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 T2 and hypointense with T1-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.[¹] 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.[²] 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.[³] MS may arise anywhere along the peripheral nerves including along the sympathetic chain, GI tract, mediastinum, and subcutaneous
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. Our review of the MS literature revealed 78 cases of sporadic spinal and 20 cases of intracranial MS. Fewer than 20 cases of cutaneous MS have been described. Previous articles have reported the propensity for extradural 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 intramedulary lesions seen extremely rarely. MS is usually a benign pathology but between 10 and 44% of reported cases have undergone malignant transformation and 24–35% recurred. Recurrence and malignancy have been reported in MS patients treated with gross total resection (GTR) and radiation, whereas non-melanotic variations of schwannomas rarely exhibit recurrence when GTR is accomplished. 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, pain and paresthesias are the most common presenting symptoms. 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, of which neither mimicked a presentation of myocardial infarction. Muscle weakness and gait disturbances are not uncommon, and a wide range of other neurologic symptoms have been reported. 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. 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. No cases of lesonal 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. This gene normally encodes the r1α regulatory subunit of protein kinase A, 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. 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 T1-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
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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 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 incisinal pain. Imaging did not
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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.[6,35,51,61] 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 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.

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 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 discrepancies 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.[20,30,54] 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 PRKARIA 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 | 48     | M   | 6 months back pain, sciatica, dysuria         | S2         | L          | GTR             | No         | No         | 6/ANED                   |
| Hou et al., 2020   | 41     | F   | 8 months neck pain, arm numbness, arm weakness | C3         | R          | GTR             | No         | 12 years later | 151/ANED                 |
| Sahay et al., 2020 | 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 | NA | NA | NA | |
| Sahay et al., 2020 | 50     | F   | 5 months leg weakness, paresthesia, bladder incontinence | C6         | R          | STR + radiation | Lungs, spinal cord, chest wall, stomach | NA | NA | NA | |
| Takatori et al., 2020 | 39     | M   | Back pain, leg numbness                       | L4         | L          | STR + radiation | No         | 22/DOD                 |
| Alamer and Tampieri, 2019 | 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   | 61     | F   | 3 years leg pain and weakness                 | L1         | Intramedullary | GTR          | NA         | NA         | NA | |
| Hu and Wang, 2018 | 40     | M   | 4 months arm numbness                         | C2         | Intramedullary | STR          | NA         | NA         | 2/AWD                   |
| Cheng et al., 2018 | 47     | M   | 14 months back pain, paresthesia, leg weakness | T4         | Intramedullary | STR          | No         | 6 years later | 72/AWD                  |
| Chandran et al., 2018 | 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                 |
| Chok et al., 2017   | 59     | M   | Buttock and leg pain                          | L4         | L          | GTR             | Lung       | 5 years later | NA | |
| Mahmood et al., 2016 | 17     | M   | NA                                            | T3         | R          | GTR             | No         | No         | 12/ANED                 |
| Khoo et al., 2016   | 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..)
| 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 | 36 | M | 3 months back pain | L5 | R | STR | No | No | 6/ANED |
| Shabani et al., 2015 | 54 | M | Incidental finding, on monitoring developed neck and arm pain | C5 | L | GTR | Lower spinal nerve root | No | 7/DOD |
| Li and Chen, 2015 | 62 | M | Incidental finding | T7 | R | GTR | No | No | 30/ANED |
| Bakan et al., 2015 | 31 | F | Back pain | T4 | R | GTR | No | No | 6/ANED |
| Torres-Mora et al., 2014 | 21 | F | NA | C7 | NA | NA | No | 1 year later | 108/ANED |
| Torres-Mora et al., 2014 | 39 | M | NA | T3 | NA | NA | No | 10/DOD |
| 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 | Lumbar/thoracic and brain | No | 48/AWD |
| Torres-Mora et al., 2014 | 47 | M | NA | C5 | NA | NA | 2 years later | 25/ANED |
| Torres-Mora et al., 2014 | 62 | F | NA | T11 | 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 | Lung and skeleton | No | 18/ANED |
| Torres-Mora et al., 2014 | 32 | M | NA | C2 | NA | NA | No | 12/DOD |
| Torres-Mora et al., 2014 | 62 | F | NA | Cauda Equina | NA | NA | No | 168/ANED |
| Torres-Mora et al., 2014 | 19 | M | NA | S1 | NA | NA | No | 7/ANED |
| Torres-Mora et al., 2014 | 30 | M | NA | S1 | NA | NA | No | NA |
| Torres-Mora et al., 2014 | 17 | F | NA | S1 | NA | NA | No | NA |
| Torres-Mora et al., 2014 | 63 | M | NA | Sacral | NA | NA | No | NA |

