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Malignant Myopericytoma of the Right Foot: A Case Report

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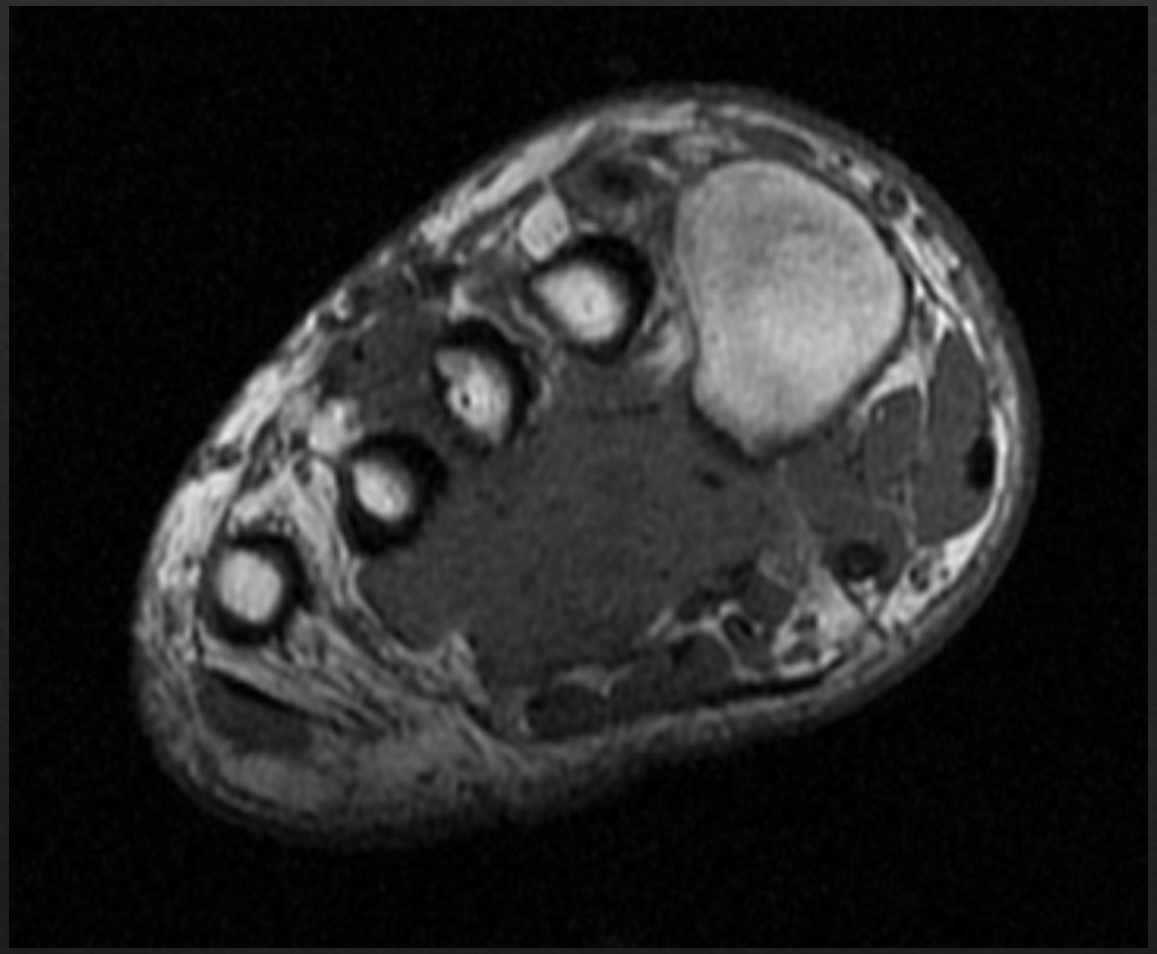
Case Background

