Leiomysarcoma – A Case Report of a Rare Pedal Soft Tissue Mass Malignancy

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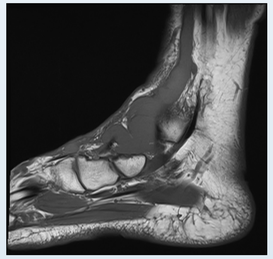
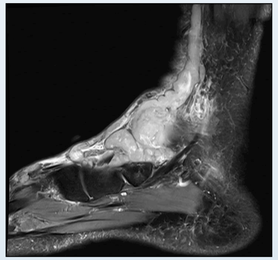
Introduction and Literature Review

Bibbo et al, in 2011, reviewed the vascular leiomyosarcoma and detailed its incidence as a rare soft tissue sarcoma, discussing the epidemiology, evaluation, and current treatment options [1]. They further went on to report a case of vascular leiomyosarcoma at the Greater Saphenous Vein (GSV) ankle distribution. A large 7cm specimen was identified and excised. Similarly, we are reporting on a case of leiomyosarcoma at the level of the GSV ankle distribution. Soft tissue sarcomas (STS) are uncommon malignant tumors that arise from connective tissues. Leiomyosarcomas are an extremely rare sub group of STS, accounting for less than 10% of STS. The most common vessel affected in the lower extremity is the GSV, with the vast majority occurring proximal to the knee joint. Research cites between 15 – 25 leiomyosarcoma cases reported in the last century [4]. The purpose of this poster is to present a rare aggressive pedal STS, in order to bring awareness of neoadjuvant therapies and report on a case of outpatient management of neoplastic disease.

Diagnostic Image 1 & 2 ; Sagittal MRI T1 & T2 enhanced image

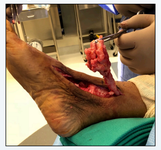




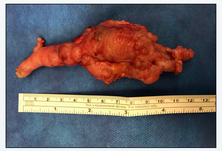
Case Presentation

A sixty-nine year old African American female with an unremarkable past medical history, and remote tobacco history presented to clinic with a growing and painful mass to the dorsal medial right foot and anterior ankle. She states the mass had arisen approximately 1 year ago. On physical exam, a tender, firm, irregularly shaped subcutaneous mass on the dorsomedial foot extending to the ankle was palpated. X-ray imaging revealed soft tissue edema dorsally in the foot with soft tissue prominence medially at the hindfoot of uncertain etiology. A Magnetic Resonance Imaging (MRI) was performed and revealed a multiseptated tubular appearing mass measuring 10cm x 5cm x 4.5cm, with exuberant enhancement. Venomalformation versus plexiform neurofibroma was suspected. The report further went on to describe a low suspicion for low grade malignancy.

Additionally, an in office aspiration biopsy was performed which showed a predominantly hypocellular specimen, composed of neutrophils and macrophages, without malignant cells. After these various modalities and diagnostic tests, the decision was made to proceed with surgical excision with biopsy.



Clinical Image 1 & 2



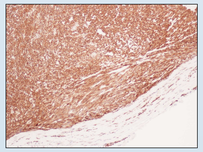
Timeline of Events

Date	Event	Details
1/12/18	1st office visit	X-Ray performed, MRI ordered, discussion of surgery
2/20/18	2nd office visit	Discussed MRI (10cm x 5cm x 4.5cm mass), office aspiration biopsy
3/13/18	3rd office visit	Biopsy reviewed (hypocellular non malignant specimen), surgery is scheduled
4/4/18	Surgery	Excision of mass, sent to pathology
4/10/18	1st post op visit	Reviewed pathology results, chest/abdominal CT ordered, MRI entire extremity ordered, referral to oncology
4/10/18 - 4/17/18	1st oncology visit	Evaluation of records, referral to University of Florida orthopedic oncology with presentation at tumor boards. Recommended repeat chest CT in 4 months, radiation therapy alone without systemic chemotherapy
4/20/18	2nd post op visit	Normal post operative healing, sutures removed
5/11/18	4th post op visit	Fully healed incision, continue oncology follow-up
10/1/18	2nd oncology visit	Repeat chest CT and extremity MRI negative, no further surgical plans, neoadjuvant therapies alone

