the literature. Predominantly benign tumours, MS are still imaging and pathological challenge and can be easily misdiagnosed with more aggressive peripheral nerve tumours.

We report a case of melanocytic schwannoma on L3 sensory rootlet with systematic literature review of nearly 200 cases presented in intracranial, paraspinal region, thoracic, abdominal or pelvic cavities and skin. Two-thirds of cases are part of Carney complex.

We present a case of a 61-year-old male with a 3-month history of low back pain, progressive numbness and stiffness in the right thigh, shin and knee, tibial and peroneal paresis causing gait disturbance and neurological claudication. MRI findings present “sand clock” type intradural extramedullary tumour formation with extension to the L3 rootlet through right L3-L4 foramen, hypointense on T2 and hyperintense on T1. Pathological diagnosis of sporadic type melanocytic schwannoma was made via immunohistological and ultrastructural analysis. Thirteen months after total resection there was clinical and MRI evidence of recurrence of the tumour. Total resection and radiosurgery was performed with a recurrence free period of 14 months.

A gold standard for melanocytic schwannoma treatment is gross total surgical resection. Despite being considered benign tumours, MS have a local or metastatic recurrence of around 13%. MRI imaging in most of the cases is insufficient and only exhaustive pathological and immunohistological examination is the key to diagnosis. Need of postoperative radiation therapy is still controversial. For the first time, a criterion for postoperative adjuvant therapy was established.

Keywords

Carney complex, melanotic, pigmented, recurrent

INTRODUCTION

Melanocytic schwannoma (MS) (melanotic schwannoma, pigmented schwannoma) was first described by Miller in

1932. It is a rare variation of the peripheral nerve sheath tumours¹ with ectodermal origin occurring predominantly in the somatic, as well as in the autonomic peripheral nervous system. There are about two hundred cases known in the literature and most of them have extracranial loca-

tion – mainly in the paravertebral region, but also in the internal organs, skin, and only dozen cases of intracranial location. Despite the fact that it is considered a benign tumour, around one-seventh of the cases have malignant transformation and 9%-42% demonstrates local or distant recurrence and metastases.²⁻⁶

According to the WHO 2016 classification, there are two subtypes of MS – sporadic and psammomatous.⁷ The first histological subtype is part of the Carney complex (autosomal dominant genetic disorder) with age predominance in the fourth decade of life, which is considered more aggressive.^{1,3} Psammomatous or sporadic subtype is typical for the fifth decade of life and has more benign course and lower recurrence rate.¹

CASE REPORT

Medical history and examination

A 61-year-old male presented with complaints of low back pain, progressive numbness and stiffness in the right thigh, shin and knee over the past 3 months, which had been causing gait disturbance and neurogenic claudication. He also reported that his legs felt cold, but no peripheral vascular pathology was observed. He was treated with oral/intra-muscular administration of COX-2 inhibitors, corticosteroids and galantamine without any significant effect. No clinical features of the Carney complex were present. The patient had a medical history of hypertension, well-

controlled asthma, myocardial infarction (1991) and transient ischemic attack in the part of the brain supplied by MCA (2016) without neurological deficit. There was no family predisposition.

Physical examination revealed bilateral thigh hypotrophy that was more prominent on the right side with a considerable difference in the circumference of both thighs of 19 mm (measured in the middle between anterior inferior iliac spine and apex of patella in supine/recumbent position). The difference in the crural circumference between the right and left calf was 14 mm (measured on the border between upper and middle third from the most prominent area of the medial condyle and medial malleolus). Neurological examination showed vertebral syndrome, right-sided L3-L4 lumbo-sacral radiculopathy with hyperesthesia and paresthesia, peroneal nerve paresis -3/5 MRC (right) and 4+/5 MRC (left). Diminished ankle and patellar reflexes bilaterally were observed. No bladder dysfunction. No clinical findings of the Carney complex syndrome (Fig. 1).

The MRI findings show a “sand-clock” type intradural, extramedullary tumour formation with extension to the L3 rootlet through the right L3-L4 foramen, with bone erosion but no bony edema present. The tumour is presented as hypointense on T2 and hyperintense on T1 with dimensions 17×18 mm (intraforaminal part) and 33×28 mm (intradural part) with spinal canal stenosis (Fig. 2).

