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Primary Spinal Malignant Melanoma Mimicking a Cervical Nerve Root Schwannoma: Case Report and Literature Review

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Abstract

Primary spinal malignant melanoma (PSMM) is a rare cancer of the central nervous system (CNS), and PSMM of the spinal nerve root is even more extraordinary. PSMM of a nerve root can mimic the radiographic appearance of benign nerve sheath tumors, thus resulting in misdiagnosis until tissue diagnosis can be made. A 53-year-old African American woman presented with pain primarily involving the left aspect of her neck and shoulder for 2 years. Magnetic resonance imaging (MRI) of the cervical spine demonstrated a T1-hyperintense, T2-hypointense, homogenously enhancing, dumb-bell-shaped, intradural extramedullary mass extending out through the left C2–3 foramen. A midline incision was used to perform a C2 and C3 laminectomy, and the mass was removed from the cavity. The histopathologic profile was consistent with the diagnosis of malignant melanoma. The present case report adds to the 110 cases of PSMM and the 20 cases of PSMM of the spinal nerve root in the existing body of literature. Radiographic and clinical features resemble that of the much more common schwannoma or neurofibroma requiring immunohistochemical analysis for definitive diagnosis. The optimal treatment for PSMM has not yet been defined due to its rarity and it is therefore important to report such cases in order to share our clinical experiences and provide data to other clinicians treating this uncommon disease.

Keywords
- spine surgery
- malignant melanoma
- nerve root
- case report
- schwannoma

Introduction

Melanoma is the sixth most common cancer in the United States with an increasing incidence.[1,2] While lung and breast cancers are more prevalent than malignant melanoma, metastasis to the central nervous system (CNS) is more likely with melanoma than with breast or lung cancers—40 to 60% of patients with malignant melanoma will be diagnosed with CNS metastases during the disease course. Autopsy studies indicate the incidence of CNS involvement may be even greater, with up to 80% of metastatic melanoma cases

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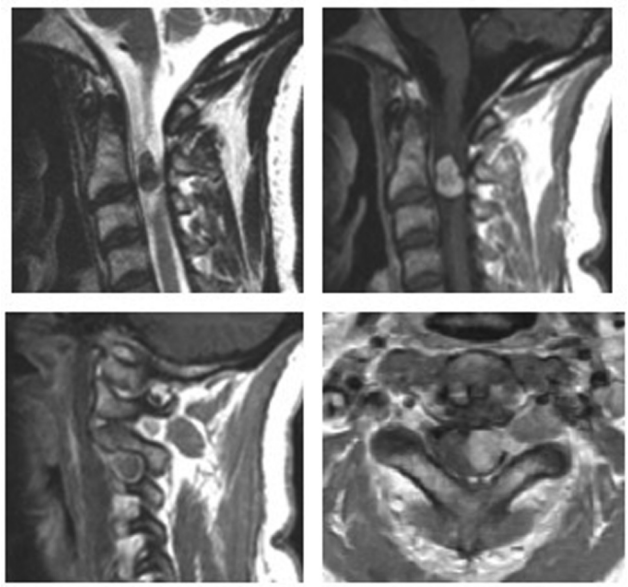

Fig. 1 Preoperative magnetic resonance imaging (MRI) of the cervical spine with and without contrast demonstrated a T1-hyperintense, T2-hypointense, homogenously enhancing, dumbbell-shaped, intradural extramedullary mass extending out through the left C2–C3 foramen.

involving the CNS.[3] While CNS involvement in metastatic melanoma is common, primary malignant melanoma (PMM) of the CNS is quite rare and accounts for approximately 1% of all melanoma cases.[4] Primary spinal malignant melanoma (PSMM) is even less common and PSMM arising from a spinal nerve root is exceedingly rare. PSMM of a nerve root can mimic the radiographic appearance of benign nerve sheath tumors, thus resulting in misdiagnosis until tissue diagnosis can be made. In this case report, we present a case of PSMM arising from a cervical nerve root mimicking a nerve sheath tumor.

Case Report

Presentation

A 53-year-old African American woman presented with pain primarily involving the left aspect of her neck and shoulder for 2 years. The pain also occasionally involved her left mastoid region and posterior aspect of her head. Over the past 2 months, the pain had been progressively worsening. On physical examination, she was noted to be full strength in all major muscle groups. She did not have any signs of myelopathy and her gait was normal. Her sensation to light touch was preserved. Magnetic resonance imaging (MRI) of the cervical spine with and without contrast demonstrated a T1-hyperintense, T2-hypointense, homogenously enhancing, dumbbell-shaped, intradural extramedullary mass extending out through the left C2–3 foramen. The intradural portion of the mass compressed the spinal cord with displacement of the cord to the right. The foraminal portion of the mass resulted in widening of the neural foramen (►**Fig. 1**). Based upon the imaging, the differential diagnosis included schwannoma and neurofibroma, and the patient was counseled on surgical resection of the lesion for which she consented.

