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Case Report

Intraspinal mesenchymal chondrosarcoma: An argument for aggressive local resection and adjuvant therapy based on review of the literature

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ABSTRACT

Background: Mesenchymal chondrosarcoma is a rare cartilaginous neoplasm that typically involves the axial skeleton. Despite a well-circumscribed appearance, this tumor has a tendency to recur both locally and with distant metastases.

Case Description: A 17-year-old patient presented with numbness and paresthesias in the lower extremities attributed to a T10-T11 intradural extramedullary mesenchymal chondrosarcoma. The patient was treated with aggressive local resection and adjuvant therapy. Here, this case and present literature are appropriately reviewed.

Conclusion: Although uncommon, intraspinal mesenchymal chondrosarcomas warrant both radical local resection and aggressive adjuvant therapy with chemoradiation to provide the greatest chance of progression-free survival.

Keywords: Adjuvant therapy, Intradural tumor, Intraspinal, Mesenchymal chondrosarcoma

INTRODUCTION

Mesenchymal chondrosarcomas are rare, primary malignant neoplasms of bone and soft tissues that arise from primitive cartilage forming mesenchymal tissue and are capable of distant metastasis.[3,10,13] With an estimated 215 cases per year in the United States, the tumor constitutes 3–10% of all chondrosarcomas.[10,14] Although it grossly appears well-circumscribed with a firm, lobulated outer surface, the tumor is typically invasive.[9] Despite aggressive local resection, these lesions commonly result in distant metastases with an approximated 10-year survival rate of only 27%.[3,10,13,14] The high mortality of the disease warrants aggressive management with wide surgical excision followed by adjuvant radiation therapy. With a very limited number of reported intraspinal cases published in the literature, here, we describe the diagnosis and aggressive surgical treatment of a thoracic intradural extramedullary T10-T11 mesenchymal chondrosarcoma in a 17-year-old male.

Patient case

A 17-year-old male presented with a 5-week onset of progressive lower extremity numbness and hyperreflexia attributed to a primary T10-T11 intradural extramedullary mesenchymal

chondrosarcoma. Contrast-enhanced T1-weighted magnetic resonance imaging (MRI) of the thoracic spine showed an intradural posterolateral right-sided lesion at the T10-11 level compressing the spinal cord anteriorly [Figures 1 and 2]. The patient underwent a T10-T11 laminectomy. The tumor was carefully debulked following a durotomy under the microscope. The lesion was completely removed, and the dura around the area of tumor involvement was coagulated. The frozen section diagnosis was a spindle cell neoplasm. The final pathologic diagnosis was extraskeletal mesenchymal chondrosarcoma.

Due to the high risk of local tumor recurrence, the patient underwent a second operation with further resection of surrounding tissues.[16] A large area of overlying dura was removed, surrounding regions were coagulated, and a large dural patch was inserted. The patient did well postoperatively and underwent adjuvant radiation and chemotherapy (fractionated proton therapy 4500 ccGe with a boost of 540 ccGe to the thoracic spine and vincristine/doxorubicin/ifosfamide × 6 cycles followed by ifosfamide/etoposide × 4 cycles). The patient tolerated these treatments well without incurring any complications.

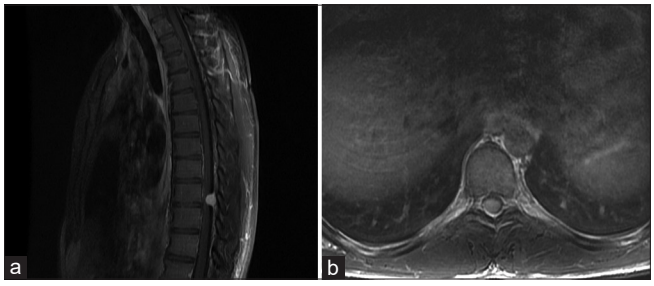

Figure 1: (a and b) Sagittal (left) and axial (right) T1-weighted gadolinium-enhanced magnetic resonance imaging of the thoracic spine reveal an intradural extramedullary homogeneously enhancing mass at the T10-11 level compressing the spinal cord.

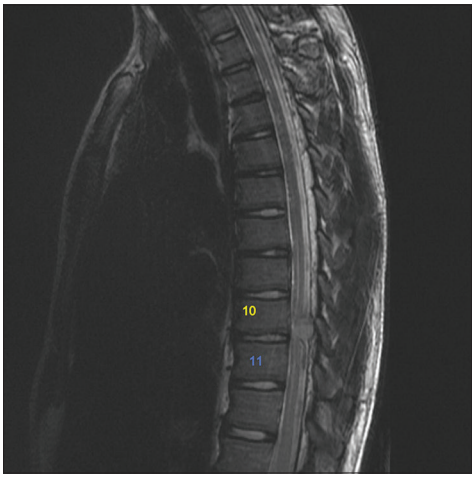

Figure 2: Sagittal T2-weighted magnetic resonance imaging of the thoracic spine reveals a well-circumscribed lesion with a dural tail and a dilated central canal but no evidence of cord edema.