First surgical treatment

L3 laminectomy, partial L4 laminectomy and medial longitudinal durotomy were performed. An unusual black coloured



Figure 1. CT findings one month after beginning of the complaint. Red arrows are pointing at the erosion of the neural foramen and the expanding isodense lesion.

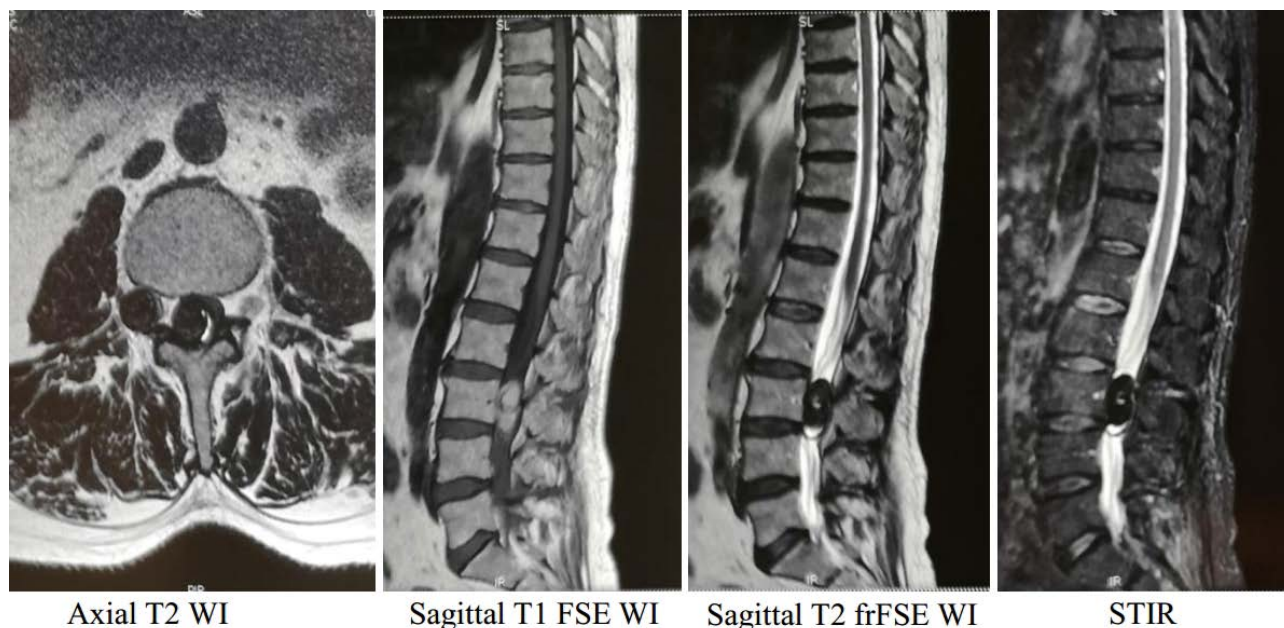


Figure 2. Pre-operative MRI findings visualizing the intradural and intra-/extraforaminal extension of the tumour.

tumour was found, well vascularized with aggressive invasion of the surrounding tissue and encapsulating the rootlets of the spinal cord, destroying the posterior L3 rootlet. The initial incision of the capsule led to leakage of stromal content with the characteristics of “olive paste”. L3 medial facetectomy was necessary because of the lateral extension of the tumour into the adjacent neural foramen (**Fig. 3**).

Intraoperative neuromonitoring and ultrasonic surgical aspiration were used. The obvious colour differentiation between the lesion and the rootlet was helpful for the perfect preservation of cauda equina. Only the L3 posterior rootlet was sacrificed because of its total destruction by the tumour. Total resection of the intradural, intra- and extraforaminal parts of the tumour was performed with no additional posterior vertebral osteosynthesis. The dura and the soft tissues were closed in a normal fashion.

