Clinical Image: Histiocyte-rich Rhabdomyoblastic Tumor

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A 70-year-old man with a history of multiple myeloma on maintenance therapy who was previously treated with induction and subsequent high-dose chemotherapy followed by autologous stem cell transplantation that resulted in a stringent complete response presented with abnormal imaging of the left lower extremity six months after transplantation. The patient did not experience any left lower extremity pain or discomfort. The physical examination was unremarkable. Magnetic resonance imaging showed an intensely enhancing enlarged and globular mass within the myotendinous junction of the tibialis anterior in the proximal leg, measuring approximately 5.8 x 1.2 cm without any surrounding inflammatory changes. (Figure.1.a) Surgical pathology showed a diffuse infiltrate of low-grade appearing cells with ample eosinophilic cytoplasm and areas that look plasmacytoid and/or rhabdoid. (Figure.1.b,c) CD138 and MUM1 were both negative, which is highly unexpected for a plasma cell neoplasm. (Figure.1.d,e) Desmin, a marker of muscle differentiation, was diffusely positive. (Figure 1.f). A diagnosis of a histiocyte-rich rhabdomyoblastic tumor was made. Treatment with surgical resection was done, and the maintenance therapy for multiple myeloma was continued. At a 12-month follow-up visit, the patient's imaging showed no evidence of recurrence.

Histiocyte-rich rhabdomyoblastic tumors are a recently described skeletal muscle neoplasm of uncertain malignant potential.1 The tumor is characterized by a pronounced proliferation of non-neoplastic histiocytes that obscure the underlying tumor, and it was identified in ten cases, predominantly in middle-aged males. These tumors have distinct features, including the presence of foamy macrophages, multinucleated giant cells, and spindled macrophages, making it challenging to identify the underlying rhabdomyoblastic tumor using typical markers. A previous study by Martinez et al. suggests that these tumors may represent a new category of skeletal muscle tumors with intermediate (borderline) malignancy. It hypothesizes that they could be termed "rhabdomyoblastic tumors of uncertain malignant potential." In that study, clinical follow-up indicated that all patients remained disease-free after treatment, with no recurrences or distant metastases observed.1 This case underscores the importance of obtaining a biopsy in cases of suspected relapse of multiple myeloma to exclude other pathologies.

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Figure 1a . Magnetic resonance imaging with intensely enhancing enlarged and globular mass within the myotendinous junction of tibialis anterior in the proximal leg, measuring approximately 5.8 x 1.2 cm without any surrounding inflammatory changes.

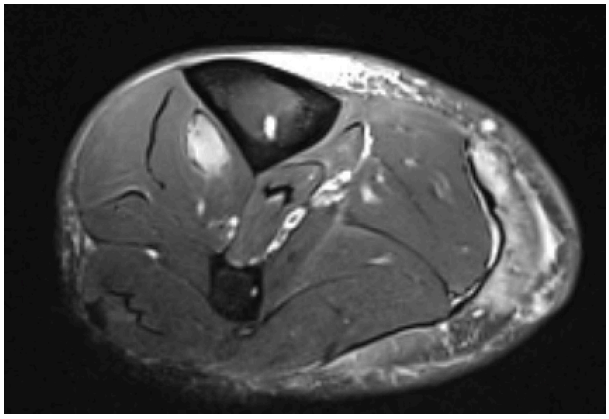


Figure 1b and c . Hematoxylin and eosin stain show diffuse infiltrate of fairly low-grade appearing cells, which have ample eosinophilic cytoplasm and have areas that look plasmacytoid and/or rhabdoid.

