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Case Report

Spinal mesenchymal chondrosarcoma: A case report of a rare malignant tumor

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ABSTRACT

Background: Mesenchymal chondrosarcoma is an uncommon malignant variant of chondrosarcoma that mainly affects the bones and cartilaginous tissues, but may rarely involve the spine. Careful preoperative planning for surgical tumor removal and spine reconstruction is mandatory and must be based on oncologic and surgical staging.

Case Description: Over 1 month, a 16-year-old female became paraplegic with a T9 sensory level and urinary dysfunction. The magnetic resonance imaging revealed an intraspinal extradural T7-T9 mass that was isointense in T1W1 and markedly enhanced with gadolinium. The patient underwent gross-total tumor resection followed by an osteoplastic laminectomy with fusion. The histological examination was consistent with a mesenchymal chondrosarcoma. She had received radiation and chemotherapy. One year later, she was readmitted for tumor recurrence with multiple metastases involving L1, the lung, and peritoneum. Despite full course of radiotherapy and chemotherapy, she died after 6 months of the second surgery.

Conclusion: Total resection of mesenchymal chondrosarcomas is the gold standard for treatment and is typically followed by radiation and/or chemotherapy. However, the status of residual tumor, local extension, and or metastases best determine the overall survival which may prove extremely limited.

Keywords: Chondrosarcoma, Mesenchymal chondrosarcoma, Metastatic chondrosarcoma, Spinal tumors

INTRODUCTION

Chondrosarcoma is the third most common bone tumor after myeloma and osteosarcoma, and the second most common malignant spinal tumor (i.e., represents 12% of all spinal neoplasms). Thoracic vertebrae most commonly site affected.[4] Surgery optimally includes marginal/wide marginal excision, but is limited due to surrounding crucial spinal structures. Metastases and recurrences are the two main problems that determine the survival rate.[1,3] Here, we describe a 16-year-old with a T7-T9 malignant chondrosarcoma who underwent gross-total tumor excision. Nevertheless, 1 year later, and despite radiation, and chemotherapy, she presented with metastatic disease and died 6 months later.

CASE REPORT

A 16-year-old girl presented with a paraplegia that had been progressing for 1 month (0/5 in the lower extremities with T7 pin level) and sphincter dysfunction (urinary retention and constipation). Thoracic magnetic resonance imaging (MRI) showed a mass between T7 and T9 that was isointense on T1W1 and markedly enhanced with gadolinium [Figure 1]. Whole-body CT scan additionally showed adenopathy of the hilus. The patient underwent an initial laminectomy from T7 to T9 with gross-total resection of an extradural tumor with transpedicular screw fixation. At surgery, the lesion was gray, firm, and vascular.

Pathology

The histology revealed round, oval, and atypical cells with evidence of mitosis, and well-differentiated islet cells with a cartilaginous component [Figure 2]. The immunohistochemistry/biomarkers revealed; CD99 (mesenchymal cells), PS100, Desmine, Myogénine, and Cytokératine, with positive, CD99. These findings confirmed the diagnosis of amesenchymal chondrosarcoma.

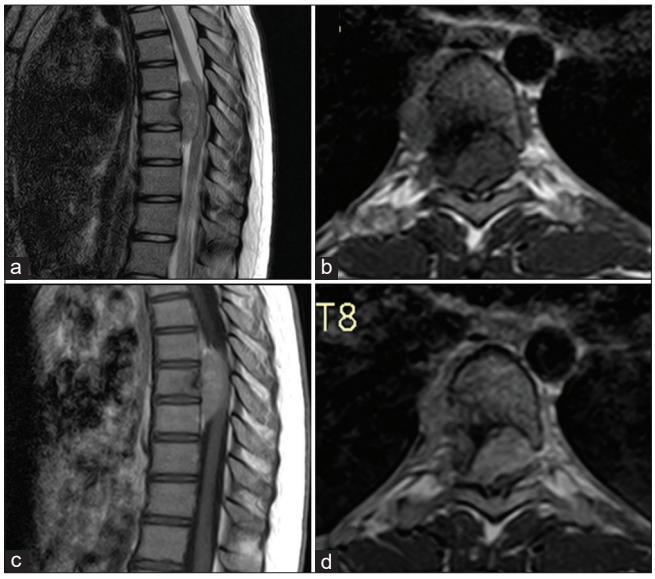

Figure 1: Spinal magnetic resonance imaging (a and b) showing extra axial mass from T7 to T9 compresses the spinal cord isointense in T1WI, high signal after gadolinium injection (c and d).

Postoperative course

Postoperatively, she remained full neurological function and the postoperative MRI confirmed total tumor resection. She received radiation and chemotherapy (32 sessions of vincristine, Actinomycin D, and Ifosfamide). However, 1 year later, she again became paraplegic (2/5 in the lower extremities with sphincter dysfunction). Spinal MRI showed a recurrent T7-T9 mass and metastatic disease affecting L1 leading to compression of the filum and conus [Figure 3]. She underwent laminectomy at L1 and recurrent decompression of T7-T9 for repeat partial tumor excision. When she developed dyspnea and abdominal pain 2 weeks later, a whole-body CT examination revealed pneumonia, ascites, and peritoneal carcinomatosis [Figure 4]. Despite further palliative chemotherapy, she died 6 months later.

DISCUSSION

Histology

Malignant mesenchymal chondrosarcomas (MCS) comprise about 10% of all chondrosarcomas. They are typically extradural and thoracic in location. Histologically, they are characterized by small round-cells with focal cartilaginous differentiation, and exhibit a unique, biphasic pattern including both mesenchymal and chondrocytic components.

