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Case Report

Hemorrhagic spinal melanotic schwannoma presenting as acute chest pain: A case report and literature review

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ABSTRACT

Background: Melanotic schwannoma (MS) is a rare variant of peripheral nerve sheath tumor. MS commonly arises along the spinal nerve sheath. Patients most often experience pain along the dermatome of the affected nerve root. Symptoms development is usually insidious. About half of MS cases are associated with Carney complex, a multi-neoplastic disorder. The remaining cases arise spontaneously. About 10–44% of these tumors undergo malignant transformation.

Case Description: We describe a case of hemorrhagic MS presenting as acute chest pain mimicking myocardial infarction, a presentation which has not yet been described in the literature. Neurologic examination did not reveal any abnormalities. Myocardial infarction was ruled out in the ER, and a chest CT angiogram was ordered for evaluation of PE or aortic dissection which revealed an intradural extramedullary dumbbell-shaped mass extending through the left vertebral foramen at the level of T8. MRI revealed a heterogenous mass that was hyperintense with T_2 and hypointense with T_1-weighted imaging. The patient underwent an open laminectomy of the left T8 and T9 vertebrae and gross total resection (GTR) of a hemorrhagic black tumor. Microscopic examination showed fascicles and nests of plump spindle cells with variable intracellular melanin. Immunohistochemistry showed the cells to be positive for S100, SOX10, HMB-45, and MART-1, confirming diagnosis of MS. Two months after the operation, the patient was doing well and is free of recurrence.

Conclusion: GTR is considered the optimal treatment for MS; radiotherapy and chemotherapy may be considered but have not been shown to improve patient outcomes.

Keywords: Carney complex, Melanotic schwannoma, Nerve sheath, Spinal neoplasia

INTRODUCTION

Melanotic schwannoma (MS) is a rare neoplastic lesion, comprising less than 1% of all nerve sheath tumors.[3] Fewer than 150 cases have been reported in the literature to date. MS is considered a variant of schwannoma which has melanogenic capacity, producing a characteristic black appearance grossly.[26] Successful identification of MS requires differentiation from other spinal neoplasms as well as other pigmented lesions, such as metastatic melanoma.

Because of the scarcity of MS cases reported in the literature, demographic and clinical data on this entity are continuing to evolve. Prior reviews have reported a male-female ratio of 1.1:1 with the highest frequency in the fourth decade.[20] MS may arise anywhere along the peripheral nerves including along the sympathetic chain, GI tract, mediastinum, and subcutaneous

areas.[20,40] Reports on the most common locations of MS are mixed, but along the spinal nerve sheath and within the cranium are considered to be the two most common.[3,32,61] Our review of the MS literature revealed 78 cases of sporadic spinal and 20 cases of intracranial MS.[39] Fewer than 20 cases of cutaneous MS have been described.[15] Previous articles have reported the propensity for extramedullary spinal MS to arise from the lumbosacral region in 47.2%, thoracic region in 30.5%, and cervical region in 22.2% of cases, with intramedullary lesions seen extremely rarely.[48] MS is usually a benign pathology but between 10 and 44% of reported cases have undergone malignant transformation and 24–35% recurred.[3,57,61] Recurrence and malignancy have been reported in MS patients treated with gross total resection (GTR) and radiation,[20,41] whereas non-melanotic variations of schwannomas rarely exhibit recurrence when GTR is accomplished.[55]

The presentation of MS is variable and dependent on the location of the tumor and involvement of local structures. As most spinal schwannomas arise from the spinal nerve sheath,[22] pain and paresthesias are the most common presenting symptoms.[4,7,9,11,18,29] When pain is the main complaint, it most commonly occurs along the back, legs, and neck; MS presenting as chest pain has only been reported in two previous cases,[12,17] of which neither mimicked a presentation of myocardial infarction. Muscle weakness and gait disturbances are not uncommon,[60] and a wide range of other neurologic symptoms have been reported.[21,42,52] Most of the reported cases of MS report symptom evolution over multiple months or years before presentation. Our review of the literature returned only one case of spinal MS with a symptom history of less than 1 month.[40] Slow growth of the tumor usually leads to mass effect on the neural elements which causes the slow symptom development in MS. One case of tumor hemorrhage leading to worsening symptoms has been reported.[64] No cases of lesional hemorrhage leading to sudden onset of presenting symptoms have been reported to date.

Half of MS cases are related to the Carney complex, an autosomal dominant inheritance multi-neoplastic syndrome resulting from a *PRKAR1A* gene mutation.[57] This gene normally encodes the r1α regulatory subunit of protein kinase A;[31] binding of this and one other regulatory subunit functions to suppress intracellular PKA activity and limit cell proliferation. In the absence of a functioning r1α, excessive PKA function leads to uncontrolled cell proliferation in various organs.

