Undifferentiated Pleomorphic Sarcoma of the Forefoot

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Purpose

To introduce a unique clinical presentation and discuss the multidisciplinary team approach, identification of pathology, and definitive surgical treatment while providing a literature review on practice guidelines for undifferentiated pleomorphic sarcoma.

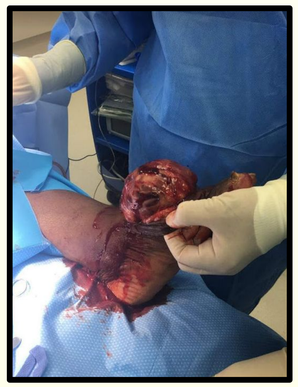
METHODS

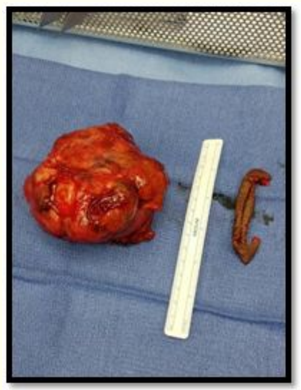
A fifty-seven (57) year-old Nigerian male with a PMH notable for HTN, obesity, and a 6.4-cm aortic aneurysm s/p open-repair in July 2018 presented with a substantial mass on the dorsal-left foot. The patient arrived at the clinic with an inability to wear regular shoe-gear; however, the utilization of open-toed shoes and sandals allowed the individual to make accommodations for this mass. A physical exam revealed a grapefruit-sized growth that did not move freely, while mild tenderness was exhibited during palpation. The patient's foot displayed the neurovascular status intact with a musculoskeletal exam revealing no attachment to adjacent tendons or decrease in ROM throughout adjacent joints. An MRI of the foot revealed an oblong, multilobulated but well-circumscribed mass that traversed the mid-foot and first metatarsal, and measured 9.0-cm in length, 5.3-cm in craniocaudal dimension, and 7- cm in transverse dimension. After these initial findings, the patient was referred to oncology where the tumor was graded as T2bN0M0 (Grade IIb) and surgical intervention was recommended for removal of the mass. Surgical-wide excision was performed utilizing a combination of sharp and blunt dissection to detach the mass from its underlying soft-tissue attachments. Stalking communication was noted at the 1st MTPJ capsule where clip ligation and transection were performed. No other communication was appreciated. Microscopic description illustrated fascicular, solid, myxoid, and storiform patterns; moreover, tumor cells were pleomorphic and multinucleated. Mitotic activity was present with abnormal mitosis, as were many extensive areas of necrosis and inflammation. Tumor cells were observed within 1-mm of margin and extended into outer adipose tissue of the clearance margin, thus suggesting that a well-circumscribed neoplasm existed. The diagnosis on the surgical pathology report ascertained that the mass was an undifferentiated pleomorphic sarcoma. Postoperatively, the patient followed-up with oncology where a PET and sarcoma mutation panel were ordered to rule out systemic metastasis. Unfortunately, the patient did not return for contiguous follow-up and sought a second opinion with an associate oncologist at a neighboring health system. Approximately one year after the initiatory surgical excision, the patient returned to the primary surgeons office and was found to be healthy without further metastasis or recurrence of the tumor. The recommendation was made that the patient should schedule yearly follow-up appointments with both his oncologist and surgeon.

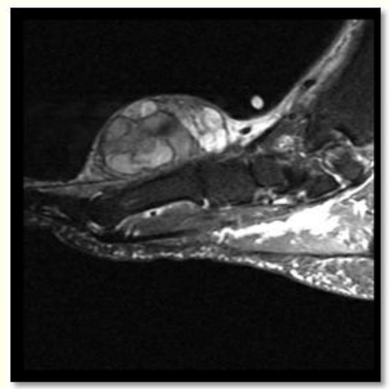
DISCUSSION

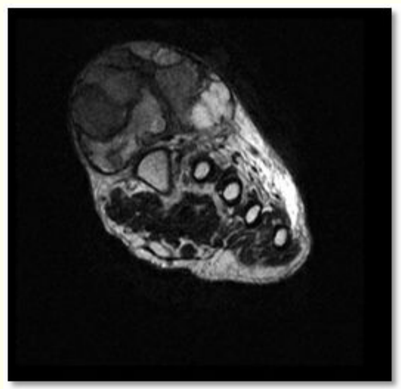
Undifferentiated pleomorphic sarcomas (UPS) account for approximately one-third of all high-grade extremity sarcomas (1). Surgical resection and adjuvant therapies such as radiation or chemotherapy are the preferred treatment for UPS, as well as other comparable high-grade extremity soft-tissue sarcomas (1). Of particular note is the uncertainty regarding true margin clearance for these tumors since they are often diagnosed after surgical excision. Unplanned excision is the gross removal of tissue without preoperative staging or the absence of professional consideration prior to removing an envelope of normal tissue around the tumor (2). Unplanned excision, even when combined with radiation therapy or chemotherapy, is inadequate for the treatment of soft-tissue sarcoma. Re-excision is necessary to lower the incidence of local recurrence (3). The rate of unplanned excision is about 40% for soft-tissue sarcoma of the extremities (2-4). No difference was found between CT and MRI in the accuracy of determining tumor involvement with adjacent soft-tissue structures, although an MRI is considered superior by medical specialists in its ability to delineate anatomy (5). Frequency and metastatic progression patterns for high-grade tumors were reviewed by Massachusetts General Hospital-Boston, MA. Their research outcomes confirmed that tumors measuring 5-cm to 10-cm had a 38% chance of distant metastasis (6). Soft-tissue sarcomas are best managed by wide surgical resection with adjunctive radiation. With this management, the local recurrence rate is reported to be below 10% (7). Unplanned-positive margin after surgical resection has been found to increase local recurrence and reduce patient survival rates (8-10). One study observed a recurrence rate of 32% for patients who underwent unplanned excision (11). In contrast, excision with a planned-positive margin in the context of radiation therapy was shown to yield a very low local recurrence rate of 3.6% (11). Existing literature analyzing soft-tissue sarcomas of the foot and ankle is deficient; Only two published studies were found to exclusively examine the foot and ankle within their research (13,14). In summary, the preceding assertions suggest the following: early diagnosis, multi-specialty treatment, and precise follow-up may reduce mortality rates in individuals with diagnosed soft-tissue sarcoma of the lower extremity.

Figures









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