Surgery

A midline incision was used to perform a C2 and C3 laminectomy. Upon opening the dura, a dark pigmented mass was identified to the left of and ventral to the spinal cord arising from a spinal nerve root (►**Fig. 2A** and **B**). The mass was gently separated from the spinal cord. The tumor capsule was coagulated using bipolar electrocautery and incised using microscissors. An ultrasonic aspirator was then used to debulk the mass internally to allow greater manipulation of the mass without pressure on the adjacent cervical spinal cord. The rostral attachment to the nerve root was identified, coagulated, and incised (►**Fig. 2C**). We then followed the

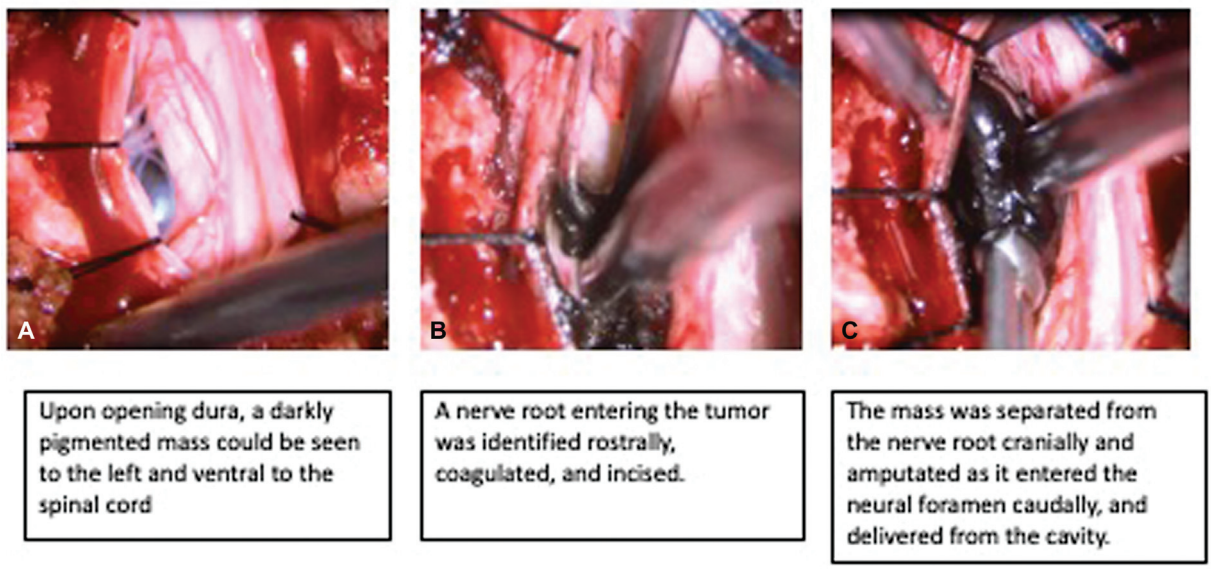

Fig. 2 (**A–C**) Intraoperative imaging when opening the dura with visualization of a dark pigmented mass to the left of and ventral to the spinal cord arising from a spinal nerve root.

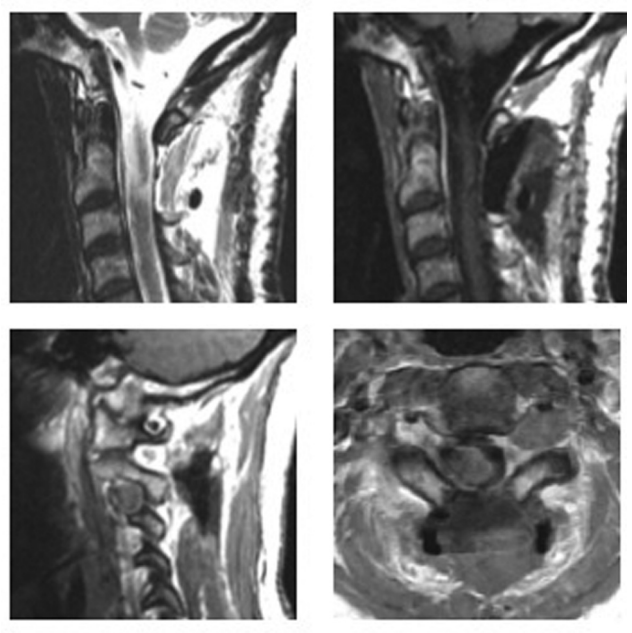

Fig. 3 Postoperative imaging showing removal of the tumor.