Historically, Carney complex-associated and sporadic MS have been reviewed together with data analysis including both etiologies.[61]

Here, we discuss a case of sporadic hemorrhagic spinal MS with the only known presentation of sudden onset chest pain mimicking myocardial infarction as well as a literature review of all reported cases of sporadic spinal MS in an attempt to expand on previously reported demographic and clinical data, as well as proper diagnosis and treatment.

CASE PRESENTATION

A 53-year-old man reported to the emergency room with a 2-day history of sudden-onset left chest pain radiating to his left back. The pain was intermittent over the 48 h and felt similar to the pain he experienced in a prior episode of pleurisy. It did not localize to any specific region of the chest and did not involve the left arm. It was partially relieved with NSAID use and resting on his left side. The patient had a 30 pack year smoking history as well as a history of illicit drug use. He had history of hypertension that was controlled with lifestyle changes and did not show any signs of end organ damage. The previous clinic visits showed his blood pressure to be under control and on admission it was 100/78. A review of systems and physical examination did not show any signs of paresthesias, numbness, weakness, or ataxia. Cardiovascular, respiratory, and neurologic physical exams were normal. The patient was given nitroglycerin and fentanyl, which eased the pain. Because of both his history of smoking and hypertension as well as his clinical presentation, a chest X-ray, EKG, and blood work-up including troponin I and D-dimer were performed which returned normal results.

Due to high suspicion for coronary ischemia and other cardiac etiologies related to his history of smoking, hypertension, and illicit drug use, the patient was admitted to further investigate his chest pain. At this point, his pain was completely resolved with fentanyl. As part of expanding the differential diagnosis for chest pain, to rule out pulmonary embolism and aortic dissection, a CT angiogram of the chest was performed and revealed a 4.4 × 2 × 2.1 cm soft-tissue mass compressing the spinal cord at the level of T8-T9. To further characterize the spinal lesion, MRI imaging was obtained and confirmed the presence of a heterogeneous mass at the left T8 that was hyperintense on T_2-weighted and hypointense on T1-weighted images [Figure 1]. Axial scans showed an intradural extramedullary dumbbell-shaped tumor, characteristic of a spinal schwannoma, at the level of spinal nerve T8.

Given the significant mass effect of the mid-thoracic spinal cord and pain symptoms, the patient underwent an open laminectomy and partial facetectomy of T8 and T9. A dark, dumbbell-shaped mass could be seen extending from the left spinal column grossly [Figure 2]. There also appeared to be a hemorrhage within the dural sac near the T8 nerve root. GTR of the lesion was accomplished with sparing of the nerve root. Post-operative histological examination showed fascicles and nests of plump spindle cells, consistent with schwannoma [Figure 3]. Variable amounts of melanin were

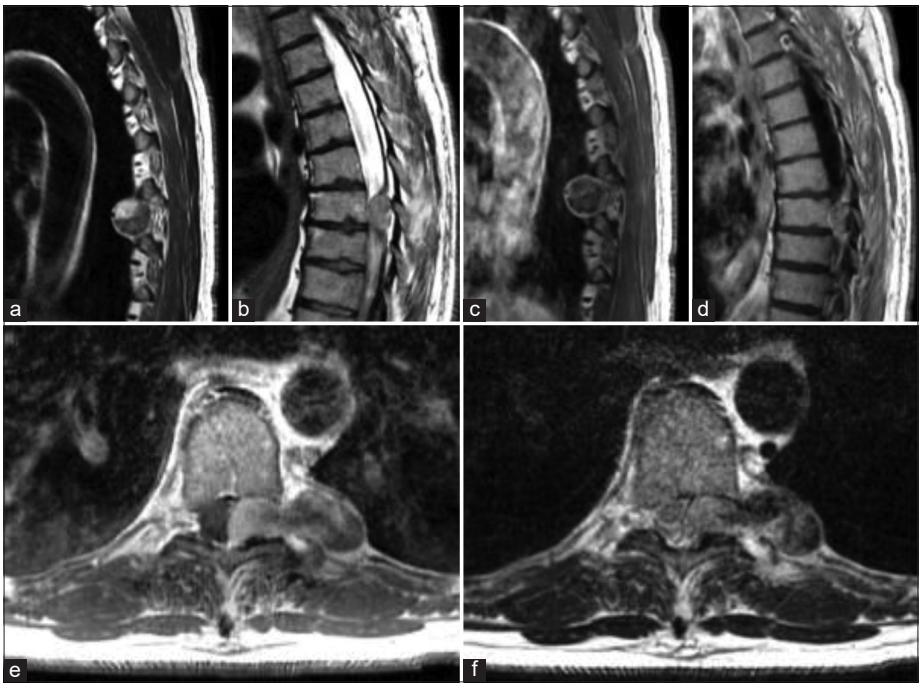

Figure 1: (a and b) Sagittal T_2-weighted MRI scans taken at the time of presentation show a slightly hyperintense heterogeneous mass at the level of T8-T9. (c and d) T1-weighted MRI scans taken after gadolinium administration show a hypointense mass with circumferential enhancement. Axial T_1 (e) and T_2 (f) images show an extramedullary intradural dumbbell-shaped mass.