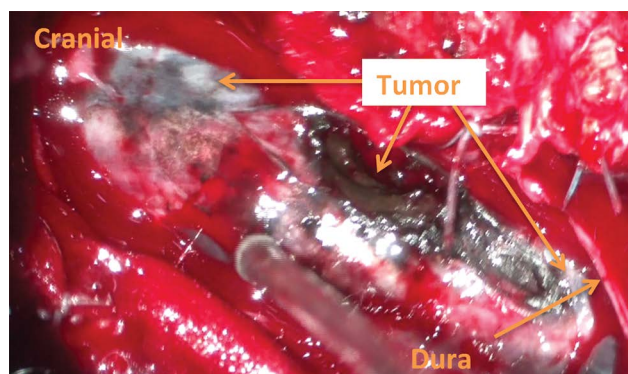


Figure 3. Intraoperative imaging of the melanocytic schwannoma. The tumour encapsulates cauda equina rootlets without clear arachnoid border. Profuse tumour bleeding was observed.

Postoperative evaluation

The patient had an uneventful postoperative period and was discharged from the hospital 6 days after surgery. No additional neurological deficit was present except mild hyperesthesia along the L3 dermatome.

Due to the “benign” histopathologic/immunohistology description of the sporadic melanocytic schwannoma and the totally resected tumour, no adjuvant therapy was considered necessary.

One course of post-surgical rehabilitation was performed. On the follow-up examination there was a complete relief of the stiffness and the numbness in both legs, especially on the right side, improvement of the L3-L4 radiculopathy with no need of painkiller administration. There was no significant difference in the circumferences of the crural musculature on the right and left lower extremities, while the peroneal nerve paresis remained on the right limb – 4/5 MRC. The patient missed his follow-up visit at 6 months and no MRI was performed.

Twelve months post-surgery, the patient developed new onset of pain with progressive exacerbation in the lower part of the thigh and his right knee, mild weakness in the quadriceps femoris muscle combined with stiffness and fatigue.

Postoperative MRI at 13 months revealed an extradural, intra-foraminal recurrent tumour with dimensions 11×25×15 mm, hypointense on T1 FSE and hyperintense on T2 frFSE was visualized (**Fig. 4**).

Second surgical treatment

A lateral facetectomy on L3-L4 was performed. Using the ultrasonic surgical aspirator and the optic magnification,

