Undifferentiated Pleomorphic Sarcoma of the Ankle: Rare Case Report and Literature Review

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Statement of Purpose

A rare high grade undifferentiated pleomorphic sarcoma of the ankle which was discovered during an elective outpatient ankle soft tissue mass excision surgery. Complete sarcoma eradication was achieved after a wide surgical excision and split thickness skin graft, with no local recurrence in a 22-month surveillance follow up.

Literature Review

Undifferentiated pleomorphic sarcoma (UPS) formerly known as malignant fibrous histocytoma is a rare soft tissue malignant sarcoma of uncertain origin. UPS presents approximately 50-70 years of age, although it can appear at any age with a slight male predominance. UPS typically presents as a non-painful soft tissue mass and commonly occurs at the lower extremity thigh, upper extremity and retroperitoneum. Unintentional weight loss and fatigue is not typical. Diagnostic work up includes MRI imaging with contrast and biopsy in order to rule out other malignant neoplasms. If sarcoma is discovered, sarcoma staging is warranted with an initial chest CT, in order to guide multimodal treatment incorporating chemotherapy, radiation, and surgery. If diagnosed early and without metastasis, UPS can be completely eradicated. Prognostic factors for mortality rates are correlated with tumor grade, depth, size, metastasis, age, histologic subtype. UPS local recurrence is 20-30%, but recurrence is lowest in the lower extremity and directly related to negative margins with surgical excision. The five year UPS survival rate is 65-70% with about 35% UPS metastases. There is scant ankle UPS literature due to its rarity with only one case report of ankle UPS which underwent a proximal limb amputation. We report an ankle UPS with a favorable limb sparing outcome.

Clinical Presentation

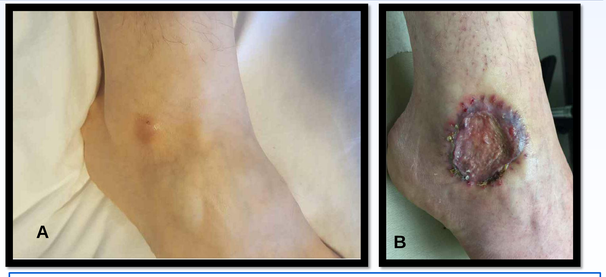

Figure 1. A: Ankle soft tissue mass presentation. **B:** 3 week status post split thickness graft after wide surgical excision with negative margins.

Magnetic Resonance Imaging

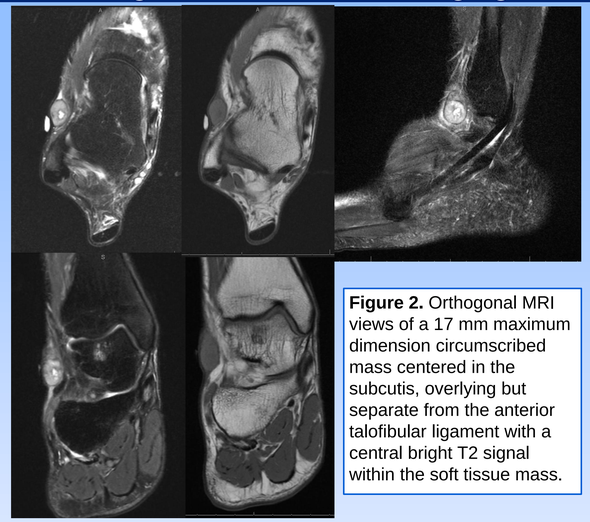

Figure 2. Orthogonal MRI views of a 17 mm maximum dimension circumscribed mass centered in the subcutis, overlying but separate from the anterior talofibular ligament with a central bright T2 signal within the soft tissue mass.