Immunohistochemistry

Typical immunohistochemistry findings include positivity of the mesenchymal portion for vimentin, Leu7, and CD99, positivity of the cartilaginous regions for S100 protein.[5] Additionally, there is accompanying positivity of new markers; Sox9, collagen Type II, and FLI-1.[2] The HEY1-

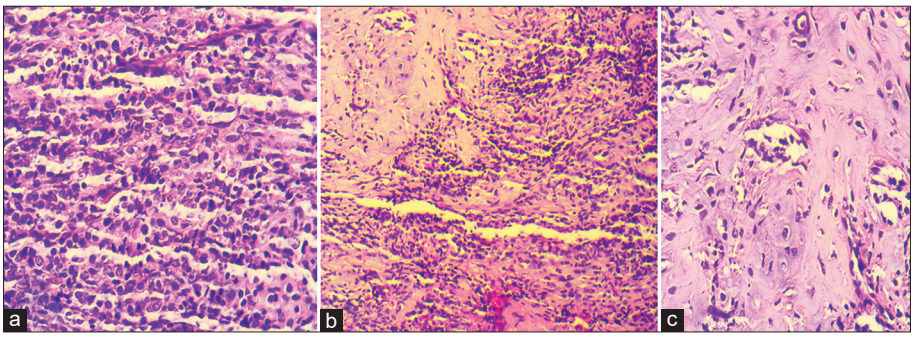

Figure 2: Histological image showing (a) sarcomatous proliferation of round and oval cells with the presence of well-differentiated cartilaginous islands, (b) cartilaginous islands, and (c) poorly differentiated sarcomatous proliferation of round and oval cells.

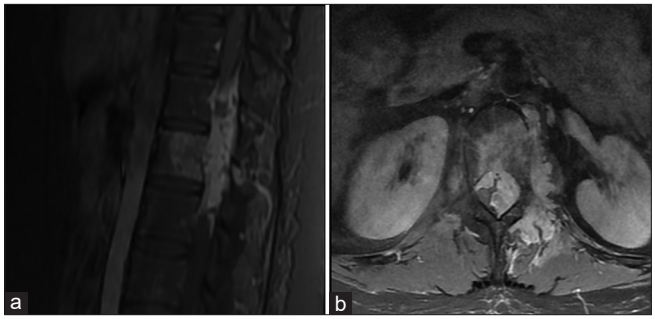

Figure 3: Spinal magnetic resonance imaging (a and b) showing metastases at L1 level.

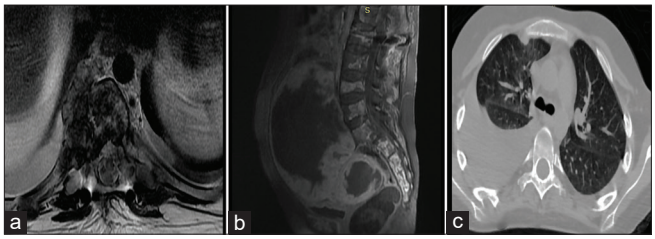

Figure 4: (a and b) spinal magnetic resonance imaging showing multiple metastatic lesions (spine and peritoneum) and mass recurrence at L1 level. (c) Chest CT-scan showing pulmonary effusion.

NCOA2 fusion gene on fluorescence *in situ* hybridization presently suggests that it will assist the diagnosis of MC, especially when the tumor has little cartilaginous component and presents as an undifferentiated small round-cell sarcoma. All of these findings help establish the diagnosis of MCS, especially when the tumor has only a minor cartilaginous component, and presents as an undifferentiated small round-cell sarcoma.

CT and MR scans for MCS

X-rays for MCS show nonspecific osteolytic changes and/or defined soft tissue or bony mass with calcification. With CT/MRI scans, MCS findings are nonspecific and must be differentiated from osteosarcomas, myeloma, or other lesions. MRI studies best identify the levels and extent of parameningeal lesions cord compression. Although gross total local resection is the treatment of choice, survival rates for MCS typically depend on the initial extent/severity of both primary and metastatic disease (i.e., to the lungs, soft tissues, and other organs). In addition, these tumors are resistant to both radiation and chemotherapy.

CONCLUSION

Malignant spinal mesenchymal chondrosarcomas are rarely spinal lesions that may cause paraplegia. Although the gold standard is gross total tumor excision, even adequately resected primary tumors are likely to recur locally and/or metastasize, thus markedly restricting long-term survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Frezza AM, Cesari M, Baumhoer D, Biau D, Bielack S, Campanacci DA, *et al*. Mesenchymal chondrosarcoma: Prognostic factors and outcome in 113 patients. A European Musculoskeletal Oncology Society study. Eur J Cancer 2015;51:374-81.
2. Hu X, Li L, Eid JE, Liu C, Yu J, Yue J, *et al*. IDH1 mutation induces HIF-1 α and confers angiogenic properties in chondrosarcoma JJ012 Cells. Dis Markers 2022;2022:7729968.
3. Miao R, Choy E, Raskin KA, Schwab JH, Nielsen GP, Deshpande V, *et al*. Prognostic factors in dedifferentiated chondrosarcoma: A retrospective analysis of a large series treated at a single institution. Sarcoma 2019;2019:9069272.
4. Schwab JH. Primary tumors of the Spine. Orthop Knowl Updat Spine 2018;4:465-72.
5. Shakked RJ, Geller DS, Gorlick R, Dorfman HD. Mesenchymal chondrosarcoma: Clinicopathologic study of 20 cases. Arch Pathol Lab Med 2012;136:61-75.

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