Leiomyosarcoma of Vascular Origin in The Foot

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Introduction

Leiomyosarcoma (LMS) is a rare, malignant tumor with prevalence of less than one in every 100,000 malignant tumors[1]. Three types of LMS have been described based on their origin. The most common LMS is soft tissue, followed by cutaneous, and the rarest type is vascular[2, 3]. LMS and its benign variant, Leiomyoma, are often found to affect the uterus and abdominal cavity [2]. LMS in the foot and ankle is extremely rare with few isolated case reports in literature. Since 1936, only 21 cases have been reported in the foot and ankle[2, 4]. This case study reports a 75 y.o. male with primary leiomyosarcoma of vascular origin (LMSv) in the hind foot with successful treatment, and a 26 month follow up.

Case Study

A 75 year-old male patient presented to clinic with the chief complaint of a painful mass to his right heel. The patient presented with a nodular mass to the anteriormedial aspect of the right calcaneal region which has been ongoing for eight months. Patient reported constant pain which he rated 2/10. The mass was semi-mobile and adhered to skin and subcutaneous tissue. Clinically, the mass was diagnosed as a ganglion cyst on initial visit. However, patient returned one month later with complaint of progressive enlargement and increasing pain. MRI was ordered and revealed a non-specific soft tissue mass with differential diagnosis of "soft tissue neoplasm including a soft tissue sarcoma, a hemangioma, neurofibroma, a tumor of the skin appendage or somewhat favored a low grade vascular lesion with two crossing vessels. Excisional biopsy is recommended."

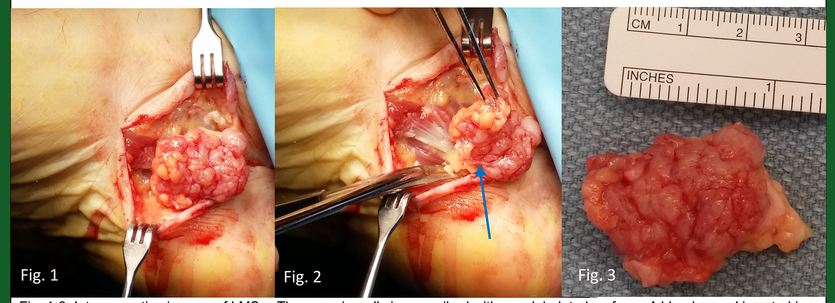
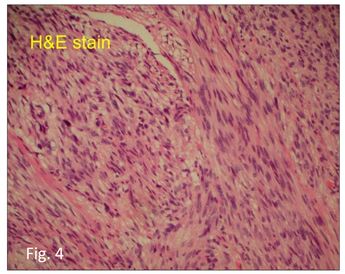


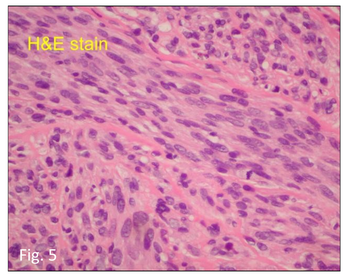
Fig. 1-3: Intra operative images of LMSv. The mass is well circumscribed with very lobulated surface. A blood vessel is noted in fig. 2 at blue arrow. The soft tissue mass measured 2.5x2.2x1.0cm.

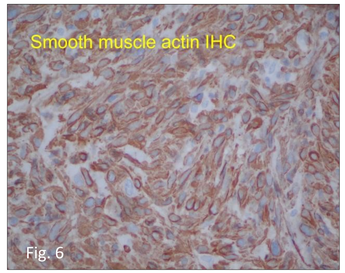
During excisional biopsy, the mass was noted to be well circumscribed with lobulated surface resembling that of a brain (fig. 1-3). The mass was found abutting the superior aspect of the abductor hallucis muscle without intramuscular invasion. Two small blood vessels within the mass was also noted (fig. 2). The mass was successfully excised in total and sent for pathological evaluation with samples of the abductor hallucis muscle for clean margins. Histological studies revealed intercrossing bundles of fascicles at low power with numerous spindle shaped cells with cigar-shaped nuclei at high power (fig. 4-5). Ancillary stains were positive for smooth muscle actin antigens and Ki-67 proliferation index was increased (fig. 6-7).