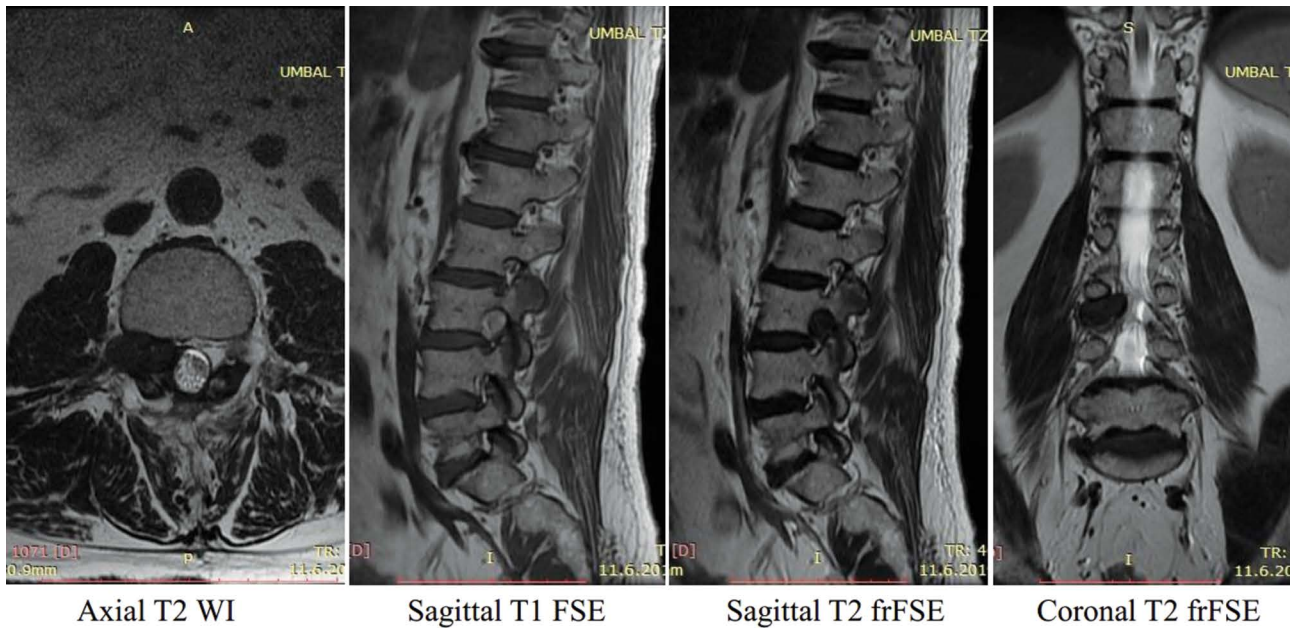


Figure 4. Postoperative MRI, 13 months after the first surgery with radiological data showing local recurrence of the tumour, encapsulating the L3 rootlet.

total resection of the tumour was achieved with sparing the motor root of L3 by using neuromonitoring. The macroscopic characteristics of the lesion were similar as described during the first surgery. No additional dural infiltration was noticed, except rootlet cuff was corroborated. The soft tissues were closed in a normal fashion.

Postoperative evaluation

The patient was discharged from the hospital 7 days postoperatively without any additional deficit, with significant pain relief, improvement of the stiffness in the right leg and the movement impairment. Three sessions of radiotherapy were given to the patient with a dose of 5Gy. An MRI study done 3 months after the surgery is shown below in Fig. 5.

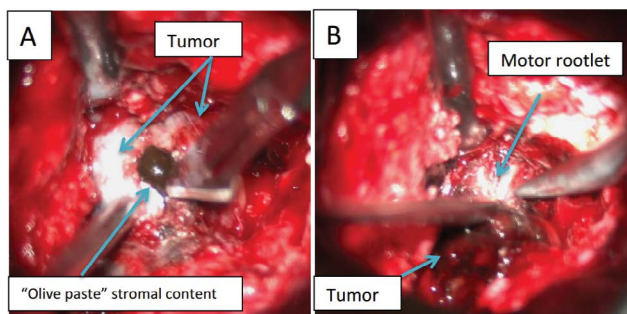


Figure 5. Intraoperative imaging in the second surgery. The lateral L4 facet joint is removed and the ganglion exposed. A) While cutting through the fibrous capsule, an interstitial liquid content “olive paste” like was pouring out; B) Careful dissection and en-block total resection of the tumour with preservation of the L3 motor rootlet.

DISCUSSION

Microstructural, types of MS, Carney complex ways of histological verification

It was originally described by Millar in 1932 as a malignant melanotic tumour of sympathetic ganglion cells.⁸ In 1961, Hodson suggested that it was a form of schwannoma.⁹ Because of its tendency to involve somatic and autonomic nerves as well as sympathetic ganglia, uniform composition of schwann cells and frequent association with neurological symptoms, the lesion is classifiable as peripheral nerve sheath tumour.⁹⁻¹¹

MS cells have microstructural, histological and immunohistochemical characteristics of both Schwann and melanocytic cells. The main theory for the etiology of MS cell is migration disorder of neural crest cells during closure of neural tube and based on the common cell lineage origin of both Schwann and melanocytic cells.¹² Other theories are based on neoplastic proliferation of Schwann cells in the dural cuff or dedifferentiation of neuroectodermal cells of pia in cases of intramedullary or cranial schwannomas.¹³

According to 2016 WHO CNS tumour classification, MS is described as a distinct type of tumour, presumed to derive from elements of neural crest lineage.¹⁴ MS is composed of cells with ultrastructural characteristic and immunophenotype of Schwann cells, but containing melanosomes in varying stages of maturation.⁸

There are two subtypes with different microscopic and ultrastructural characteristics: the psammomatous and the sporadic subtype. Psammomatous melanotic schwannoma (PMS) is named for its content of laminated calcospherule

“psammoma bodies”.¹² Around 55% to 60% of people with PMS have a Carney complex (CC), despite some reported PMS cases without CC features.