mass caudally to its point of exit into the C3 neural foramen and amputated it as it exited the spinal canal. The mass was then delivered from the cavity. The intradural space was irrigated and inspected for any remaining tumor. Residual tumor was intentionally left in the foraminal and extraforaminal spaces due to the much higher surgical morbidity of removing this portion and the persistent belief that this represented a benign mass. The dura was closed primarily using 4–0 silk suture and fibrin sealant (►**Fig. 3**).

Hospital Course

The patient was admitted to the neurosurgical floor postoperatively and recovered without complication. She remained at her neurologic baseline without any neurologic deficit and did report some interval improvement in her baseline left shoulder pain. She was discharged home on postoperative day 2. At her 2-week follow-up visit, the preoperative neck and shoulder pain improved by more than 50% and she had expected incisional soreness.

Pathology

The histopathologic slides demonstrated infiltrating atypical melanocytes with prominent melanin pigmentation, nuclear pseudoinclusions, pleomorphic nuclei, and binucleation. The tumor stained positive for HMB45, Mart-1, SOX-10, and S-100 (►**Fig. 4**) and negative for epithelial membrane antigen (EMA), synaptophysin, and glial fibrillary acidic protein (GFAP). This histopathologic profile was consistent with the diagnosis of malignant melanoma.

Follow-up

A thorough skin examination did not reveal any lesions concerning for melanoma of the skin. MRI of the brain with and without contrast likewise did not demonstrate any lesions or enhancement of the leptomeninges. The patient underwent a whole-body positron emission tomography (PET)/computed tomography (CT) scan that demonstrated prominent fluorodeoxyglucose (FDG) uptake in the laminectomy bed consistent

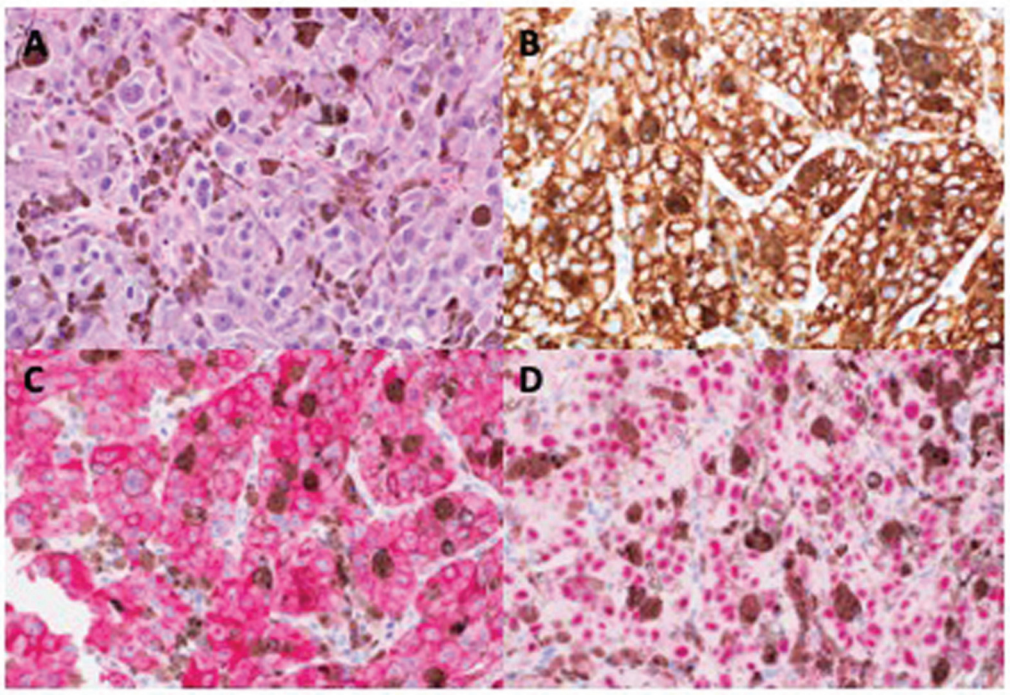

Fig. 4 The tumor is composed of infiltrating atypical melanocytes with prominent melanin pigmentation, abundant eosinophilic and finely granular cytoplasm, nuclear pseudoinclusions, pleomorphic nuclei, large eosinophilic nucleoli, and binucleation. (**A**) Hematoxylin and eosin (H&E), ×400. (**B**) HMB-45, ×400. (**C**) Mart-1, ×400. (**D**) SOX-10, ×400.