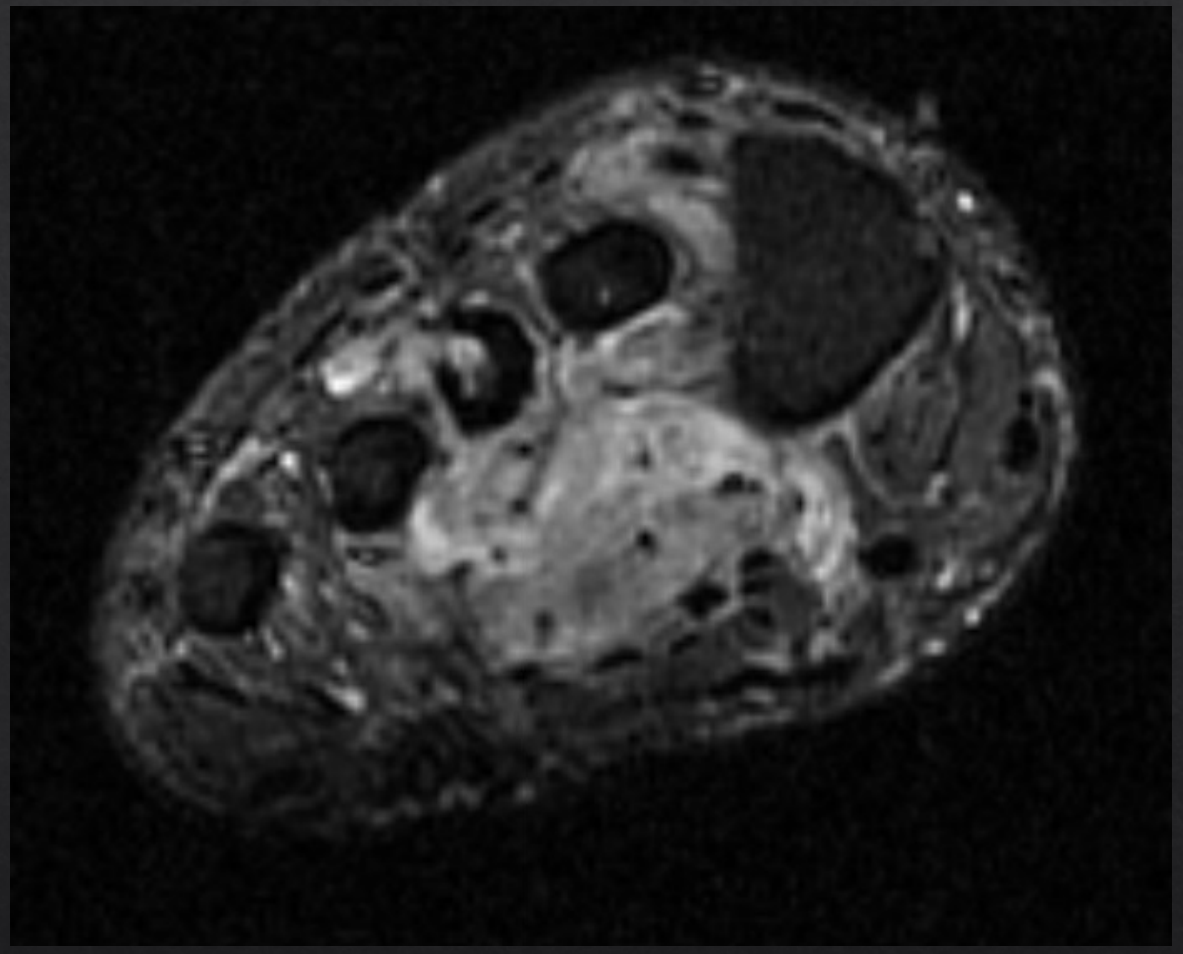
- 70-year-old male with PMHx of HTN, prostate cancer s/p prostatectomy 9 years ago, and bilateral bunionectomy
- Presents to outpatient orthopedic clinic with complaints of worsening right foot pain for the past 4-5 months
- Worse w/ activity but can occur anytime during the day
- Physical exam revealed small, poorly circumscribed mass on plantar aspect of right foot
- XR obtained with no acute findings
- MRI w/ Contrast of R foot obtained