First described in 1985 and lately named after its discoverer, Carney complex is an autosomal dominant disease caused in approximately 70% of cases by inactivating germline mutations of the gene encoding the type 1A regulatory subunit of cyclic adenosine monophosphate (AMP)-dependent protein kinase A (PRKAR1A).¹⁵ Male-to-female ratio is 1:1.4¹⁶ but there is no information about race predominance. The clinical presentation of the CC are myxomas of the heart (65%), skin and breast³ spotty pigmentation of the skin on lips, inner and outer corners of the eyes, the conjunctiva (membrane lining) of the eye, and around the genital area 65% are typical¹⁷. Endocrine hyperactivity including ACTH-dependent Cushing’s syndrome caused by primary pigmented nodular adrenocortical disease, sexual precocity, and congenital osteo-chondromyoxoma³ are also presented.

Most of the symptoms of CC are presented between the end of the first and third decades of life.

Presentation

The median age of MS presentation is the third or fourth decade for PMS and the fourth or fifth decade for non-syndromic subtype.¹²

In more than 60% of cases, the tumours arise in the paravertebral area from somatic or autonomous spinal nerve roots.¹² The sporadic type is more often affiliated to somatic spinal nerve roots and ganglia in the paravertebral area with intradural extension causing pain and radiculopathy. The psammomatous subtype is presented frequently in association with autonomous plexi and ganglia in the para-

vertebral area but also in gastrointestinal tract and, lung, skin and pelvic organs, causing obstruction of their normal passage as a first clinical sign. Melanotic schwannomas can also arise from cranial nerves and can present in the soft tissues of the trunk and extremities, chest wall and palate. An intramedullary localization is exceptional but also has been encountered.¹³ Patients with the Carney complex may develop multifocal melanotic schwannomas.³

In the sporadic type, MS has clinical presentation that pertains to the position and relation to the neural structures or internal organs. The most frequent symptom in MS in the vertebral region is radiculopathy in nearly 92% combined with back pain 85% of cases whereas mass lesion with or without cranial nerve deficit is the most frequent appearance in intracranial MS. Great variety of symptoms can be caused in thoracic and abdominal cavities but none of them are pathognomonic. A major sign is mass lesion and obstruction of the internal organs outflow.

Microscopic appearance

To the unaided eye, melanotic schwannomas are generally circumscribed, may appear encapsulated, and range from gray or brown to jet black in colour.

Melanocytic schwannomas are rounded or ovoid in shape, most often covered by a thin, fibrous capsule. Sometimes stroma includes liquefied debris resembling “olive paste” (Fig. 6). Most tumours are up to 5 cm in diameter. Small thin-walled cysts on the surface of the tumour sometimes are possible to be observed. The cut surface is homogeneous, with a soft texture and the color varies from greyish to black. Foci of hemorrhage, necrosis, and cystic changes may be appreciated, as may attachment to nerves.¹² The tumour can reach the bone



Figure 6. A post-second surgery MRI at 3 months and 2 sessions of adjuvant therapy. The tumour is totally removed with no imaging signs for local recurrence.

structures tightly and can sometimes erode them.¹ Under magnification, MSs are well encapsulated and are composed of spindle and focally epithelioid tumour cells, forming mainly fascicular structures and focally small nests. Tumour cells have moderately abundant eosinophilic cytoplasm and centrally located rounded, ovoid and focal elongated nuclei with finely dispersed chromatin and visible delicate nucleoli. In rare cases, larger, pleomorphic tumour cells with prominent nucleoli can be seen; mitoses are rare. There is scant hyalinised stroma with abundance of melanophages. The typical characteristics of the schwannomas such as bodies of Verocay, microcysts, and the abundance of thick, hyalinized vessels in the stroma, are absent.¹⁸

The MS associated with the Carney complex is characterized by a more specific morphology. Thick cords of adipocyte-like cells with light cytoplasm and scattered numerous psammoma bodies are typical.