Case Study Continue

Pathology reported the 2.5x2.2x1.0 cm soft tissue mass is a LMSv Grade 1 (FNCLCC grading) with 5 mitotic figures per 10 high powered fields and pathologic staging pT1NXMX . The abductor hallucis muscle sample was found to be unremarkable. Oncology and plastic surgery were immediately consulted.







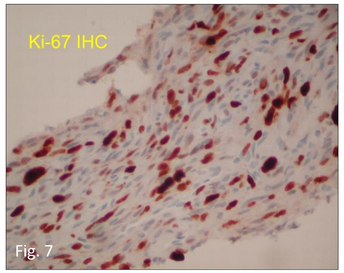


Figure 4 & 5: Numerous spindle-shaped cells with evident of cigar-shaped nuclei at higher power.

Figures 6 & 7: Ancillary stains showing presence of smooth muscle actin antigens, consistent with smooth muscle derivation and increased Ki-67 index indicating malignancy nature.

Computer tomography (CT) of the chest with contrast was obtained with unremarkable results. Following through evaluation by the oncology team, the tumor was classified as Stage 1A (T1b N0 M0) LMS. A plastic surgeon along with the oncology team formulated a treatment plan, and the patient underwent neoadjuvant external beam irradiation using intensity-modulated radiotherapy (IMRT) technique with appropriate margins, 5 fields, 6mV photons, 50 Gy at 2 Gy per fraction in 25 fractions with a 1cc customized bolus. Following completion of radiation treatment, wide excision of the lesion site was performed and margins were determined clear via pathology. The plastic surgeon reconstructed the wide excision site using an autologous split thickness skin graft harvested from patient's ipsilateral thigh. At one year follow up, patient had negative chest CT and body PET-CT scans, and excisional site fully healed without any complication (fig. 8). At two years follow up, patient continue to do well, and without signs of metastasis. He was able to return to everyday normal activity level and is happy with outcome.

Discussion

LMS are malignant soft tissue tumors that can arise from any smooth muscle in the body, however, most are commonly found in the uterus and abdominal cavity[2]. LMS can be further divided into three types: soft-tissue, cutaneous, and vascular[3,6]. LMSv is the rarest type of LMS, making up less than 2% of all LMS[1,5-7]. The majority of LMSv occurring in the extremity are found proximal to the knee and in large vessels such as the Inferior vena cava and Great saphenous vein[6,7]. Wide excision is treatment of choice. However, with involvement of large vessels, excision can range from difficult to impossible; making the prognosis of LMSv very poor. Fortunately for our patient, the LMSv was found in the foot involving small end vessels which are amenable to wide excision and radiation treatment.

LMS in the foot, though extremely rare, can clinically mimic other soft tissue masses such as ganglion cyst, lipoma, fibroma, etc. Further advanced imaging and/or diagnostic tests should be performed if suspicions of possible malignancy arise. Stock et al. described sonographic features of LMS of the foot, however, they concluded the features remain non-specific and differentiation from other benign or malignant neoplasms was not possible based solely on sonographic features[8]. Magnetic Resonance Imaging (MRI) is the preferred advanced imaging modality for sarcomas affecting the extremity such as the one in our case[9-11]. Despite being non-specific, MRI can differentiate muscle groups, bone, vascular structures, tendons, ligaments, and involvement of the lesion(s) with surrounding tissues. In addition, MRI can help differentiate benign lesions such as lipomas, hemangiomas, schwannomas, neurofibromas, and intramuscular myxomas from their malignant variants[10, 11].

A differential diagnosis that includes malignancy should drastically change the treatment plan and dictates speedy consultation of appropriate specialties involvement. In our case, the differential diagnosis via MRI included soft tissue sarcoma with excisional biopsy recommended. This resulted in a definitive diagnosis of LMSv and triggered consultations of appropriate specialties such as oncology and plastic surgery. Our timely and multidisciplinary approach to this particular rare case of primary LMSv of the foot afforded us a favorable outcome and a grateful patient.

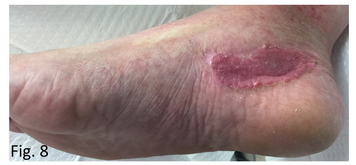


Figure 8: Healed surgical site at one-year follow up. Measuring 10x2cm.

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