with postoperative changes but did not demonstrate any other sites of FDG uptake to indicate additional lesions. Based upon these radiographic findings and the histopathologic profile, a diagnosis of PSMM was made. The patient was treated with adjuvant fractionated radiation therapy (RT) with 35 Gy in five fractions to the resection bed and residual tumor left in the foraminal/extraforaminal space. At 8 months since surgery, the patient is doing well with continued improvement of her left neck and shoulder pain. MRI completed at 5 months after surgery demonstrated no progression of the residual tumor and no intradural recurrence.

Literature Review

We identified in the existing body of literature a total of 112 reported cases of PSMM involving the neural elements, excluding 16 cases of PSMM isolated to the vertebral body (►**Table 1**).[5–27] These 112 cases involved the extradural, intradural extramedullary, and/or intramedullary compartments (►**Table 1**). The thoracic spine was the most common location for PSMM ($n = 45$, 40.2%) followed by cervical ($n = 39$, 34.8%), lumbosacral ($n = 16$, 14.3%), and the thoracolumbar junction or conus medullaris ($n = 12$, 10.7%; ►**Table 1**). PSMM of the spinal nerve root is even more uncommon; including the presently reported case, we identified only 21 cases reported in the literature that identified the tumor arising from the nerve root (►**Table 1**). Among PSMM of the nerve root, the most common location was the cervical spine ($n = 10$, 47.6%) followed by the lumbosacral spine ($n = 7$, 33.3%), and an even distribution between the thoracic ($n = 2$, 9.5%) and thoracolumbar junction or conus medullaris ($n = 2$, 9.5%).

Treatment data were available for 76 cases reported in the literature. All 76 patients underwent surgical resection. Gross total resection (GTR) was achieved in 33 (43.4%) patients, subtotal resection (STR) in 41 (53.9%) patients, and the extent of resection was not reported in the remaining 2 (2.6%) patients. Thirty-five (46.1%) patients underwent adjuvant treatment, which consisted of fractionated RT or chemotherapy or both. The majority of patients ($n = 41$, 53.9%) did not receive any adjuvant treatment. Of the 35 patients who received adjuvant treatment, the most common treatment modality was fractionated RT alone in 23 patients (65.7%). Eight (22.9%) patients received both RT and systemic therapy and four (11.4%) patients received systemic therapy alone. The median fractionated RT dose was 45 Gy (range: 30–60 Gy). Agents used for systemic therapy included dacarbazine, levamisole, temozolomide, cisplatin, carmustine, interferon alpha, interferon beta, and anti-PD1 antibody.

We identified 64 studies that reported survival data for 84 patients. The overall survival (OS) of PSMM varied widely in the literature, ranging from less than 1 month to 25 years from diagnosis with a median survival after diagnosis of 17 months. At the last follow-up, 72.3% of patients were alive. One-year and 3-year OS were 83 and 55%, respectively.

Discussion

The first description of PSMM was reported by Hirschberg in 1906.[28] Since then, only 110 cases of PSMM involving the neural elements have been reported. While PSMM represents an extremely rare cancer, PSMM of the spinal nerve root is even more uncommon; including the presently reported case, we identified only 21 such cases reported in the literature. Whereas PSMM in general was most commonly found in the thoracic spine, PSMM of the spinal nerve root was most commonly located in the cervical spine. The first description of PSMM arising from a spinal nerve root was described by Kiel et al in 1961 in a 33-year-old woman with a melanoma of the left C5 nerve root.[29] The lesion localized to the intradural extramedullary space and extended laterally into the left C5–6 neural foramen. The authors performed a laminectomy for tumor resection and noted a darkly pigmented tumor that involved the left C5 nerve root. In the present case report, we describe a case of a PSMM arising from the C3 nerve root, mimicking a cervical schwannoma, treated with STR and fractionated radiotherapy.