Intensive melanin accumulation in MS tumour cells is the major difficulty in pathologic diagnosis (**Fig. 7**). Malignant melanoma, neurofibroma, pigmented protuberating dermatofibrosarcoma and melanocytoma are part of the differential diagnosis spectrum and additional histochemical and immunohistochemical studies are often used.^{12,18}

Argentaffin stain impregnation (Fontana-Masson) is widely used and specific for melanin demonstration. Perls' staining (ferric salts demonstration) and PAS can be used in differential diagnosis which should be negative in MS (**Fig. 7**).

Pathological findings play an important role in distinguishing intracranial or intramedullary MS from malignant melanoma. MS is mainly composed of spindle to epithelioid cells growing in short fascicles and sheets and variably abundant melanin pigment.^{4,8} Unlike malignant melanoma which usually has frequent mitosis, MS has rare mitosis. Melanin pigment was present in all cases but showed similar variability, with some cases containing only

small amounts of intracytoplasmic melanin and other cases showing massive melanin deposition at times obscuring cellular detail.^{4,19}

Highly specific immunohistochemical studies like S100, HMB45, Melan A, Vimentin, and SOX-10 can be used. The result is diffuse positive reaction in tumour cells with Ki 67 under 5%. Histologic features of ample cytoplasm, cytoplasmic process, and indiscernible cell border as well as low proliferative index contribute to the diagnosis of MS rather than malignant melanoma.¹⁹ We recommend additional molecular tests like BRAF V600E – positive marker in 90 % of malignant melanoma and negative in MS.

Imaging characteristics and differential diagnosis

The sporadic type of schwannomas typically presents on neuroradiologic study as variably contrast-enhancing paravertebral masses with frequent involvement of adjacent neural foramina and, in some cases, mushroom-like or “dumbbell” profiles indistinguishable from those common to conventional nerve sheath tumours.^{20,21} Psammoma body – rich examples may evidence calcification best appreciated in computed tomography. MRI appearance of MS depends on content of melanin in melanosomes. In MS with lesser than 10% cells with melanin pigmentation presentation is hardly distinguishable from schwannomas (hypointense/isointense on T1WI and isointense/hyperintense on T2 WI) whereas high content of melanin present with hyperintensity on T1 WI and hypointensity on T2WI. Contrast enhancement after gadolinium administration may be homogenous or heterogeneous based on tumour cellularity, hemorrhage, and cystic changes (**Table 1**).¹ Due to the variations in blood supply and vascularization of the tumour, some MS can be misdiagnosed for cavernoma. Susceptibility weighted images (SWI) and gradient recalled echo (GRE) can be helpful in differential diagnosis.