Initial Imaging Findings

- Large infiltrating mass at the plantar aspect of the foot measuring approximately 9 x 4 x 3 cm
- Heterogeneously increased T2 and decreased T1 signal
- Immediately deep to metatarsals, abutting flexor digitorum brevis muscles and tendons
- Dorsal extension of the mass between 1st, 2nd, 3rd, and 4th metatarsals
- Associated cortical bone loss at the dorsal/lateral aspect of the 3rd metatarsal
  - *Indicating mass invasion of 3rd metatarsal*
- Benign etiologies – aggressive fibromatosis and giant cell tumor of tendon sheath
- Malignant etiologies – fibrosarcoma, chondrosarcoma, or less likely synovial cell sarcoma



Cor T1



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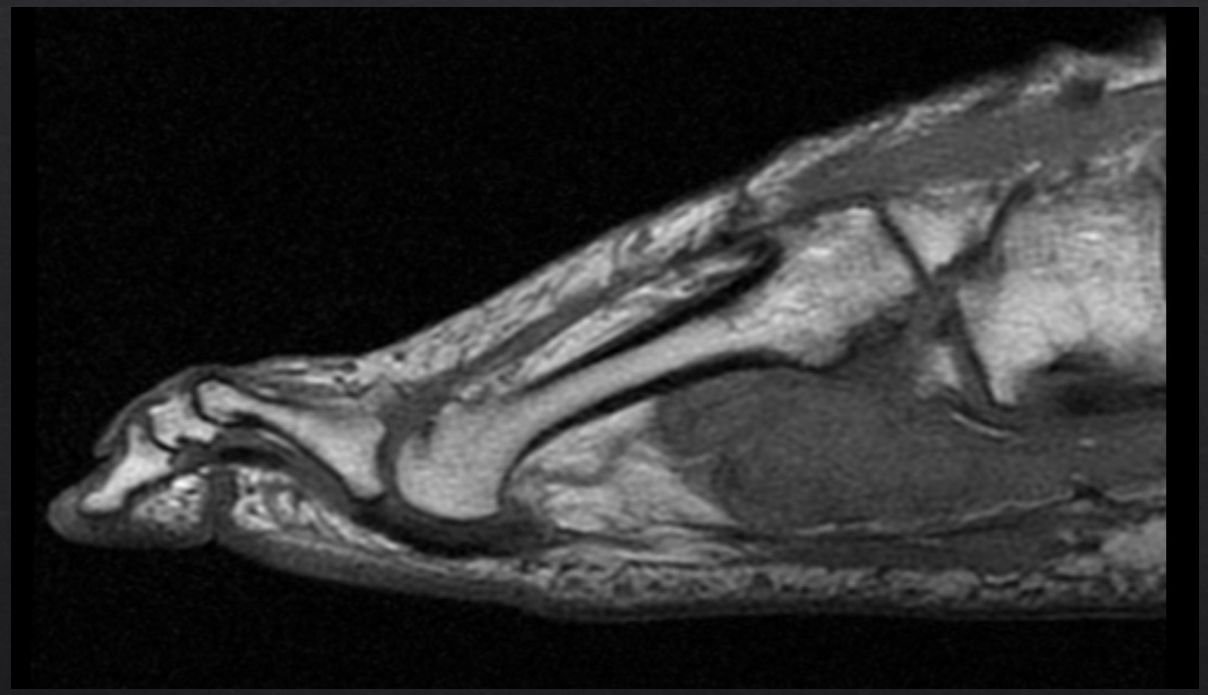
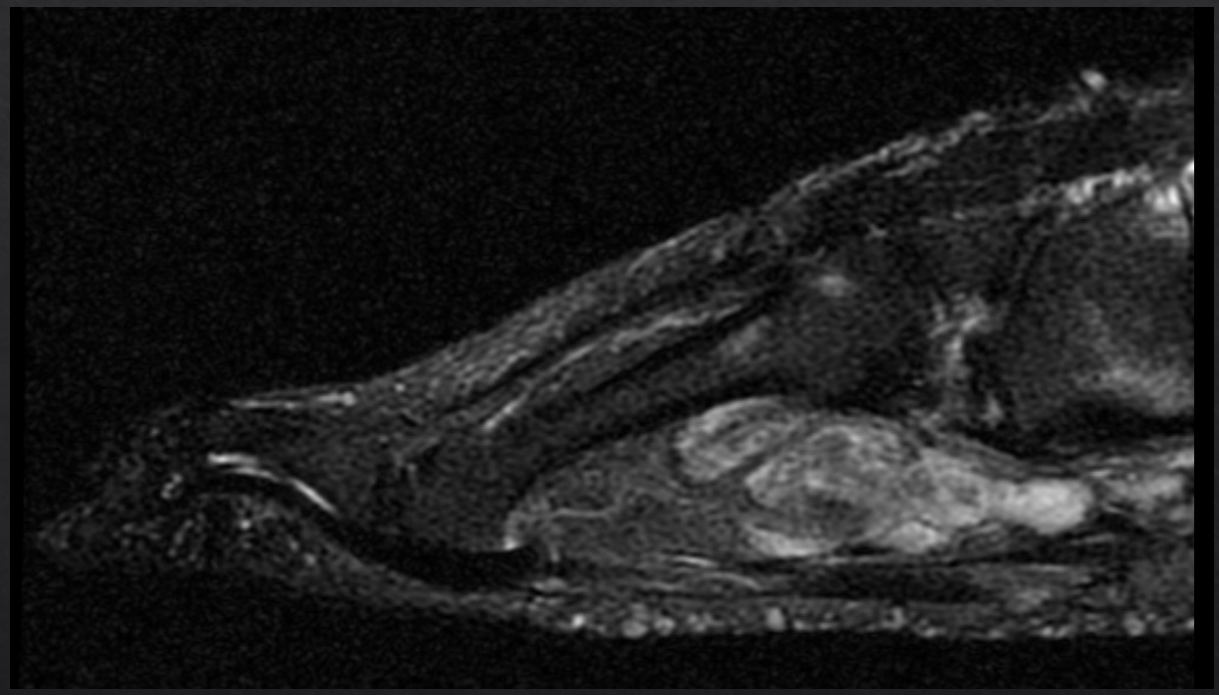