DISCUSSION

Background

Mesenchymal chondrosarcomas are rare tumors, constituting 25% of all chondrosarcomas in children and adolescents.[4,10] These tumors arise from the pluripotent mesenchymal cell which gives rise to all forms of connective tissue, including the dura and the leptomeninges.[8] Previous intraspinal cases have been described in patients younger than 20 years of age [Table 1]. While older patients more often have mesenchymal chondrosarcomas in bone, younger patients typically have extraskeletal lesions.[10]

Diagnosis

On MRI imaging, these tumors often have extraosseous extension, lytic lesions, poorly defined periosteal reaction, and mottled calcification.[6] Notably, intraspinal mesenchymal chondrosarcomas with dural attachment have a more favorable prognosis potentially due to early diagnosis from signs and symptoms of acute cord compression.[12] Histopathologic diagnosis is essential for differentiating cartilage forming mesenchymal tumors including intraspinal mesenchymal chondrosarcomas from other solid tumors such as meningiomas and other cartilaginous lesions,[9] which is critical as mesenchymal chondrosarcomas have a higher probability of recurrence and metastasis.[12]

Management

Mesenchymal chondrosarcoma is known to have a late recurrence, sometimes even more than 20 years after the primary tumor occurs.[12] Surgical management is essential for local control of disease, and better survival rates are seen with wide surgical resection.[2,12] Given the high frequency of local recurrence and the potential presence of tumor cells in the cerebrospinal fluid, postoperative radiation to the tumor bed may be vital in maintaining or prolonging the disease-free period and decreasing the risk of metastasis.[9,15] De Amorim Bernstein *et al*. used a 60 Gy dose of neoadjuvant and adjuvant treatment to achieve a 79% 10-year overall survival rate.[5] In addition, Kawaguchi *et al*. found that the rate of local recurrence was reduced in another cohort of patients receiving 50–59 Gy doses of radiotherapy postoperatively.[11] Here, we utilized postoperative adjuvant radiotherapy in an effort to prevent local recurrence.

The use of chemotherapy in the treatment of mesenchymal chondrosarcomas in the literature is controversial.[16] Frezza

Table 1: Previously reported cases of intradural extramedullary mesenchymal chondrosarcoma.

Case	Author	Year	Age	Gender	Level	Presenting symptoms	Treatment	Survival
1	Derenda	2017	22	F	T12-L1	Bladder/bowel incontinence, radiculopathy, paresthesias, areflexia	GTR+RT	Recurrence, 4 years
2	Chen	2016	26	F	L4	Radiculopathy, weakness, urinary incontinence, hyporeflexia	GTR	Recurrence, 5 months
3	Yang	2016	33	F	L2-3	Hypothesia	GTR	Disease free, 3 years
4	Bishop	2015	12.8	F	Unknown	Unknown	STR+CT+RT	AWD, 4.3 years
5	Andersson	2014	10	F	T4	None	GTR+RT	Disease free, 2 years
6	Turel	2013	6	M	T9	Back pain, weakness, hyperreflexia,	GTR	Unknown
7	Lee	2014	17	M	Multiple lesions, C7-L5	Radiculopathy, paresthesias	Biopsy	Unknown
8	Iida	2014	10	F	L4	Radiculopathy, hypoesthesia	GTR	Disease free, 3 years
9	Patniak	2012	46	M	Multiple lesions, disemmination	Weakness, bowel/bladder incontinence, altered mental status	Biopsy+VP shunt	Deceased, 5 days
10	Bae	2011	25	M	T7	Back pain, weakness	GTR+CT+RT	Disease free, 2 years
11	Belhachmi	2008	13	F	T7-8	Back pain, weakness, hypothesia	GTR	Disease free, 2 years
12	Li	2007	3	F	T11-L1	Weakness, bowel/bladder incontinence	GTR+RT	Disease free, 2 months
13	Chen	2005	11	M	C7-T2	Unknown	GTR	Disease free, 2 years
14	Kotil	2005	40	M	T12	Back pain	GTR	Disease free, 26 months
15	Platania	2003	42	F	T12-L2	Back pain, weakness, urinary incontinence	GTR+RT+CT	Disease free, 6 years
16	Huang	2003	21	M	T8	Back pain, numbness, weakness, hyporeflexia	GTR+RT+CT	Disease free, 8 months
17	Berberoglu	1996	9	F	L4-S2	Weakness, hypoesthesia, hyporeflexia	Biopsy+RT+CT	Deceased, 6 months
18	Rushing	1995	21	F	L5	Back pain, weakness, urinary incontinence, numbness	GTR	Disease free, 1 year
19	Ranjan	1994	52	M	C3-6	Weakness, spasticity, urinary incontinence, hyporeflexia	GTR	Unknown
20	Huckabee	1991	7	F	L3	Back pain, weakness	GTR	Unknown

et al. demonstrated fewer recurrences in patients with localized disease receiving chemotherapy.[7] In contrast, Bishop *et al*. suggested that mesenchymal chondrosarcomas are rather chemoresistant with only one in six patients in their cohort responding to treatment, and De Amorin Bernstein *et al*. demonstrated no statistically significant improvement in disease-free survival.[1,5,11,14] Nakashima *et al*. and Kawaguchi *et al*. also did not observe benefits with chemotherapy in their cohorts. While Cesari *et al*. demonstrated some effectiveness of chemotherapy, reporting that disease-free survival in patients between 5 and 10 years after surgical excision was 76% with chemotherapy and 17% without chemotherapy, the overall survival rate at 10 years between the two cohorts was not statistically significant.[2] Given the young age of the patient in our case, adjuvant chemotherapy was included in the aggressive treatment regimen.

CONCLUSION

Intradural extramedullary mesenchymal chondrosarcomas are rare tumors that require prompt diagnosis, aggressive wide surgical excision, and adjuvant radiation and chemotherapy to achieve the best outcomes. Here, a 17-year-old male with a T10-T11 mesenchymal chondrosarcoma underwent a secondary operation with total *en bloc* resection, including dural removal with patch grafting plus adjuvant chemoradiation to avoid tumor recurrence.

Declaration of patient consent

Patient's consent not obtained 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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