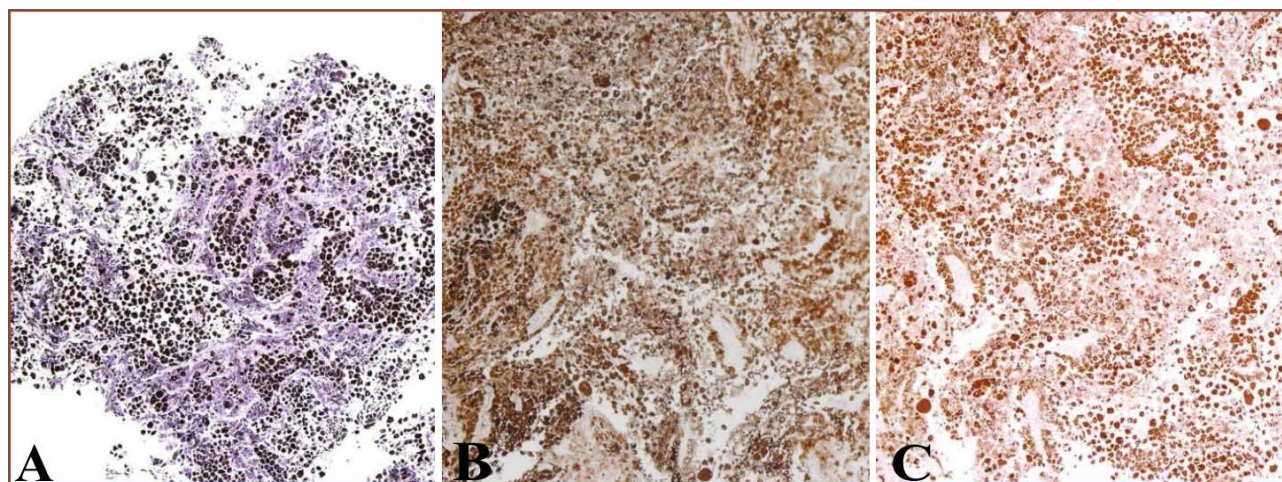


Figure 7. Melanocytic schwannoma microscopic appearance. **A)** H&E staining on 4× magnification – clearly visible black grouped melanosomes. **B)** Melan A staining with positive reaction 4× magnification; **C)** Fe staining (Perls' staining) negative 4× magnification.

Table 1. Radiological differentiation between melanoma, schwannoma, and melanocytic schwannoma using standard MRI sequences T1WI, T2WI and T1 with gadolinium enhancement

Histology	Melanoma	Schwannoma	Melanocytic schwannoma*
WI			
T1 WI	Isointense / hyperintense	Hypointense / isointense	Isointense / hyperintense
T2 WI	Isointense / hypointense	Isointense / hyperintense	Isointense / hyperintense
T1 + contrast	Hyperintense	hyperintense	hyperintense
Homogeneity	+/-	+/-	+/-

Course – metastasis malignant transformation

Operative treatment is the only method for treatment. MS amenability for frequent recurrence (6%-35%) and metastatic spread (9%-42%) is uncommon compared to its “benign” clinical and histological characteristics.²⁻⁶ According to some authors, there is a need of a remodelling paradigm around melanocytic schwannoma’s benignancy, but there are no certain criteria for distinguishing the benign from the malignant type. This may be the cornerstone of creating an adequate guideline for adjuvant treatment. Combination of worrisome histologic features (large, vesicular nuclei, with macronucleoli; brisk mitotic activity; and necrosis) raises concern of aggressive behavior.²² In some series including patients with intradural lesions, malignant PMS developed recurrence of the tumour within 8 months postoperatively, but the patient without malignant tumour was disease free 16 months postoperatively.²³

Only elevated mitotic activity (Z2 mitotic figures/10 HPF) was found to be associated with a higher risk for aggressive behavior and metastasizing in MS, but the lack of it does not corroborate benign course of the disease.⁴ The overall mortality of MS is 10%-17% of all cases in different series.^{3,5,6} Possible ways for local or distant metastasis is by blood flow or via CSF seeding.²³

Need for postoperative adjuvant therapy

Whereas no clear pathological criteria for malignancy are established, the need of adjuvant therapy is disputable and considered controversial by some authors. The RFP (recurrence-free period) in intracranial and intramedullary lesions is between 3 and 20 months. Fifty-six percent of patients underwent incomplete resection and 89% did not receive adjuvant therapy.¹ Comparing data and adjunct to our case, we consider radiation therapy is advocated in cases when: (1) subtotal or partial resection is made with imaging and clinical proof of local recurrence; (2) partial, subtotal or total resection^{24,25} with high mitotic index (Z2 mitotic figures/ 10 HPF)⁴ and aggressiveness^{10,19} local or distant metastasis. In complex decision of radiotherapy care must be taken for adjacent crucial neural structures and possible consequences in developing postradiation complications. In these cases, SRT may be a useful method of choice. In patients with incomplete tumour resection or in those with local

recurrence, SRT also may opt as a single adjuvant therapy. Immunotherapy may be useful to metastatic/malignant MS with therapy used in malignant melanoma.

CONCLUSIONS

Melanocytic schwannoma can be easily described as a tumour in the middle between schwannomas and melanomas depending on which component of the tumour is more presented the more likely the course of the disease will undergo. Meticulous anamnesis and clinical examination is the key to distinguishing sporadic MS from Carney complex related cases. MS are rare tumours and can easily be misdiagnosed with more frequent tumours like melanoma, neurofibroma and hemangiomas based only on radiological signs. That may change and postpone the course of the treatment. Pathological examination using Melan A, Perls staining are keystone for diagnosis of MS. Additional immunohistochemical staining S-100, HMB-45, Vimetin and SOX-10 are also helpful in the pathologic diagnosis corroboration. The gold standard treatment is total resection but the risk for additional neurological deficit must be weighted. Radiological adjuvant therapy is possible and sometimes mandatory especially in MS with aggressive and metastatic behavior. Because of its rarity and no standardized data, suitable guidelines for treatment (especially adjuvant radiotherapy) have not been created until now.