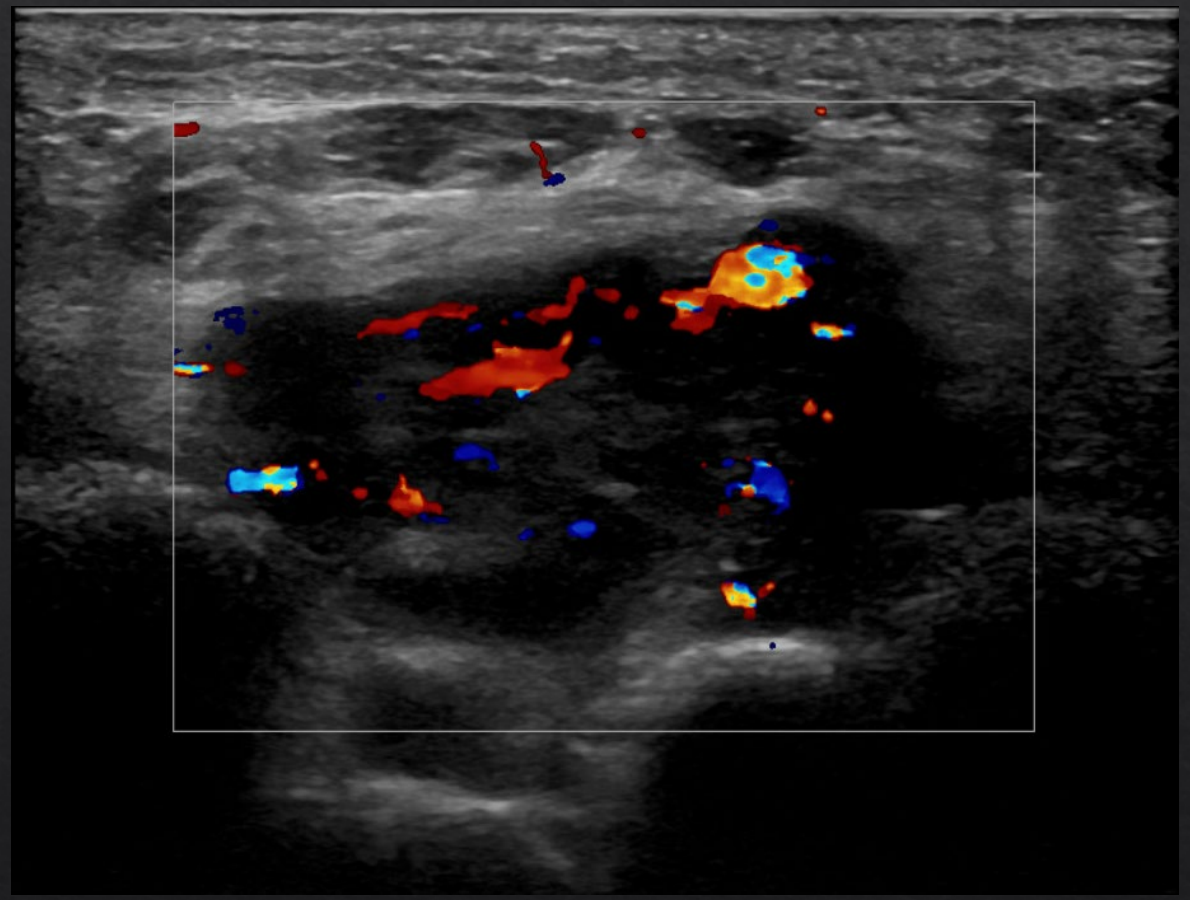
Sag T1



Sag T2

Referral with U/S Biopsy

- Patient referred from outside orthopedic clinic to SHMC for appointment with orthopedic oncologist
- U/S guided biopsy ordered after referral visit
- U/S biopsy results from SHMC
  - Atypical spindle cell lesion → sent to Cleveland Clinic for 2nd opinion
    - Read as "atypical myopericytic neoplasm"



Open Biopsy

- Wanted better characterization compared to U/S biopsy
- Cleveland Clinic – "atypical myopericytic neoplasm"
  - Spindled cells, hemangiopericytic-like vascular pattern with areas of myofibroma nodules and muscular walled vessels, rare mitotic activity. "Presence of cytologic and nuclear pleomorphism raises concern for "malignant" or more aggressive form… best diagnosis is atypical and complete excision is recommended"
- Patient presented at tumor board at SHMC
- Decision for 2nd opinion from Harvard

2nd Opinion on Open Biopsy

- Highlights from Dr. Chris Fletcher's team at Harvard
  - Spindle cell neoplasm with perivascular distribution
  - Neoplastic cells form part of vessel walls
  - Substantial subset w/ nuclear atypia beyond expected in benign myopericytic neoplasm
  - (+) for SMA and focal (+) caldesmon
  - **"Given the degree of cytologic and nuclear atypia, I believe that we have no alternative but to regard this as a malignant myopericytoma."**

Myopericytomas; What are they?

