Malignant Myopericytoma in a Military Member

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Disclosures

Dr. Hall holds a minority equity stake in Veradermics Inc.

We have no other relevant disclosures.

Case Presentation

A 29-year-old active-duty military male in basic training presented to his primary care clinic with a two week history of a tender lesion on his right thigh. The patient reported trauma as a possible inciting event. Physical exam revealed an isolated 15mm erythematous, firm, tender, domed nodule on the right lateral thigh. An infectious abscess was suspected and incision and drainage was attempted. Upon incision, an adipose-like tumor was discovered and the procedure was aborted. Dermatology was consulted for evaluation of a possible lipoma. At presentation to the dermatology clinic the above lesion was identified with an overlying 1cm incision with hemorrhagic crust. A shave biopsy was performed which revealed an underlying encapsulated adipose-like tumor. The procedure was then converted to an excision and the residual mass was easily dissected and removed.

Histology

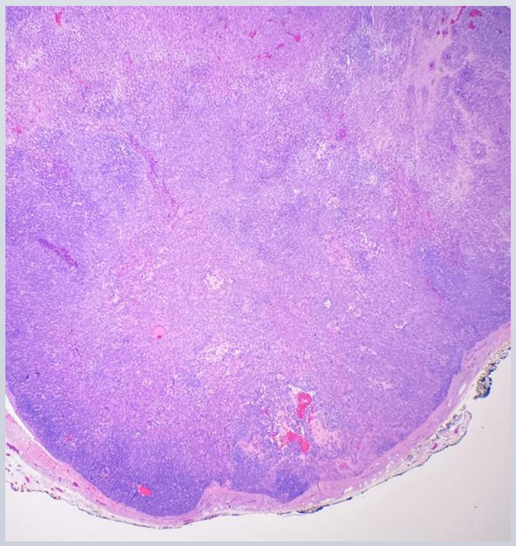

Figure 1: H&E 40x. There is a highly cellular tumor with concentric growth around vascular channels.

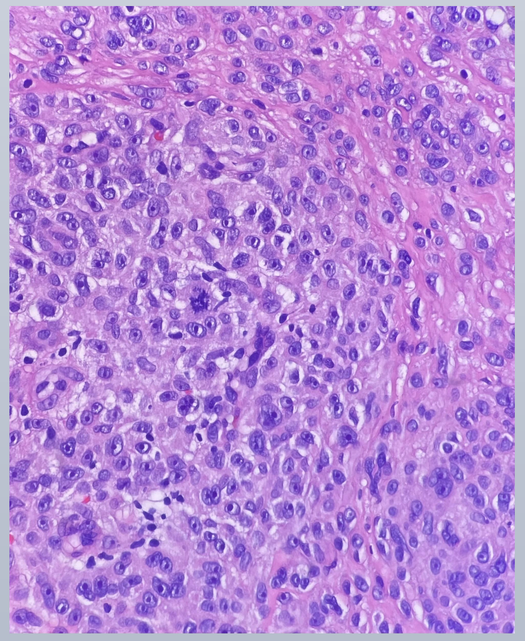

Figure 2: H&E 200x. The tumor demonstrates eosinophilic to amphophilic cytoplasm, irregular vesicular nuclei, prominent nucleoli, and multifiocally scattered mitoses.

Histology (cont.)

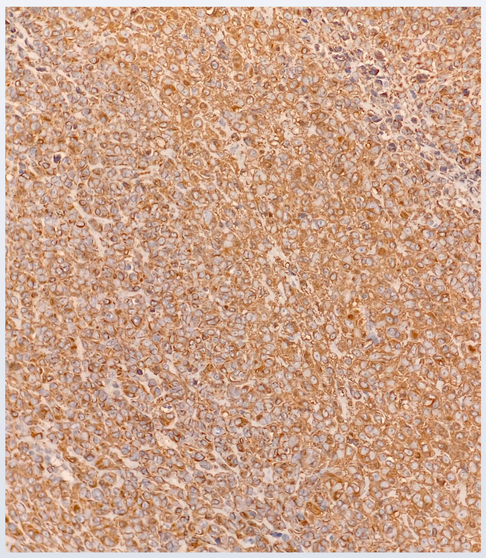

Figure 3: Strong positivity with SMA.