Diagnosis

The diagnosis of PSMM of a spinal nerve root is challenging because the radiographic features resemble that of schwannoma or neurofibroma, and melanoma of the nerve root, as described above, is exceedingly rare. Melanoma within the spinal column is characterized on MRI by T1-weighted hyperintensity, T2-weighted iso- or hypointensity, and mild homogenous enhancement after gadolinium administration, mimicking the radiographic findings of benign nerve sheath tumors.[30,31] Definitive diagnosis, therefore, requires immunohistochemical analysis. Malignant melanoma demonstrates melanocytes with melanin pigmentation on hematoxylin and eosin (H&E) staining and positive staining for HMB-45, S-100, Mart-1, and SOX-10. Schwannoma, on the other hand, is characterized by alternating areas of compact spindle cells with nuclear palisading (Antoni A) and hypocellular areas with myxoid stroma (Antoni B) on H&E staining, and positive staining for S-100 and SOX-10. Importantly, schwannoma demonstrates low-grade cytologic features, whereas malignant melanoma demonstrates cytologically malignant cells. Once a diagnosis of spinal melanoma is confirmed, a thorough workup is required to identify whether the tumor represents a primary tumor without metastases versus metastatic melanoma. This workup should include a thorough skin examination and whole-body PET/CT to identify additional melanotic lesions. MRI of the neuraxis is also indicated to rule out other lesions within the CNS. The ultimate diagnosis of PSMM can then be made according to the criteria described by Hayward: (1) absence of melanoma outside of the CNS, (2) absence of melanoma in another area of the CNS, and (3) histologic confirmation of malignant melanoma.[32] PSMM portends a better prognosis than metastatic melanomas that involve the CNS, making this differentiation between PSMM and metastatic malignant melanoma critical for patient counseling and treatment.[33–35]

Table 1 Literature review of primary spinal malignant melanoma arising from nerve sheath

Author	Year	Age (y)	Sex	Recurrence or metastases (mo)	Survival (mo)	Alive at last follow-up	Level	Location	Extent of resection	Radiation	Systemic therapy
Hirschberg	1906	67	F			Dead	Thoracolumbar	IM			
Boit	1907	51	M				T8–11	Extradural			
Esser	1907	32	M				T1–2	IDEM			
Kawashima	1910	26	F				Cervical	IDEM			
Lindborn	1912	45	F				C1–3	IDEM			
Koelichen	1916	25	M				C7	IM			
Ringertz	1926	61	F				Thoracic	IM			
Schmid	1926	71	M				T7–8	IM			
Bau-Prussak	1929	29	M				Thoracic	IDEM			
Bell	1930	48	F				C7–T1	IDEM			
de Blasi	1930	71	F				T7–8	IM			
Van Bogaert	1933	38	M				T6	Extradural			
Schnitker	1938	49	F		6		T9–10	IDEM			
Da Costa	1939	55	F				T6	IM			
Moersch	1940	55	F		120	Alive	T5				
		49	M		2		T1				
Ray and Foot	1940	29	F		300	Alive	L2				
Garcin	1941	52	M				Cauda equina	IDEM			
Mackay	1942	32	F				Cervicomedullary	IM			
Woods	1944	62	F		6		T9				
Bakody	1950	45	M		228	Alive	L2–4				
Castaner Vendrell	1950	52	F				Cauda equina	IDEM			
Forbes	1950	57	M		2		Thoracic	IM			
Kissel	1950	25	F		11		Cervical	IDEM			
De Assis	1951	26	M		11		Cauda equina	IDEM			
Declich	1952	34			8		T5–6				
King	1952	53	M		4	Dead	Cauda equina	Extradural/IDEM	STR		
		47	M		6	Dead	Cauda equina	IM exophytic	STR		

Table 1 (*Continued*)