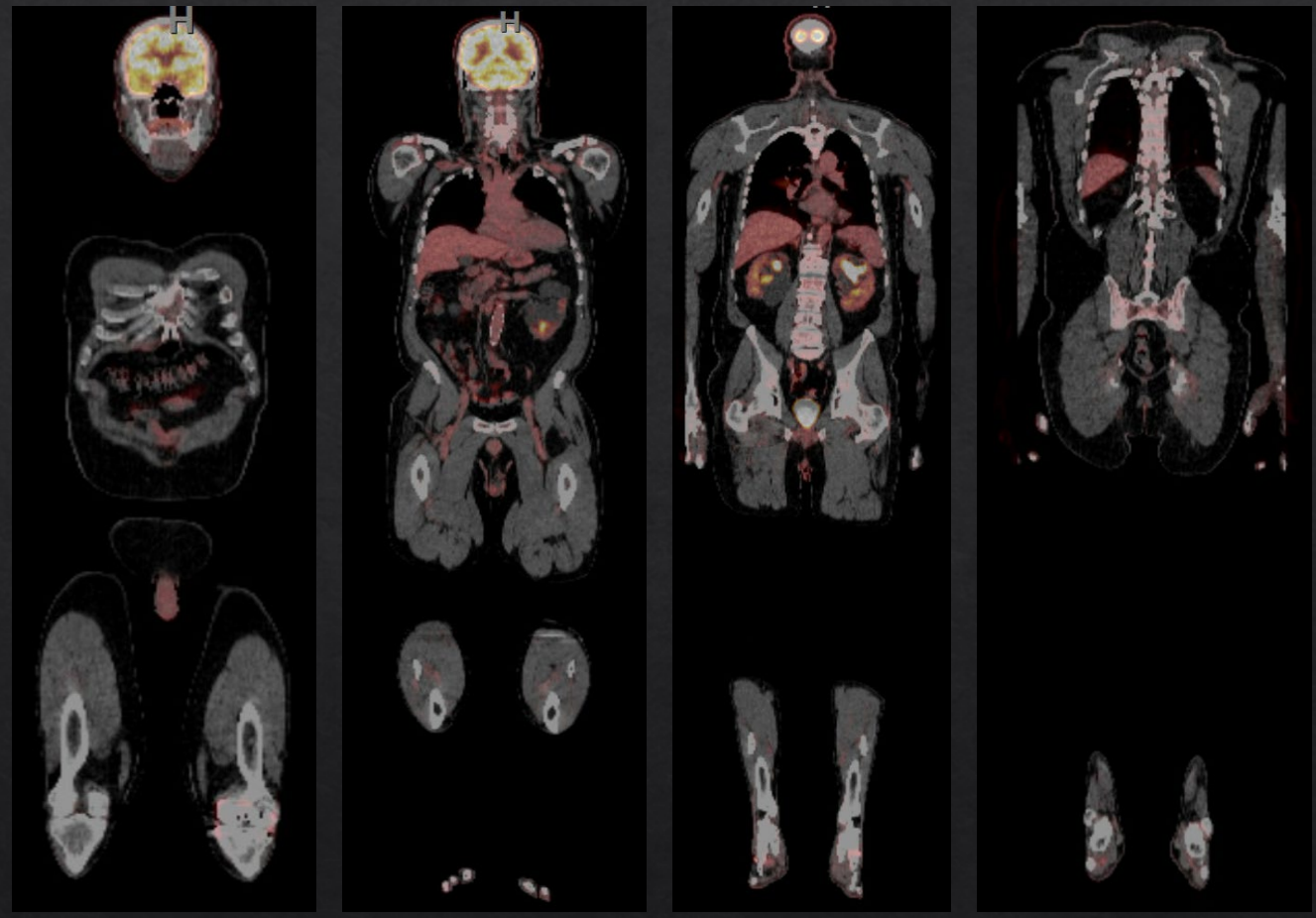
- Rare benign soft tissue neoplasms that originate from perivascular smooth muscle-like myoid cells[1]
  - First designated its own classification in 2002 by WHO[2]
- Benign myopericytomas typically present as a solitary, well-demarcated, and slow growing painful small mass over several years[1]
- Most frequent location is in the extremities often in subcutaneous tissues[3]
- Affects all ages, but peaks >50 yrs with slight male predominance[4]
- Treatment is with simple excision

Malignant Myopericytomas

- Extremely rare, aggressive form of myopericytoma with an estimated 15 cases reported in the literature as of July 2021[5]
- Defined with deeply infiltrative growth, marked atypia, increased mitotic index and high cellularity with necrosis[2]
  - Often with (+) SMA stains[7]
- Will typically see marked concentric growth around vessels
- Often mimic more aggressive lesions such as sarcomas and has similar histologic findings as other perivascular tumors[7]
- Typically very aggressive lesions that can metastasize to various sites[5, 6]
  - Reported cases of brain, skeletal, liver, lung
  - Case series where 4 patients developed metastases and 3 died within 1 year of development[7]
- Tx involves extensive surgery with adjuvant therapy due to their aggressive course, but there is no standardized treatment[4, 8]

Back to Our Patient

- Recommended amputation of right foot rather than resection given anatomic location
- Patient requested second opinion with radiation oncology
  - Did not believe radiation or chemotherapy would provide better quality of life than amputation
- Elected for conservative management with serial MRI and PET scans
- Most recent MRI one month ago with small interval growth in mass but no further infiltration in metatarsals
- Follow up PET CT with no metastasis x2 (~1.5 yrs since diagnosis)



Why This Case is Important

- Not many case reports with in-depth findings regarding specific MRI sequences
- Keep malignant myopericytomas in the differential with quickly progressing, painful soft tissue lesions in the extremities
- Highlights importance of histopathologic diagnosis as well as open biopsies

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Questions?