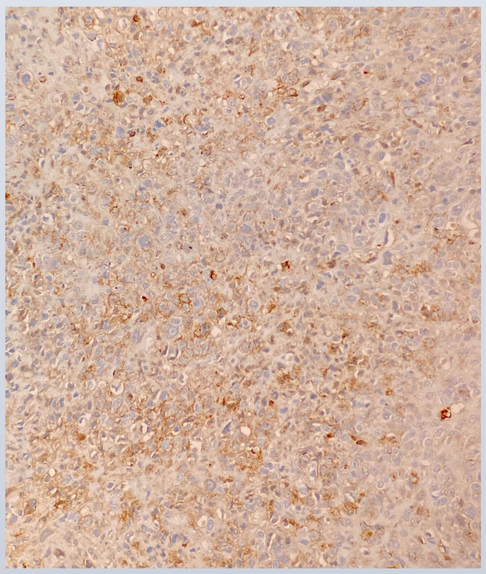

Figure 3: Multifocal and weak staining with EMA.

Clinical Presentation

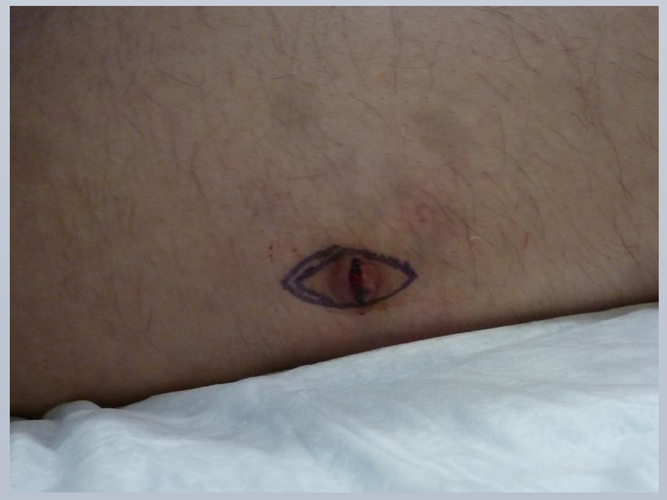

Figure 5: Erythematous nodule with overlying incision with hemorrhagic crust. The original surgical plan is outlined.

Histology

The histopathologic features demonstrated a highly cellular neoplasm with irregular vesicular nuclei, prominent nucleoli, and multifocally scattered mitoses. The cells demonstrated a palely eosinophilic to amphophilic cytoplasm with concentric growth around vascular channels. Staining of the lesion revealed multifocal weak staining for EMA and strong positivity for SMA, with negative staining for pan-keratin, S100, SOX10, ALK, CD30, CD34, CAM5.2, and desmin. The architecture and staining pattern of the lesion was most consistent with a diagnosis of malignant myopericytoma.

Discussion

Malignant myopericytoma is an exceedingly rare diagnosis with approximately 10 cases now reported in the English literature since first reported by McMenamin and Fletcher. [1] Lesions have typically been described as aggressive and rapidly growing masses involving the proximal extremities but have also been noted in the mediastinum and heart. [2] The reported cases have been predominantly male. [3] The tumors have been highly aggressive with a predilection for metastasis and recurrence. [1, 2, 3]

There is no definitive treatment for malignant myopericytoma. Reported treatments have included wide local excision with or without adjuvant chemotherapy and/or radiation. Chemotherapy and radiation have not demonstrated a clear benefit but may be considered. [4] A complete workup for staging and individualized treatment is recommended. [3]

Our patient received a PET CT for evaluation of possible metastases. A non-specific area of enhancement was noted in the right buttock without definitive lesion and biopsy of the area was recommended. Definitive excision of the primary tumor was discussed with Dermatology and Orthopedic Oncology with plan for a wide local excision. Evaluation by Medical Oncology and Radiation Oncology was planned.

Following his diagnosis and before definitive treatment could be initiated, the patient returned to his hometown to pursue treatment in a different health system. His records were transferred and he provided confirmation that he established care with his hometown healthcare facility. Following his transfer, further follow-up was lost. His ultimate treatment and ongoing health status remain unknown.

References

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2. Chen W, Han L, Pang H, Duan L, Zhao Z. Primary malignant myopericytoma with cancer cachexia: Report of the first case and review of literature. Medicine (Baltimore). 2017 Dec;96(49):e9064. doi: 10.1097/MD.0000000000009064. PMID: 29245316; PMCID: PMC5728931.

3. Patrick A, Soares-de-Almeida L, Heinz K. Malignant Myopericytoma: Report of a New Case and Review of the Literature. Am J Dermatopathol. 2016 Apr;38(4):307-11. doi: 10.1097/DAD.0000000000000463. PMID: 26863056.

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