Author	Year	Age (y)	Sex	Recurrence or metastases (mo)	Survival (mo)	Alive at last follow-up	Level	Location	Extent of resection	Radiation	Systemic therapy
Perino	1953	40	M				T10–12	IDEM			
Roca de Vinals	1954	50	F				Lumbosacral	IDEM			
Gros and Rotgen	1956						T12–1				
Gibson	1957	51	F				Thoracolumbar	IM	No surgery		
Leger	1957	62	M		42	alive	T6				
Zimmerman and Adams	1958	42	M		4		T9–10				
Lang and Bridge	1959		M		surgical mortality	Dead	Cervical	IM	GTR		
Hirano	1960	42	M	No	6.5	Dead	T8–10	IM		6,000 rads (cobalt)	No
Kiel[a]	1961	33	F	10	25	Dead	C4–6	IDEM	STR		
Holaday[a]	1968	20	F	9	12	Dead	S2	Extradural			
Clifford	1968	64	M	18, metastasis	24	Dead	C4	IDEM	GTR	No	No
Jung	1974	62	F				C2–5	IM	GTR		
Ozden	1984	30	F	No	16	Alive	T7–T10	Extradural		No	Carmustine, dacarbazine, and levamisole
		15	F	No	18	Alive	C1–6	IDEM			
Larson	1987	73	M	No	84	Alive	T6–8	IM	STR	50 Gy	No
		63	M	36	156	Dead	T9	IM	STR	60 Gy	No
		67	F	No	1	Alive	T9–11	IM	STR	45 Gy	No
		57	F	No	30	Dead	C1–3	IM	STR	50 Gy	No
		69	F	No	45	Dead	T9–10	IM	STR	No	No
Schneider[a]	1987	68	F	No	10	Alive	L3–4	Extradural	GTR	Yes	No
Yoo	1987	20	F		17	Alive	C7–T1	IM	STR		
Skarli[a]	1994	20	F	No	36	Alive	C5–6	IDEM	GTR	No	No
Bae	1996	41	M		14	Alive	C3–5	IM	STR	50 Gy	No
Magni	1996	64	M		18	Alive	T8	IM	GTR		
Francois	1998	62	M	No	28	Alive	T8–9	IM	GTR	No	No

(*Continued*)

Table 1 (*Continued*)

Author	Year	Age (y)	Sex	Recurrence or metastases (mo)	Survival (mo)	Alive at last follow-up	Level	Location	Extent of resection	Radiation	Systemic therapy
Salame	1998	76	F	No	21	Alive	T9–10	IM	STR	30 Gy, 8 fractions	No
Salpietro	1998	62	M	Brain metastasis	14	Dead	C3	IM	STR	44 Gy, 22 fractions	No
Brat	1999	71	F	No	22	Alive	T10		GTR		
		52	M	16	16	Alive	C1		STR	40 Gy	No
		20	F	20	20	Alive	C4		STR	NO	NO
		57	F	8	8	Dead	C4		STR	54 Gy (details NS)	No
		53	M	2	2	Alive	Lumbar		STR	NO	NO
Farrokh	2001	80	F	No	9	Alive	T12–L1	IM	STR	NO	NO
Sanz-Trelles[a]	2003	26	M	No	24	Alive	L3	IDEM	GTR	NO	NO
Kwon[a]	2004	45	F	No	8	Alive	C6–7	Extradural	GTR	60 Gy	No
Montinaro[a]	2004	57	F	3, metastasis	3	Alive	L1–2	IDEM	GTR		
Naing[a]	2004	42	F			Alive	L2	Extradural	STR	30 Gy over 2 wk	Interferon alpha
Kounin	2005	41	F	No	3	Alive	C2–4	IDEM	GTR		
Kanatas[a]	2007	76	F	No	6	Alive	C6–7	IDEM	STR	30 Gy, 10 fractions	No
Mekni	2007	34	M		3	Alive	T6–8		STR		
Unal	2007	37	F	No	6	Alive	T7	Extradural	GTR	No	Yes
Nishihara	2009	31	M	216	216	Alive	T6	IM	STR	50 Gy	Interferon beta, intrathecal dacarbazine
Roh[a]	2009	65	M				C6–7	Extradural	STR		
Jo	2010	68	F	No	6.5	Dead	T7–8	Extradural	STR	30 Gy, 10 fractions	Interferon
Kim	2010	34	F	No	36	Alive	T4	IM	GTR	No	No
Kolasa	2010	57	F	9	12	Alive	T10	IM	GTR	No	Yes
Lee	2010	39	M	No	17	Alive	C1–6	IDEM	GTR	45 Gy, 25 fractions	Interferon alpha
Lee[a]	2010	71	F				C6–7	IDEM	STR		

Table 1 (*Continued*)