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Рецидивирующая меланоцитарная шваннома L3 Rootlet – клинический случай и обзор литературы

Георги К. Георгиев¹, Иван В. Тодоров¹, Тодор П. Шамов¹, Иван В. Крушовенски¹,
Албена Д. Факирова², Боряна Н. Илчева², Илиан Т. Георгиев¹, Николай Д. Георгиев¹,
Владимир С. Пранджев¹, Тихомир Н. Ефтимов¹

¹ Клиника нейрохирургии, Военно-медицинская академия, София, Болгария

² Кафедра патологии, Военно-медицинская академия, София, Болгария

Адрес для корреспонденции: Георги К. Георгиев, Военно-медицинская академия, ул. „Георги Софийски“ №3, 1606 София, Болгария;
E-mail: georgi_krasimirov91@abv.bg; Тел.: +359 879 171 561

Дата получения: 23 июня 2020 ♦ **Дата приемки:** 30 июля 2020 ♦ **Дата публикации:** 30 июня 2021

Образец цитирования: Georgiev GK, Todorov IV, Shamov TP, Krushovenski IV, Fakirova AD, Ilcheva BN, Georgiev IT, Georgiev ND, Prandjev VS, Eftimov TN. L3 rootlet recurrent melanocytic schwannoma – case report and literature review. Folia Med (Plovdiv) 2021;63(3):448-56. doi: 10.3897/folmed.63.e55295.

Резюме

Меланоцитарная шваннома (МШ) (меланоцитарная шваннома, пигментированная шваннома), впервые описанная Миллером в 1932 году, представляет собой редкую разновидность опухолей оболочек периферических нервов эктодермального происхождения, возникающую в основном в соматической, но также и в вегетативной периферической системе, при этом в литературе описано около 200 случаев. Хотя это в основном доброкачественные опухоли, МШ остается образной и патологической проблемой, и её легко ошибочно принять за более агрессивные опухоли периферических нервов.

Мы сообщаем о случае меланоцитарной шванномы сенсорного корня L3 с систематическим обзором почти 200 случаев, описанных в литературе, представленных в интракраниальной, параспинальной области, грудной, брюшной и тазовой полостях и на коже. Две трети случаев являются частью комплекса Карни.

Мы представляем случай 61-летнего мужчины с трехмесячной историей боли в пояснице, прогрессирующим онемением и ригидностью в правом бедре, икре и колене, парезом большеберцовой кости и малоберцовой кости, вызывающим нарушение походки и неврологическую хромоту. Результаты МРТ представляют интрадуральное экстрамедулярное опухолевое образование типа «песочных часов» с распространением на корешок L3 через правое отверстие L3-L4, гипоинтенсивное на T2 и гиперинтенсивное на T1. Патологическая диагностика спорадической меланоцитарной шванномы проводилась с помощью иммуногистологического и ультраструктурного анализа. Через тринадцать месяцев после тотальной резекции были клинические и МРТ свидетельства рецидива опухоли. Тотальная резекция и радиохирургия были выполнены с безрецидивным периодом в 14 месяцев. Золотым стандартом лечения меланоцитарной шванномы является тотальная хирургическая резекция всех опухолей. Хотя МШ считается доброкачественной опухолью, она имеет местный или метастатический рецидив примерно в 13% случаев. МРТ в большинстве случаев бывает недостаточно, и только полное патологическое и иммуногистологическое обследование является ключом к постановке диагноза. Необходимость в послеоперационной лучевой терапии остаётся спорной. Впервые разработан критерий послеоперационной адъювантной терапии.

Ключевые слова

Комплекс Карни, меланотический, пигментный, рецидивирующий