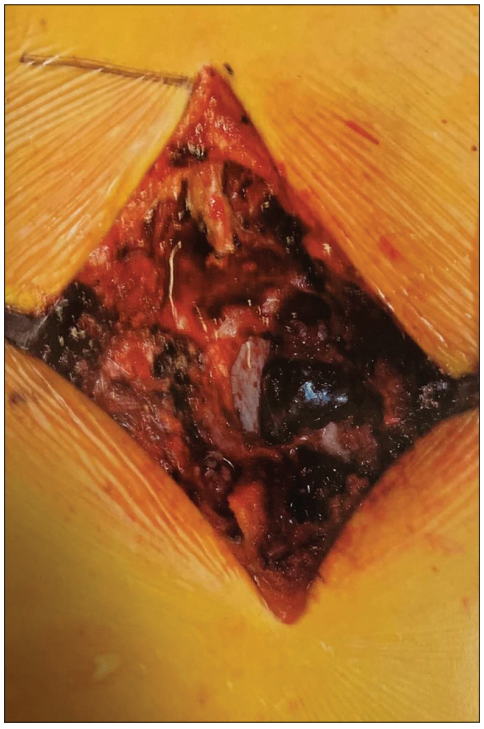

Figure 2: Intraoperative view of the dark, dumbbell-shaped tumor with apparent hemorrhage within the dural sac near the left T8 spinal nerve.

also seen within tumor cells [Figure 3], leading to further immunohistochemical staining. Tumor sections showed

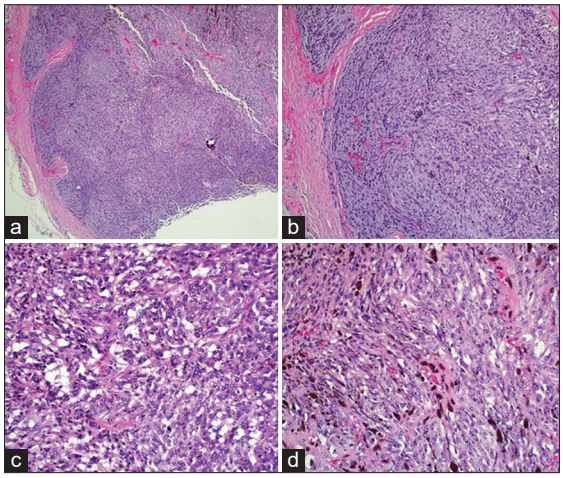

Figure 3: (a) Tumor is circumscribed with cells in fascicles and nests. (b) Fascicles and nests composed of plump spindle cells. (c) Adipose-like cells admixed with tumor cells. (d) Melanin pigment deposition in the tumor cells (hematoxylin-eosin, original magnifications ×40 (a), ×100 (b), ×200 (c and d)).

positive expression of S100, SOX10, HMB-45, and MART-1 [Figure 4], compatible with a diagnosis of MS.

A complete history and review of systems did not reveal any family history or clinical signs of Carney complex in this patient. He was discharged 1 day after surgery. We followed up with the patient 2 months after surgery. He was doing well other than some persisting incisional pain. Imaging did not

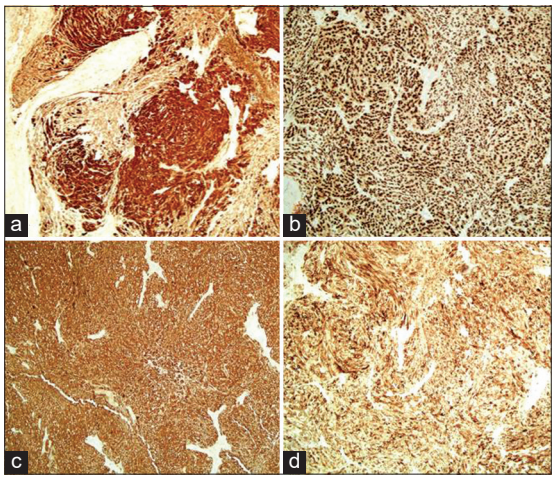

Figure 4: (a) S100 expression in tumor cells. (b) SOX10 expression in tumor cells. (c) HMB-45 expression in tumor cells. (d) MART-1 expression in tumor cells (original magnification ×100 (a through d)).

show any local recurrence of the tumor [Figure 5]. He was counseled on his options regarding radiation and referred to another institution for a second opinion due to the rarity of this diagnosis. He has subsequently been lost to follow-up 6 months after surgery.