Author	Year	Age (y)	Sex	Recurrence or metastases (mo)	Survival (mo)	Alive at last follow-up	Level	Location	Extent of resection	Radiation	Systemic therapy
Fuld	2011	62	M	No	11	Alive	C2–3	IM	STR	30 Gy, 10 fractions	No
Jaiswal	2011	40	M	No	4	Alive	C1–2	IDEM	GTR		
		16	M	No	4	Alive	C1–5	IDEM	GTR		
Katalinic[a]	2011	30	M	Yes, metastasis	204	Dead	T7	Extradural	GTR	No	No
Cicuendez	2012	82	F		2	Dead	L2		STR	Yes	No
Ganiusmen[a]	2012	49	F	Metastasis	48	Alive	L3	Extradural	GTR	Yes	Temozolomide
Yan[a]	2012	44	F				L2–4	IDEM	GTR		
Yu	2012	48	M	No	2	Dead	C2–6	IDEM	STR	No	No
Jeong	2013	42	M	22	22	Alive	T2	IDEM	STR	No	No
Sinha[a]	2013	55	M	No	38	Alive	L4	Extradural	GTR		
Cetinalp	2014	47	F	No	9	Alive	T9–L1	IM	GTR	No	No
Li	2014	57	F				T4–T5	IDEM	GTR		
Marx	2014	54	F	No	24	Alive	C2–C3	IDEM	GTR	No	No
Beculic[a]	2015	54	M	No	1	Dead	C5	IDEM/ Extradural	GTR	No	No
Liu[a]	2015	39	M	No	7	Alive	T9–10	IDEM	STR	No	No
		47	M	No	76	Alive	C4–5	IDEM	GTR	No	No
		76	M	No	67	Alive	L2–3	IDEM	GTR	No	No
Mallick	2015	28	M	24	24	Alive	T8–9	IDEM		30 Gy, 10 fractions	Temozolomide
Agarwalla	2016	51	F	No	84	Alive	T7–8	IM	STR	Yes	No
Hering	2016	57	F	No	24	Alive	T12	IDEM	STR	40 Gy, 16 fractions	No
Wu	2017	47	M	24	25	Dead	C2–6	IDEM	GTR	No	No
		47	M	8	10	Dead	T12–L1	IDEM	STR	No	No
		51	M	12	14	Dead	C1–C2	IM	STR	45 Gy, 25 fractions	No
		23	F	No	72	Alive	T6–7	IM	STR	45 Gy, 25 fractions	No
		39	F	No	96	Alive	T4–6	IDEM	GTR	No	No
		57	F	No	38	Alive	C5–6	IDEM	STR	45 Gy, 25 fractions	No

(*Continued*)

Table 1 (*Continued*)

Author	Year	Age (y)	Sex	Recurrence or metastases (mo)	Survival (mo)	Alive at last follow-up	Level	Location	Extent of resection	Radiation	Systemic therapy
		44	F	No	52	Alive	T2–3	IDEM	STR	45 Gy, 25 fractions	No
Martinez[a]	2017	47	M	No	76	Alive	C4–5	IDEM	GTR	No	No
Iga	2018	39	M	No	24	Alive	C2–5	IDEM	STR	No	Anti-PD1 antibody
Wuerdeman	2018	64	F	No	96	Alive	T8	IM	STR	50.4 Gy	No
Zou[a]	2018	42	F	No	16	Alive	C8	Extradural	GTR	Yes	Temozolomide and cisplatin
Chatterjee	2019	78	M	No	18	Alive	C7	IM	GTR	No	No
Hironaka	2019	39	M	n/a	14	Dead	L1–S5	IDEM	No surgery		
Sharma[a]	2019	67	F	9	9	Alive	L1–2	IDEM	STR	No	No
Yoshizaki	2019	49	M	No	60	Alive	T12	Extradural	STR	36 Gy	Dacarbazine
Hanft[a]	2023	53	F	No	8	Alive	C2–3	IDEM	STR	Yes	No

Abbreviations: GTR, gross total resection; IDEM, intradural extramedullary; IM, intramedullary; STR, subtotal resection.
[a]Cases of PSMM arising from the spinal nerve root.