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Table 1: (Continued).

| Authors, year       | PT age | Sex | Symptoms                                                                 | Nerve root | Tumor side | Treatment      | Metastasis | Recurrence | Follow-up (months/status) |
|---------------------|--------|-----|---------------------------------------------------------------------------|------------|------------|----------------|------------|------------|-------------------------|
| Torres-Mora et al., | 40     | F   | NA                                                                        | L3         | NA         | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Torres-Mora et al., | 52     | F   | NA                                                                        | Thoracolumbar | NA      | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Torres-Mora et al., | 28     | M   | NA                                                                        | T10        | NA         | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Torres-Mora et al., | 75     | M   | NA                                                                        | L2         | NA         | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Torres-Mora et al., | 47     | F   | NA                                                                        | T12        | NA         | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Torres-Mora et al., | 57     | F   | NA                                                                        | L3         | NA         | NA             | NA         | NA         | NA                      |
| 2014                |        |     |                                                                           |            |            |                |            |            |                         |
| Mohamed et al.,     | 43     | M   | 2 years leg weakness                                                      | T9         | L          | GTR            | No         | No         | 3/AWD                   |
| 2014[44]            |        |     |                                                                           |            |            |                |            |            |                         |
| Mahesh et al.,      | 67     | M   | 2 weeks leg weakness, constipation, dysuria                               | T10        | R          | STR + radiation | No         | No         | 12/AWD                  |
| 2014[49]            |        |     |                                                                           |            |            |                |            |            |                         |
| Chen and Gu, 2013   | 47     | M   | Back pain, chest pain, leg weakness and numbness, gait disturbance         | T3         | L          | GTR            | No         | No         | 6/ANED                  |
| 12[2]               |        |     |                                                                           |            |            |                |            |            |                         |
| Faria et al., 2013  | 32     | F   | 6 months neck pain, arm weakness                                          | C5         | L          | GTR + radiation | Lungs      | 6 months later | 9/DOD                  |
| 20[20]              |        |     |                                                                           |            |            |                |            |            |                         |
| Yokota et al., 2012 | 64     | M   | 3 years arm paresthesia, gait disturbance                                 | C7         | L          | STR            | Bone, lungs | 9 months later | 12/DOD                  |
| 2012[65]            |        |     |                                                                           |            |            |                |            |            |                         |
| Hoover et al., 2012 | 62     | F   | Several year thigh pain, leg weakness                                     | T11        | Intramedullary | GTR         | No         | No         | 10/AWD                  |
| 2012[63]            |        |     |                                                                           |            |                  |            |            |            |                         |
| Zhao et al., 2012   | 46     | M   | 1 year neck pain, hand weakness                                          | C7         | L          | GTR + radiation | No         | No         | 16/ANED                 |
| 2012[64]            |        |     |                                                                           |            |            |                |            |            |                         |
| Shields et al., 2011| 65     | F   | Back pain                                                                | T7         | R          | STR + radiation | No         | No         | 8/DOD                   |
| 2011[36]            |        |     |                                                                           |            |            |                |            |            |                         |
| Shields et al., 2011| 33     | M   | Back pain, leg radiculopathy                                             | L5         | R          | STR + radiation | Lung       | 8 months later | 48/DOD                  |
| 2011[39]            |        |     |                                                                           |            |            |                |            |            |                         |
| Izquierdo et al.,   | 29     | F   | Leg paresthesia, gait disturbance, muscle spasms                          | T8         | NA         | GTR            | No         | No         | 12/ANED                 |
| 2011[23]            |        |     |                                                                           |            |            |                |            |            |                         |
| Rotin et al., 2010  | 61     | M   | NA                                                                        | C3         | NA         | NA             | NA         | NA         | NA                      |
| 2010[51]            |        |     |                                                                           |            |            |                |            |            |                         |
| Arvanitis, 2010[6]  | 36     | M   | Back pain, weight loss, leg weakness                                      | L3         | R          | STR x 2        | NA         | NA         | NA                      |
| 2011[41]            |        |     |                                                                           |            |            |                |            |            |                         |
| Azarpira et al., 2009| 37    | M   | 8 months back pain                                                        | L2         | L          | GTR            | NA         | NA         | NA                      |
| 2009[36]            |        |     |                                                                           |            |            |                |            |            |                         |
| Mouchaty et al., 2008| 56    | F   | Quadriplegia                                                              | T12        | Intramedullary | GTR         | No         | No         | 12/AWD                  |
| 2008[37]            |        |     |                                                                           |            |            |                |            |            |                         |