DISCUSSION

Sporadic spinal MSs are a rare entity. As more cases are reported, demographic and clinical data are evolving and may change evaluation and treatment of MS. Some prior case reports of MS do not provide in-depth data on patient symptoms, treatment, and follow-up.[8,35,51,61] Furthermore, previous studies have analyzed sporadic and Carney-associated cases of MS as one entity; it may be appropriate to consider these two etiologies separately, as preliminary data suggests variable presentations and histopathology. Accordingly, all patients who present with MS should undergo a search of clinical and family history for signs of Carney complex. Here, we review the epidemiology, diagnosis, and treatment of all sporadic spinal MS cases in the English literature from 1979 to 2020 [Table 1].

MS is considered a tumor of young adults, with a reported mean age of 38 years.[20,34] Carney-associated tumors affect even younger patients, as some reports claim a peak incidence in the third decade.[32,43] Our review of all cases of sporadic spinal MS showed a mean age of 44 years, with a range from 17 to 75 years [Figure 6]. The previous reviews have also reported mixed results on sex predilection: Faria *et al.* reported a 1.1:1 M:F ratio in a review of all cases of MS, while reviews by Kusters *et al.* and Gulati *et al.* reported no sex predilection.[20,23,36] Our analysis reveals a male predominance at a 1.55:1 M:F ratio. These findings may be due to an increase in sample size of reported cases of MS, or to our analysis of sporadic cases only.

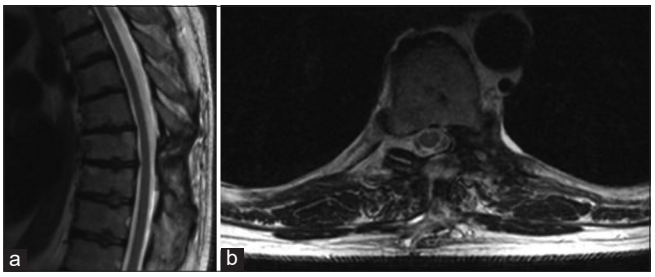

Figure 5: Sagittal (a) and axial (b) T2-weighted MRI images taken 2 months after the removal of a spinal MS tumor.

MS can occur anywhere along the peripheral nerve sheaths.[20] Our review revealed 79 cases of sporadic spinal and 20 cases of intracranial MS. About 70% of intracranial MS tumors are found within Meckel's cave or along the cerebellopontine angle.[2] Peltier *et al.* conducted a review in 2005 that found spinal MS to arise from the lumbosacral region in 47.2% of cases, thoracic in 30.5%, and cervical in 22.2%, with intramedullary tumors considered a separate and rare occurrence.[1,13,28,38,45,53] Our review showed a similar distribution: about 33.8% were thoracic, 27.3% lumbar, 26% cervical, and 13% sacral [Figure 7]. Many previous reviews also reported intramedullary MS to be only of "rare" occurrence without reporting its incidence relative to extramedullary MS.[48] In our review, we found that 11.4% of reported cases of sporadic spinal MS were intramedullary. These discretions may be due to the specificity of our review to only sporadic cases of MS, excluding those associated with Carney complex. The previous reports have not compared these demographic or clinical data between sporadic and Carney-associated cases.

Although several theories have been proposed, the etiology of MS is unknown. Some of the more popular theories include melanomatous transformation of neoplastic Schwann cells, phagocytosis of melanin by Schwann cells, two distinct proliferating cell lines of Schwann cells and melanocytes, and a genetic mutation to a common precursor of melanocytes and Schwann cells as they both arise from neuroectoderm and the cells migrate together.[20,50,54] The development of hemorrhage in our case as well as the propensity of malignant melanoma to bleed[49] may suggest a common precursor cell between melanoma and MS.

Half of MS cases are related to the Carney complex, an autosomal dominant inheritance multi-neoplastic syndrome resulting from a *PRKAR1A* gene mutation.[57] Carney complex is associated with cardiac myxomas, spotty skin pigmentation, blue nevi, and adrenal, testicular, and pituitary adenomas.[10] MS arising in relation to the Carney complex are much more likely than sporadic cases to have psammomatous calcifications upon histological examination.[14]

The presenting symptoms of MS are highly dependent on the location of the tumor. Spinal MS most commonly presents

Table 1: Reported cases of sporadic spinal melanotic schwannoma.