Management

Management of PSMM, and of malignant melanoma in general, requires a multimodality, multidisciplinary treatment approach that includes surgery, RT, targeted therapy, and immunotherapy. A challenging aspect of the treatment of PSMM is that the diagnosis is unlikely to be known prior to surgical intervention because these rare tumors mimic benign tumors of the nerve sheath or meninges (i.e., schwannoma, meningioma). Additionally, because they are primary lesions, there are no lesions elsewhere in the body as would be observed with metastatic disease. Surgeons, therefore, often go into surgery without high suspicion for PSMM. The tailoring of treatment thus does not begin until malignant melanoma is confirmed on immunohistochemical analysis and further workup does not reveal any other melanotic lesions, confirming the diagnosis of PSMM. Because the number of PSMM cases in the literature is so low, there is no accepted standard treatment. If PSMM is somehow known prior to surgery (e.g., biopsy of an extradural lesion or extradural portion of dumbbell-shaped tumor), one may consider aggressive resection to achieve GTR with spinal column stabilization and reconstruction, as necessary.[35] More likely, however, surgeons will resect as much tumor as possible safely without risk of neurological injury, perhaps leaving residual tumor if it is adherent to the spinal cord or if it extends into and/or out of the neural foramen, as we did in our presented case. The question then becomes what to do with the residual tumor. The literature suggests that surgical resection followed by adjuvant fractionated RT may be a reasonable treatment paradigm. Our literature review revealed that 45% of patients underwent adjuvant treatment following surgical resection and the median RT dose was 45 Gy. However, a greater majority of patients who underwent STR went on to receive adjuvant treatments compared to patients who underwent GTR of the lesion (60 vs. 21.9%). All patients with subtotally resected lesions who did receive adjuvant treatment received RT, with or without systemic therapy, except for one patient who received chemotherapy only. In this case, if we had known that the pathology was melanoma, which the darkly pigmented nature of the tumor did suggest, would we have performed a facetectomy with instrumented fusion in an attempt for a GTR? This is a difficult argument to make as there is significant additional morbidity of this extended operation, including increased operative time, increased blood loss, upfront instrumentation secondary to facet takedown, oft-challenging cerebrospinal fluid (CSF) leak repair from resection of the extradural tumor and widened nerve root sleeve, dissection in proximity to the vertebral artery, and persistent possibility of still leaving tumor behind. In drawing upon the growing body of literature that supports the notion of separation surgery in epidural melanoma metastatic cases (as with other malignant metastases) followed by adjuvant radiation, we believe similar logic applies in this case. In essence, this was an intradural version of a separation surgery operation, and we are hopeful that the patient will experience long-term progression-free and OS with this combination of STR and adjuvant radiotherapy.

Survival

The OS of PSMM varied widely in the literature. The majority of publications, however, reported only on a single patient. There exist only three publications that reported on a case series of five or more patients. The earliest, in 1987, reported on a series of five patients with intramedullary PSMM who were treated with subtotal surgical resection followed by adjuvant fractionated radiotherapy (45–60 Gy) in four patients, achieving a mean OS of 6.7 years (median: 45 months).[33] Brat et al reported on their series of primary melanocytic neoplasms of the CNS, including five cases of PSMM.[36] GTR was achieved in one case with no tumor recurrence after 22 months of follow-up. The remaining four tumors underwent STR with two receiving adjuvant fractionated RT. All subtotally resected tumors were found to have recurrence during follow-up. One-year recurrence-free survival (RFS) was 60% (median RFS: 16 months). Finally, Wu et al reported on seven patients with PSMM.[34] Treatment included surgical resection with adjuvant fractionated RT (45 Gy in 25 fractions) for STR. One-year and 3-year RFS were 71.4 and 57.1%, respectively. One-year and 3-year OS were 85.7 and 71.4%, respectively. Of note, all three patients who had tumor recurrence died within 2 months of recurrence diagnosis. Taken altogether, one may conclude from the available data that, although limited due to the small sample size, median survival following a diagnosis of PSMM is around 17 months. Compared to the dismal historical median survival of metastatic melanoma to the CNS of about 4 to 5 months (4.7 months in Davies et al[37] and 5.2 months in Raizer et a[38]) and more recently 6 to 12 months (Kotecha 6 BRAF 9, McHugh 8, Rauschenberg 12)[36,39–41] with combined modern treatment modalities consisting of surgery, radiotherapy, targeted therapy, and/or immunotherapy,[37,38,40,41] PSMM appears to portend a better prognosis. As targeted therapies and immunotherapies continue to evolve, prognosis of both PSMM and metastatic melanoma to the CNS can be expected to improve.

Conclusion

PSMM is a rare cancer of the CNS, and PSMM of the spinal nerve root is even more extraordinary. The present case report adds to the 110 cases of PSMM and the 20 cases of PSMM of the spinal nerve root in the existing body of literature. Radiographic and clinical features resemble that of the much more common schwannoma or neurofibroma requiring immunohistochemical analysis for definitive diagnosis. PSMM may require adjuvant treatment postoperatively to limit recurrence or metastases, unlike benign nerve sheath tumors, which makes the diagnosis crucial for patient survival. The optimal treatment for PSMM has not yet been defined due to its rarity, and it is therefore important to report such cases in order to share our clinical experiences and provide data to other clinicians treating this uncommon disease.

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Conflict of Interest
None declared.

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