



## Case Report

A SCITECHNOL JOURNAL

# A Case of Primary Cutaneous Leiomyosarcoma Managed with Mohs Surgery

Jolie Krooks<sup>1\*</sup> and Andrew Styperek<sup>2</sup>

### Abstract

Our patient presented with Primary Cutaneous Leiomyosarcoma (PCL), a rare malignant tumor of smooth muscle cells that are more commonly found in visceral structures. In contrast to tumors originating in deeper structures, which have high rates of metastasis and mortality, primary cutaneous tumors are typically only complicated by high rates of local recurrence due to inadequate excision of tumor margins. To minimize recurrence, wide local excision has been the gold standard treatment. Mohs surgery, employed in this case, is becoming a popular alternative to decrease morbidity and improve cosmetic outcome.

### Keywords

Cutaneous leiomyosarcoma; Mohs surgery; Malignant tumor

### Introduction

PCL is just one of over 100 subtypes of soft tissue sarcomas that comprise 1% of adult malignancies and are notorious for high rates of recurrence and metastasis following complete resection [1]. Leiomyosarcoma may be found anywhere in the body where there is smooth muscle. The uterus is the most common site of involvement and is implicated in about 50% of cases [2]. The non-visceral disease is even rarer and is thus less studied. Clinical course, work-up, and treatment depend on the site of involvement. The diagnosis of primary cutaneous tumors requires both clinical context and immunohistochemistry. Treatment is limited to local excision with frequent follow-up to assess for recurrence.

Our case is unique because the PCL arose out of a biopsy-proven leiomyoma. While leiomyoma is typically considered a benign tumor distinct from PCL, malignant transformation has been rarely identified by other scattered case reports [3-7].

The case highlights the rare, malignant potential of leiomyoma as well as the need for adequate tissue to correctly diagnosis the malignancy. Only a full-thickness incisional biopsy was able to identify the tumor.

### Case presentation

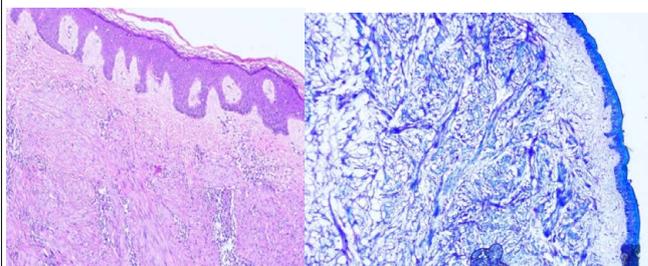
An otherwise healthy 67-year-old Caucasian male was referred to us due to a mildly painful, red lesion on the right lower extremity that had "been present for years," but that he had noted to be increasing

in tenderness and size. This lesion had previously been proven to be a leiomyoma on biopsy. He had not identified any remitting or exacerbating factors. Other than the cutaneous lesion, review of systems and physical exam were nonremarkable. The patient's evolving right distal pretibial lesion was noted to be a tender, hyperpigmented dermal nodule approximately 2.8 cm × 2.1 cm in diameter (Figure 1).

An incisional biopsy revealed malignant cells with smooth muscle differentiation with scattered mitoses and nuclear and cytologic atypia. In contrast, the leiomyoma section demonstrated a tumor limited to the dermis with minimal atypia and pleomorphism and scarce mitoses. Immunohistochemistry of the current section was strongly positive for smooth muscle actin and Ki-67 with >20% proliferation index. The tumor predominantly involved the dermis, though focal involvement of the subcutis was also noted. Lateral margins were diffusely involved (Figure 2a and 2b). PCL was diagnosed based on the patient's presentation of an evolving leiomyoma and consistent immunohistologic findings. Mohs surgery was advised due to the tumor's location. The preoperative diameter was 3.5 cm × 1.5 cm (Figure 3). The lesion was closed after two stages with negative margins. Excision reached the adipose tissue with a primary surgical defect of 6 cm × 5 cm, which was then closed by a full-thickness skin graft and advancement flap. The final surgical defect was 12 cm × 5 cm (Figure 4). The decision to defer additional treatment with radiation was made due to the absence of positive margins and suspicion for potential spread, though follow-up with an oncologist was still



**Figure 1:** Evolution of right pretibial lesion to a 2.8 cm × 2.1 cm hyperpigmented dermal nodule.



**Figure 2:** (a): Atypical spindle-shaped cells and scattered mitoses within the dermis with minimal penetration of the subcutis (H and E stain, 10X magnification); (b): Immunohistochemistry stain for smooth muscle actin and Ki-67 (10X magnification).