Authors, year	PT age	Sex	Symptoms	Nerve root	Tumor side	Treatment	Metastasis	Recurrence	Follow-up (months/status)
Present case	53	M	2 days back pain	T8	L	GTR	No	No	5/ANED
Nagashima *et al.*, 2020[46]	48	M	6 months back pain, sciatica, dysuria	S2	L	GTR	No	No	6/ANED
Hou *et al.*, 2020[27]	41	F	8 months neck pain, arm numbness, arm weakness	C3	R	GTR	No	12 years later	151/ANED
Sahay *et al.*, 2020[52]	35	M	1.5 months back and leg pain	L2	R	STR+radiation	No	No	6/ANED
Sahay *et al.*, 2020	44	M	Tingling and numbness in upper limbs	C2	NA	STR+radiation	No	3.5 years later	48/ANED
Sahay *et al.*, 2020	35	F	Thigh pain	L3	L	STR	Psoas, paraspinal muscles, lungs	5 months later	36/AWD
Sahay *et al.*, 2020	50	F	5 months leg weakness, paresthesia, bladder incontinence	C6	R	STR+radiation	NA	NA	NA
Takatori *et al.*, 2020[59]	39	M	Back pain, leg numbness	L4	L	STR+radiation	Lungs, spinal cord, chest wall, stomach	No	22/DOD
Alamer and Tampieri, 2019[2]	45	F	Back pain	T6	Intramedullary	GTR	No	No	23/ANED
Alamer and Tampieri, 2019	54	F	Back pain	S3	NA	GTR	No	No	15/ANED
Li and Dai, 2018[38]	61	F	3 years leg pain and weakness	L1	Intramedullary	GTR	NA	NA	NA
Hu and Wang, 2018[28]	40	M	4 months arm numbness	C2	Intramedullary	STR	NA	NA	2/AWD
Cheng *et al.*, 2018[13]	47	M	14 months back pain, zonesthesia, leg weakness	T4	Intramedullary	STR	No	6 years later	72/AWD
Chandran *et al.*, 2018[11]	35	M	Back pain, foot drop	L2	L	STR	No	10 months later	NA
Chandran *et al.*, 2018	25	M	Neck pain	C2	Intramedullary	GTR	No	No	60/ANED
Choi *et al.*, 2017[14]	59	M	Buttock and leg pain	L4	L	GTR	Lung	5 years later	NA
Mahmood *et al.*, 2016[41]	17	M	NA	T3	R	GTR	No	No	12/ANED
Khoo *et al.*, 2016[33]	36	F	4 years hip pain, leg pain	L5	L	STR x2	Brain and meninges	10 months later	NA
Khoo *et al.*, 2016	20	M	4 years back pain	S1	L	STR	NA	NA	NA

(*Contd...*)

Table 1: (*Continued*).

Authors, year	PT age	Sex	Symptoms	Nerve root	Tumor side	Treatment	Metastasis	Recurrence	Follow-up (months/status)
Khoo *et al.*, 2016	46	M	2 years back pain, leg pain, leg numbness	L3	L	STR+radiation	Brain	2 years later	24/DOD
Guzel *et al.*, 2016[24]	36	M	3 months back pain	L5	R	STR	No	No	6/ANED
Shabani *et al.*, 2015[56]	54	M	Incidental finding, on monitoring developed neck and arm pain	C5	L	GTR	Lower spinal nerve root	3 months later	7/DOD
Li and Chen, 2015[37]	62	M	Incidental finding	T7	R	GTR	No	No	30/ANED
Bakan *et al.*, 2015[7]	31	F	Back pain	T4	R	GTR	No	No	6/ANED
Torres-Mora *et al.*, 2014[61]	21	F	NA	C7	NA	NA	No	No	300/ANED
Torres-Mora *et al.*, 2014	39	M	NA	T3	NA	NA	No	1 year later	108/ANED
Torres-Mora *et al.*, 2014	47	M	NA	L4	NA	NA	Lung, liver, pleura, meninges, and ribs	No	10/DOD
Torres-Mora *et al.*, 2014	61	M	NA	T7	NA	NA	No	No	10/DOD
Torres-Mora *et al.*, 2014	47	M	NA	C5	NA	NA	Lumbar/ thoracic and brain	2 years later	48/AWD
Torres-Mora *et al.*, 2014	62	F	NA	T11	NA	NA	No	No	25/ANED
Torres-Mora *et al.*, 2014	27	M	NA	L2	NA	NA	Lungs, thoracic lymph nodes, and abdomen	4, 6, 7, 8, 10 years later	128/AWD
Torres-Mora *et al.*, 2014	32	F	NA	L5	NA	NA	No	No	18/ANED
Torres-Mora *et al.*, 2014	32	M	NA	C2	NA	NA	Lung and skeleton	No	12/DOD
Torres-Mora *et al.*, 2014	62	F	NA	Cauda Equina	NA	NA	No	No	168/ANED
Torres-Mora *et al.*, 2014	19	M	NA	S1	NA	NA	No	No	7/ANED
Torres-Mora *et al.*, 2014	30	M	NA	S1	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	17	F	NA	S1	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	63	M	NA	Sacral	NA	NA	NA	NA	NA

(*Contd...*)

Table 1: (*Continued*).