(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 | 30 | F | 6 months neck pain and spasms | C3 | R | GTR | No | 12 months later | 12/AWD |
| Er et al., 2007 | 54 | M | Hypoesthesia, pain, weakness of arm and leg | C1 | R | GTR | No | No | 24/ANED |
| De Cerchio et al., 2006 | 53 | M | Chest pain, arm pain | T9 | R | GTR | No | No | 12/AWD |
| Tawk et al., 2005 | 61 | M | 2 years leg weakness | T7 | NA | GTR | No | No | 12/AWD |
| Santaguida et al., 2004 | 35 | M | Neck stiffness, arm paresthesia | C5 | | GTR | | 3 months later | 12/AWD |
| Goasguen et al., 2003 | 66 | F | Pyramidal syndrome of all 4 limbs | C3 | NA | NA | NA | NA | 24/ANED |
| Cummings et al., 2000 | 51 | M | 8 months low back pain | S2 | L | Declined surgery | No | No | No |
| Vallat-Decouvelaere et al., 1999 | 35 | F | 3 years low back pain | L4 | NA | GTR | Bone, lymph nodes | 4.5 years later | 52/ANED |
| 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 | 72/DOD |
| Vallat-Decouvelaere et al., 1999 | 45 | F | 1 year back pain | T6 | L | GTR | Lung, bone, liver | No | 72/DOD |
| Vallat-Decouvelaere et al., 1999 | 41 | F | 4 years low back pain | S1 | L | STR | No | No | 72/DOD |
| Hollinger et al., 1999 | 47 | M | 3 years back and leg pain | T12 | L | GTR | | No | 12/ANED |
| Acciarri et al., 1999 | 44 | F | 10 years leg weakness and numbness | T2 | | Intramedullary | GTR | No | 4/AWD |
| Bosman et al., 1995 | 43 | M | NA | L4 | NA | NA | NA | NA | 24/ANED |
| Bouziani et al., 1994 | 46 | M | Bilateral sciatica | NA | NA | STR | No | No | 24/ANED |
| Krichen et al., 1993 | 27 | F | NA | C7 | R | NA | NA | No | 72/ANED |
| Iizuka et al., 1988 | 58 | F | 3 months gait disturbance, back pain, leg numbness | T10 | R | GTR | Bone, lymph nodes | No | 24/ANED |
| Erlandson, 1985 | 36 | M | 3 years back and hip pain, foot paresthesia | S1 | L | GTR | No | No | 24/ANED |
| Paris et al., 1979 | 49 | F | 2 years arm pain | C8 | R | GTR+radiation | No | No | 24/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
with pain correlating with the affected dermatome, often accompanied by paresthesias and muscle weakness of the same region. The development of these symptoms is often insidious, and many patients present with months or years of development of symptoms. 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. 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 T1-weighted MRI and a variable isointense to hyperintense signal on T2-weighted images. This varies from non-MS, which often appears hypointense on T1-weighted images. MS may be heterogenous on imaging, a finding previously ascribed to tumors that have associated intradural hemorrhage. 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. 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. 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. 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. The amount of melanin present within cells varies greatly between cases. Some tumors, including our case, may exhibit adipocyte-like cells due to cytoplasmic vacuolization. More commonly, the lesion may include trapped adipose tissue. 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                                                   | 35–55 years                                          |
| Clinical associations           | Neurofibromatosis 2                                           | Carney Complex                                      |
|                                | Paresthesias and pain most common presenting symptoms         | Paresthesias and pain most common presenting symptoms |
| Radiologic presentation        | Schwannomatosis                                               |                                                     |
|                                | T1: Hypointense due to presence of melanin                     |                                                     |
|                                | T2: Hyperintense due to presence of melanin                     |                                                     |
|                                | May be heterogeneous on both MRI and CT                       |                                                     |
|                                | May be heterogeneous on both MRI and CT                       |                                                     |
|                                | due to presence of mixed Antoni A/B tissue                     |                                                     |
|                                | due to presence of mixed Antoni A/B tissue                     |                                                     |
| Recommended treatment          | Resection if symptomatic                                      | Resection if symptomatic                             |
|                                |                                                               | Adjunct chemotherapy and radiation may be considered |
| Prognosis                      | Metastasis exceedingly rare in patients with neurofibromatosis 2 | Metastasis in 32.7%                                  |

MS is a rare neoplasm that is often associated with Carney complex but develops sporadically in about half of reported cases. Thirty-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.

### CONCLUSION

MS is a rare neoplasm that is often associated with Carney complex but develops sporadically in about half of reported cases. Thirty-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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Nil.

### Conflicts of interest

There are no conflicts of interest.

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