Case Report

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

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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. With an estimated 215 cases per year in the United States, the tumor constitutes 3–10% of all chondrosarcomas. Although it grossly appears well-circumscribed with a firm, lobulated outer surface, the tumor is typically invasive. Despite aggressive local resection, these lesions commonly result in distant metastases with an approximated 10-year survival rate of only 27%. 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. 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 cGy with a boost of 540 cGy 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.

DISCUSSION

Background

Mesenchymal chondrosarcomas are rare tumors, constituting 25% of all chondrosarcomas in children and adolescents. These tumors arise from the pluripotent mesenchymal cell which gives rise to all forms of connective tissue, including the dura and the leptomeninges. 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.

Diagnosis

On MRI imaging, these tumors often have extraosseous extension, lytic lesions, poorly defined periosteal reaction, and mottled calcification. 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. 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, which is critical as mesenchymal chondrosarcomas have a higher probability of recurrence and metastasis.

Management

Mesenchymal chondrosarcoma is known to have a late recurrence, sometimes even more than 20 years after the primary tumor occurs. Surgical management is essential for local control of disease, and better survival rates are seen with wide surgical resection. 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. De Amorim Bernstein et al. used a 60 Gy dose of neoadjuvant and adjuvant treatment to achieve a 79% 10-year overall survival rate. 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. 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.
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.[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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