Authors, year	PT age	Sex	Symptoms	Nerve root	Tumor side	Treatment	Metastasis	Recurrence	Follow-up (months/status)
Torres-Mora *et al.*, 2014	40	F	NA	L3	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	52	F	NA	Thoracolumbar	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	28	M	NA	T10	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	75	M	NA	L2	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	47	F	NA	T12	NA	NA	NA	NA	NA
Torres-Mora *et al.*, 2014	57	F	NA	L3	NA	NA	NA	NA	NA
Mohamed *et al.*, 2014[44]	43	M	2 years leg weakness	T9	L	GTR	No	No	3/AWD
Mahesh *et al.*, 2014[40]	67	M	2 weeks leg weakness, constipation, dysuria	T10	R	STR+radiation	No	No	12/AWD
Chen and Gu, 2013[12]	47	M	Back pain, chest pain, leg weakness and numbness, gait disturbance	T3	L	GTR	No	No	6/ANED
Faria *et al.*, 2013[20]	32	F	6 months neck pain, arm weakness	C5	L	GTR+radiation	Lungs	6 months later	9/DOD
Yokota *et al.*, 2012[63]	64	M	3 years arm paresthesia, gait disturbance	C7	L	STR	Bone, lungs	9 months later	12/DOD
Hoover *et al.*, 2012[26]	62	F	Several year thigh pain, leg weakness	T11	Intramedullary	GTR	No	No	10/AWD
Zhao *et al.*, 2011[64]	46	M	1 year neck pain, hand weakness	C7	L	GTR+radiation	No	No	16/ANED
Shields *et al.*, 2011[57]	65	F	Back pain	T7	R	STR+radiation	No	8 months later	8/DOD
Shields *et al.*, 2011	33	M	Back pain, leg radiculopathy	L5	R	STR+radiation	Lung	2 years later	48/DOD
Izquierdo *et al.*, 2010[42]	29	F	Leg paresthesia, gait disturbance, muscle spasms	T8	NA	GTR	No	No	12/ANED
Rotin *et al.*, 2010[51]	61	M	NA	C3	NA	NA	NA	NA	NA
Arvanitis, 2010[4]	36	M	Back pain, weight loss, leg weakness	L3	R	STR x2	NA	NA	NA
Azarpira *et al.*, 2009[6]	37	M	8 months back pain	L2	L	GTR	NA	NA	NA
Mouchaty *et al.*, 2008[45]	56	F	Quadriplegia	T12	Intramedullary	GTR	No	No	12/AWD

(*Contd...*)

Table 1: (*Continued*).

Authors, year	PT age	Sex	Symptoms	Nerve root	Tumor side	Treatment	Metastasis	Recurrence	Follow-up (months/status)
Marton *et al.*, 2007[43]	30	F	6 months neck pain and spasms	C3	R	GTR	No	12 months later	12/AWD
Er *et al.*, 2007[18]	54	M	Hypoesthesia, pain, weakness of arm and leg	C1	R	GTR	No	No	24/ANED
De Cerchio *et al.*, 2006[17]	53	M	Chest pain, arm pain	T9	R	GTR	No	No	24/ANED
Tawk *et al.*, 2005[60]	61	M	2 years leg weakness	T7	NA	GTR	No	3 months later	11/DOD
Santaguida *et al.*, 2004[53]	35	M	Neck stiffness, arm paresthesia	C5	Intramedullary	GTR	Meninges	4.5 years later	52/ANED
Goasguen *et al.*, 2003[21]	66	F	Pyramidal syndrome of all 4 limbs	C3	NA	NA	NA	NA	NA
Cummings *et al.*, 2000[16]	51	M	8 months low back pain	S2	L	Declined surgery	NA	NA	NA
Vallat-Decouvelaere *et al.*, 1999[62]	35	F	3 years low back pain	L4	NA	GTR	Bone, lymph nodes	6 years later	72/DOD
Vallat-Decouvelaere *et al.*, 1999	27	M	Low back pain	L5	L	GTR	Lung, pleura	No	72/DOD
Vallat-Decouvelaere *et al.*, 1999	34	M	1 year neck pain, paresthesia, and weakness	C1	L	STR	Lower spinal nerve roots	No	84/AWD
Vallat-Decouvelaere *et al.*, 1999	45	F	1 year back pain	T6	L	GTR	Lung, bone, liver	No	36/DOD
Vallat-Decouvelaere *et al.*, 1999	41	F	4 years low back pain	S1	L	STR	No	No	72/ANED
Hollinger *et al.*, 1999[25]	47	M	3 years back and leg pain	T12	L	GTR	No	No	12/ANED
Acciarri *et al.*, 1999[1]	44	F	10 years leg weakness and numbness	T2	Intramedullary	GTR	No	No	4/AWD
Bosman *et al.*, 1995[8]	43	M	NA	L4	NA	NA	NA	NA	NA
Bouziani *et al.*, 1994[9]	46	M	Bilateral sciatica	NA	NA	STR	NA	NA	24/ANED
Krichen *et al.*, 1993[35]	27	F	NA	C7	R	NA	NA	NA	72/ANED
Iizuka *et al.*, 1988[29]	58	F	3 months gait disturbance, back pain, leg numbness	T10	R	GTR	NA	NA	NA
Erlandson, 1985[19]	36	M	3 years back and hip pain, foot paresthesia	S1	L	GTR	NA	NA	NA
Paris *et al.*, 1979[47]	49	F	2 years arm pain	C8	R	GTR+radiation	No	No	48/ANED