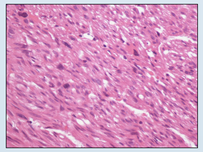
Histologic Report

Tumor Summary:
Size: 11cm x 4.4cm x 4cm
Histologic type & Grade: Leiomyosarcoma, Grade 2/3
Mitotic rate: 10 to 12 mitosis per 10 high-power fields
Necrosis: Present, approximately 15%
Margins: Involved the inked surgical margins
Lymphovascular invasion: Not identified
*Immunohistochemical studies show that the neoplastic cells are diffusely strongly positive for smooth muscle actin and desmin but negative for SOX-10, CD117 and CD34 and estrogen receptor
**This profile is consistent with a well-differentiated leiomyosarcoma of soft tissue

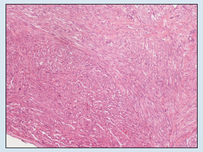
Histologic Image 1



Histologic Image 2



Histologic Image 3



Discussion

Research has shown that the recurrence rate of STS, including leiomyosarcoma, is likely 80% within the first two years of treatment. This stresses the importance of close post treatment surveillance, including periodic MRI and computed tomography (CT) [3]. The long term survivorship for STS of the foot, following surgical treatment, have five year survival rates of approximately 80% [3].

The overall goals of treating extremity STS are to minimize local recurrence, perioperative morbidity and mortality, and to maximize function and long-term survival. To achieve these goals, radiation therapy (RT) combined with wide surgical resection is recommended for most patients with high grade STS.

MRI is the most useful test for the initial local assessment of a STS [2]. The main site of distant metastatic disease is within the chest for most histologic tumor types and assessment for distant disease is carried out with chest CT. The added value of fluorodeoxyglucose-positron emission tomography (FDG-PET) or integrated PET-CT is still up for debate.

Wide surgical excision of the primary tumor without violation into tumor tissue, and with preservation of surrounding neurovascular structures is essential. Violation of the tumor is associated with a higher local failure rate even if RT is used [5]. In patients treated with combined surgery and RT, the status of the surgical margins influences the rate of recurrence and overall outcome [8].

The role of chemotherapy continues to be studied and debated. Adjuvant radiation is often used to aid with local control when it is assumed that adequate margins will be difficult to achieve. The adjunctive therapies (chemotherapy and/or RT) chosen are based upon the size and grade of the tumor, institutional expertise, experience, and physician preferences [6]. Wide local excision of STS involves excision of the sarcoma with an adequate margin of uninvolved tissue [6]. Following resection of small tumors, primary closure of the wound may be possible. Although it is rarely needed, primary amputation may be indicated for patients with large locally advanced or recurrent STS not amenable to resection or in those with severe medical comorbidities. If amputation is not accepted, RT alone is an alternative treatment for STS [6].

In this case study, the patient underwent a surgical excision which revealed a massive malignant leiomyosarcoma which did return with positive surgical margins. Immediately following excision of the sarcoma, repeat full limb MRI was ordered, CT chest was ordered, and the patient was referred to an orthopedic oncologist. The case was presented at tumor boards and a decision was made for adjuvant radiotherapy with plans for repeat MRI, and chest CT four months after the initial surgical date. Both the repeat MRI and chest CT were negative for advancement or recurrence.

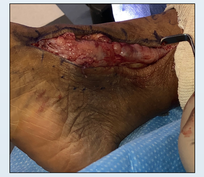
The patient was followed closely for care and monitoring of the surgical site. The patient had an uneventful postoperative course and was transitioned to full weight bearing with normal wound healing appreciated.

In the setting of positive margins post operatively in the specimen, the treating team needs to weigh the pros and cons of re-resection versus additional radiation, with an end goal of preservation of functionality. Patients with positive margins are treated with higher doses of adjuvant RT, however, the team must consider the potential negative effects of further irradiating native host tissues. Further excision to achieve negative margins may compromise neurovascular bundles or future limb salvage attempts.

Current recommendations for primary limb amputation are somewhat poorly defined. General indications include distal disease that encompasses the majority of the extremity. In this setting, often times a functional prosthetic is a better option over a risky salvage attempt in compromised local tissues or host. Therefore, amputation should not always be considered a failed outcome. Additionally, adjuvant RT is supported by oncologic treatment centers. Even if a patient did not receive any RT initially, RT has been shown to significantly improve overall outcome in patients with positive margins [7].

Conclusion

Clinical Image 3



The essential component of treatment for STS is surgical resection with negative margins, combined with a multidisciplinary approach inclusive of RT and chemotherapy. The optimal therapy regimen is highly variable and patient specific. A high level of suspicion must be maintained with atypical soft tissue mass presentation and in patients with risk factors. Modern advanced imaging, the use of adjuvant RT, and improved surgical training will significantly impact limb salvage in patients with extremities effected by STS.

References

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