Histopathology

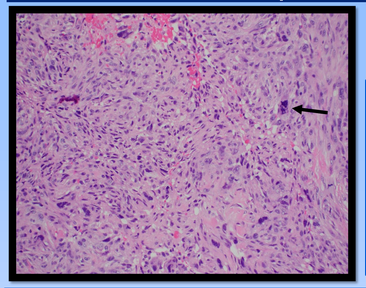

Figure 3. Histologic specimen of the excised ankle soft tissue mass demonstrating prominent pleomorphism with an atypical mitosis viewed under 100x magnification with H&E stain

Tumor Pathology Report:

- H&E sections demonstrate a pleomorphic and spindle cell neoplasm with scan seams of collagenous stroma.
- The tumor cells grow haphazardly and in a storiform arrangement indistinct cell borders and a high degree of nuclear pleomorphism including bizarre multinucleated cells with marked hyperchromasia.
- Focal foam cells are present.
- Mitoses are readily identified (8 mf/10 hpf) including atypical forms.
- No obvious line of differentiation is noted on H&E.

Intra-operative Specimen

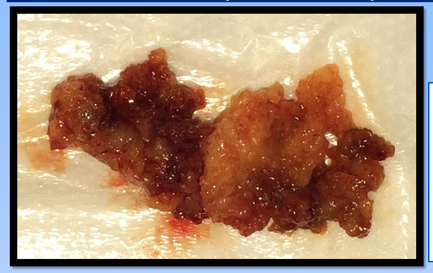

Figure 4. Intraoperative photo of the 2x2cm ankle soft tissue mass excised and sent to pathology with rusty brown color with effervescence.

Case Report

Forty three year old Greek male presented with an eight-month history of atraumatic right anterolateral ankle soft tissue mass which was enlarging and became symptomatic for the past three months due to shoe gear discomfort. The firm, well circumscribed and semi-mobile soft tissue mass was aspirated in clinic with only minimal sanguineous yield. An ankle MRI revealed a 15x10x17mm well circumscribed superficial soft tissue mass with heterogeneously increase T2 signal and a central small portion of a bright T2 fluid signal. There was mild peripheral enhancement surrounding the lesion. The lesion was also separate and superficial to the lateral ankle ligaments. Radiology determined the lesion as a cyst or lipoma.

After discussion of risks, benefits and alternatives, he elected for a right ankle soft tissue mass excision given persistent shoe gear irritation. Intraoperatively, the mass appeared rusty brown color that was not adherent to the deeper tissues or structures. The Kaiser Permanente (KP) pathology report revealed high grade undifferentiated pleomorphic sarcoma with positive margins. University of California San Francisco (UCSF) external pathology consultation confirmed diagnosis and recommended complete excision with negative margins and a metastatic work up. Chest computer tomography (CT) revealed an indeterminate solitary 2mm of the right lower lobe nodule which was determined as low risk due to nodule size being less than 4mm with a plan for serial surveillance. The KP Musculoskeletal Surgical Oncologist did not recommend any radiation or chemotherapy given the ankle tumor was less than 5 cm and superficial.

He underwent a 2.0 cm wide margin excision with multiple intraoperative frozen sections that confirmed negative margins. Negative pressure wound therapy was initiated until his wound matured with granulation tissue. A month later, he underwent a split thickness skin graft with Plastic surgery. He healed his anterolateral ankle wound uneventfully. He did not have any signs of local recurrence or interval lung nodule changes with repeat ankle MRIs and alternating chest CTs and chest XRs, every 3 months for a follow up of 22 months.

Discussion

- To our knowledge, this is the first high grade undifferentiated pleomorphic sarcoma of the ankle with a favorable outcome with eradication of the sarcoma and preservation of the foot and limb. There was no metastases or local recurrence in a 2 year follow up.
- Undifferentiated pleomorphic sarcoma is a rare soft tissue sarcoma that can present as a common soft tissue mass of the foot and ankle. Clinical aspiration attempts and advanced imaging may still not identify soft tissue lesion. Unplanned surgical excision of abnormal soft tissues are important to send to pathology in order to rule out malignancy and to discuss with musculoskeletal surgical oncologists.
- Long standing mass with a pre-op MRI with some enhancement warrants a needle biopsy or intra-operative frozen section to rule out sarcoma and need for wide resection. Thus, UPS early detection and diagnosis can lead to favorable outcomes in terms of sarcoma staging and need for metastatic work up, radiation, chemotherapy and surgical excision.

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Acknowledgements

We thank the KP and UCSF pathology departments for their integral role for diagnosis and collaboration.