GTR: Gross total resection, ANED: Alive with no evidence of disease, STR: Subtotal resection, NA: Not available, AWD: Alive with disease, DOD: Dead of disease

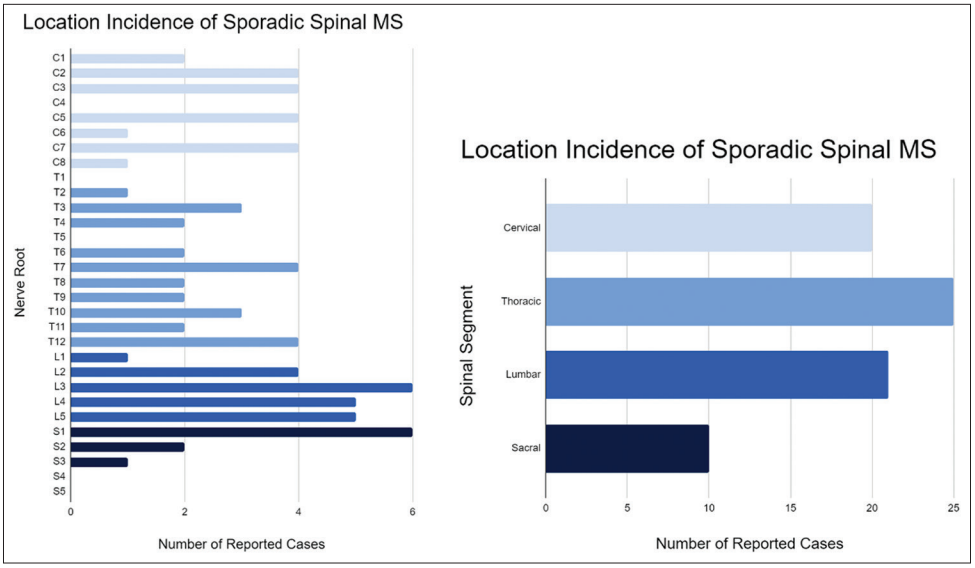

Figure 6: Bar graph visualizing reported cases of sporadic spinal melanotic schwannoma organized by primary nerve root affected. Each shade represents one of the four spinal segments.

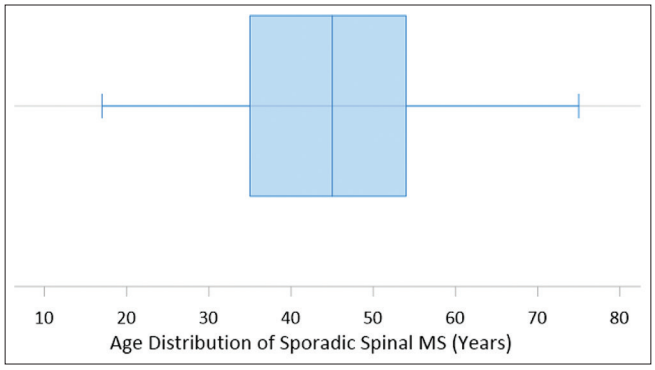

Figure 7: Box plot showing quartiles of patient age in reported cases of sporadic spinal melanotic schwannoma.

with pain correlating with the affected dermatome, often accompanied by paresthesias and muscle weakness of the same region.[3] The development of these symptoms is often insidious, and many patients present with months or years of development of symptoms.[6,16,19,25,33,44,63] Our case appears to be the most acute onset of symptoms reported, as our patient developed acute chest pain over 2 days. The acute development of symptoms may have been due to hemorrhage within the tumor leading to rapid compression of spinal nerve roots. To the best of our knowledge, hemorrhage of a spinal MS has been described only once previously.[64] Furthermore, the presentation of chest pain mimicking myocardial infarction without any associated neurological deficits has never been described as symptomatology for spinal MS.

Common radiologic features of MS include a hyperintense signal on T_1-weighted MRI and a variable isointense to hyperintense signal on T_2-weighted images.[58] This varies from non-MS, which often appears hypointense on T_1-weighted images [Table 2]. MS may be heterogenous on imaging, a finding previously ascribed to tumors that have associated intradural hemorrhage.[64] The characteristic appearance of MS is as a "dumbbell-shaped" tumor on axial view that may be intramedullary or extramedullary and intradural. Greenberg describes a 6-type classification system adapted from Asazuma *et al.* of schwannomas based on foraminal extension.[5,22] The tumor described in this case would be classified as type IIb due to extradural growth and constriction of the tumor at the vertebral foramen.