Figure 1b

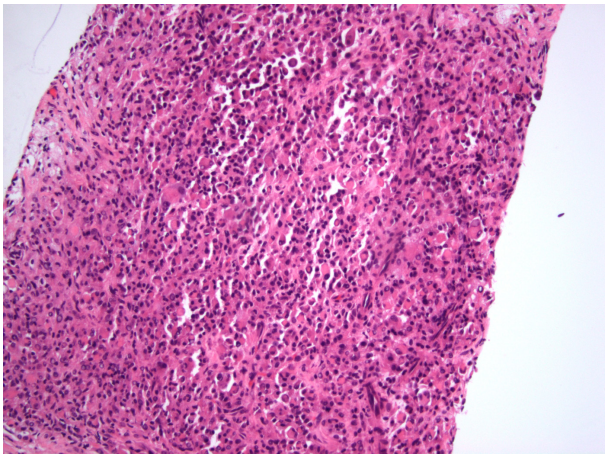


Figure 1c

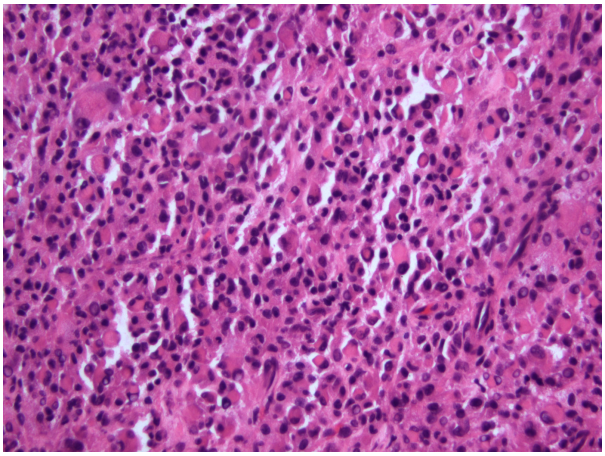


Figure 1d . positive desmin stain

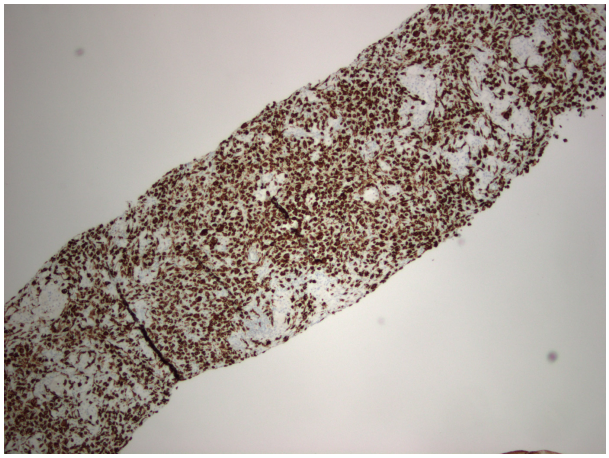


Figure 1e . negative CD-138 stain

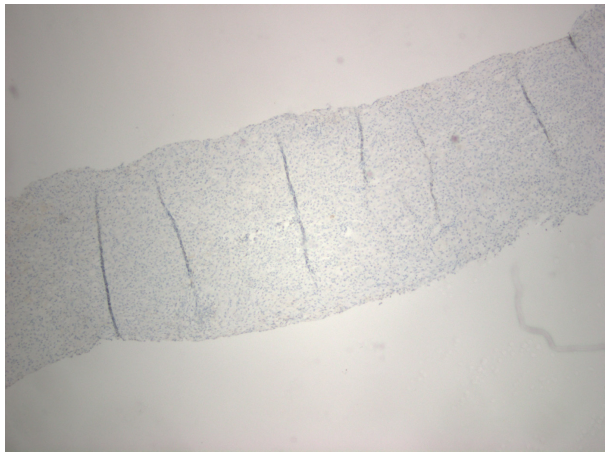
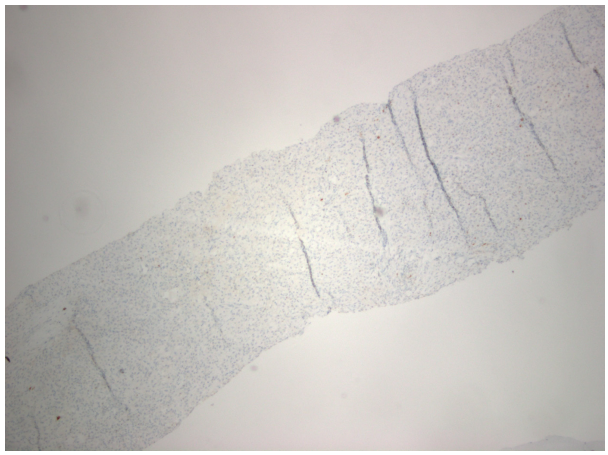


Figure 1f . negative MUM1 stain



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1 Martinez AP, Fritchie KJ, Weiss SW, et al. Histiocyte-rich rhabdomyoblastic tumor: rhabdomyosarcoma, rhabdomyoma, or rhabdomyoblastic tumor of uncertain malignant potential? A histologically distinctive rhabdomyoblastic tumor in search of a place in the classification of skeletal muscle neoplasms. Mod Pathol. 2019;32(3):446-457. doi:10.1038/s41379-018-0145-0