On gross examination, the tumors have been described as dark brown or black in color, sometimes with hemorrhagic components, cyst formation, or necrosis.[61] They are most often round or ovoid and are surrounded by a thin, fibrous membrane arising from a nerve root; however, they are occasionally lobulated.[3] Erosion or remodeling of the surrounding bone may occur, which further lends credence to the usually slow growth of these lesions.

Classical morphology of MS includes sheets of spindled and epithelioid cells with fascicles of eosinophilic cytoplasm, occasional psammoma bodies, and melanosomes in various stages of maturation within neoplastic cells.[10] The amount of melanin present within cells varies greatly between cases.[3] Some tumors, including our case, may exhibit adipocyte-like cells due to cytoplasmic vacuolization [Figure 3c]. More commonly, the lesion may include trapped adipose tissue.[61] Unlike typical schwannomas, MS tend to lack extensive vasculature. Mitotic activity in these tumors is generally low, but in the Torres-Mora series, elevated mitotic activity of ≥2 figures/10 HPF) was the only clinicopathologic variable associated with aggressive behavior of MS. Lack of mitotic activity, however, was not associated with a benign

Table 2: Comparison of typical spinal schwannoma and melanotic spinal schwannoma.

	Typical spinal schwannoma	Melanotic spinal schwannoma
Peak incidence	40–60 years[34]	35–55 years
Clinical associations	Neurofibromatosis 2 Paresthesias and pain most common presenting symptoms Schwannomatosis	Carney Complex Paresthesias and pain most common presenting symptoms
Radiologic presentation	T1: Hypo/isointense T2: Hyperintense May be heterogeneous on both MRI and CT due to presence of mixed Antoni A/B tissue	T1: Hyperintense due to presence of melanin[34] T2: Hyperintense May be heterogeneous on both MRI and CT due to presence of mixed Antoni A/B tissue
Recommended treatment	Resection if symptomatic	Resection if symptomatic Adjuvant chemotherapy and radiation may be considered
Prognosis	Metastasis exceedingly rare Recurrence common in patients with neurofibromatosis 2	Metastasis in 32.7%

course. Immunohistochemical staining of MS is most often positive for S100, SOX10, HMB-45, Melan-A, p16, and Vimentin.[62] Negative staining for GFAP may be used to differentiate MS from typical schwannoma. Likewise, molecular testing of MS will be negative for *BRAFV600E* mutations to differentiate from malignant melanoma of the spine.[3,30] Some clinical series have suggested that *PRKAR1A* genetic testing may be helpful in determining if the tumor is related to Carney complex, as the microscopic presence of psammoma bodies has poor predictive value for the presence of other features of Carney complex.[61]

At this moment, the only clinical or pathological factors predictive for the prognosis of MS are mitotic activity[61] and advanced age.[20] Classical malignant histologic features such as nuclear abnormalities and necrosis are considered worrisome for future malignant behavior of MS.[50] The previous reviews of both sporadic and Carney complex-associated MS have reported metastasis rates from 15%[62] to 42%.[61] Our review of only sporadic spinal MS revealed metastases in 32.7% of cases,[56] 57.9% of those including metastases to the lungs. Due to expanding data that show more aggressive behavior than previously thought, Torres-Mora *et al.* proposed the reclassification of MS to "melanotic schwannian tumor."

GTR with or without radiation therapy is the favored treatment for MS.[37,46] Depending on the size and local invasion of the tumor, GTR may or may not be possible without inducing significant iatrogenic neurological deficit. In cases of subtotal resection or particularly malignant-appearing tumor, adjuvant therapy has been employed with positive outcomes;[40,47,52,59,64] however, a definite mortality benefit has not yet been shown. The use of chemotherapy in MS has not been thoroughly studied. Because of the potential for malignant transformation to occur more than 10 years after resection,[27] patients should undergo long-term monitoring with serial imaging.

CONCLUSION

MS is a rare neoplasm that is often associated with Carney complex but develops sporadically in about half of reported cases. Seventy-nine cases of sporadic MS arising along the spine have now been described. The presentation of spinal MS varies but most commonly includes an insidious onset of back, leg, or neck pain associated with the affected dermatome over months to years. Our case represents the only case of MS to date that presented as acute chest pain mimicking myocardial infarction and suggests that hemorrhagic spinal lesions should be considered in the differential diagnosis of acute chest pain, especially when cardiac workup is negative. Our review of sporadic MS cases showed a male preference as well as an average age of 44 years, slightly older than previously described. We also found that 11% of reported cases of sporadic spinal MS were intramedullary. Immunohistochemical staining should be used to differentiate MS from malignant melanoma. Gross total excision with long-term serial imaging is recommended.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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