\*Corresponding author: Jolie Krooks, Florida Atlantic University, Charles E. Schmidt College of Medicine, Boca Raton, FL, U.S.A, Tel: 856-625-4996; E-mail: jkrooks2014@health.fau.edu

Received: December 09, 2019 Accepted: December 20, 2019 Published: January 06, 2020



**Figure 3:** Preoperative tumor 3.5 cm x 1.5 cm on the right distal pretibial region.



**Figure 4:** The final surgical defect was 12 cm x 5 cm after suture removal.

advised. There have been no signs of recurrence or metastasis on 6-month follow-up and imaging.

## Discussion

Consistent with our patient's demographic, PCL most commonly affects caucasian middle age-elderly males [8]. Patients may present with cutaneous PCL as a primary neoplasm or as a site of metastasis. Most commonly, cutaneous PCL presents as a solitary, well-circumscribed, painful nodule on the hair-bearing regions of the extremities, particularly the thigh and upper arm. Lesions rarely exceed 2 cm [9-11]. However, presentation is highly variable and non-specific; several variations from the above description have been described. Thus, the diagnosis of PCL requires a combination of clinical presentation with biopsy. Cutaneous and subcutaneous PCL must also be further distinguished with MRI, as tumors extending into deeper tissue warrant further imaging to stage and assess for metastasis.

Histologically, PCL needs to be differentiated from other spindle-cell neoplasms presenting with cutaneous findings based on clinical picture and immunohistochemistry. Differential diagnoses of cutaneous spindle-cell tumors include dermatofibroma (DF); dermatofibrosarcoma protuberans (DFSP); malignant fibrous histiocytoma; Kaposi sarcoma; angiosarcoma; spindle cell squamous cell carcinoma; and desmoplastic melanoma. The cells of PCL, Kaposi sarcoma and angiosarcoma are arranged in a fascicular pattern, while DF and DFSP cells are arranged in a whirled pattern. The atypical, infiltrative tumor cells of PCL have elongated, blunted nuclei and

homogenous eosinophilic cytoplasm [10,12]. Whereas cutaneous PCL consists of poorly defined proliferating spindle-shaped tumor cells proliferating within the stroma, subcutaneous PCL cells are typically more clearly defined and vascular [4]. Distinguishing microscopic features from leiomyoma include increased number of tumor cells with oval, hyperchromatic nuclei; pleomorphism; mitotic figures; and minimal stromal fibers between tumor cells. Vascular walls are notably thinner [13]. Immunohistochemistry is instrumental in differentiating PCL from other cutaneous spindle-cell tumors. Specifically, PCL is positive for SMA, desmin, vimentin, and h-caldesmon [12,14]. Of note, further molecular classification of PCL in itself is dependent on anatomic location and has prognostic significance; however, further research with larger sample sizes are suggested for more clinical significance [15].

The gold standard for treatment has been wide excision, which typically requires skin graft for closure to reduce recurrence (42%) [10]. Recommendations regarding the width of tumor-free margins vary from >1 cm up to 2-5 cm. Mohs surgery has become an appealing option in order to reduce recurrence and improve cosmetic outcome, though current knowledge is limited to studies with small sample sizes. Of the largest studies we noted (11 patients with 4.5-year follow-up [16] and 14 patients with 5.6-year follow-up) [9], no patients experienced recurrence. Adjuvant radiotherapy and chemotherapy is typically not warranted in patients with the cutaneous disease [17].

A full skin exam is warranted at diagnosis and with each follow-up, as the most common form of metastasis is to other dermatologic sites [9]. In patients with primary cutaneous lesions, follow-up skin exams with local ultrasound are recommended every 3 months within 3 years of excision, every 6 months after 3-5 years, with a return to standard annual exams. Further imaging is warranted in patients with deeper involvement [18].

Depth of invasion is an essential prognostic factor. The involvement of the subcutis was minimal in our patient, though follow-up with an oncologist was advised. Patients with tumors extending to subcutaneous tissue have a higher recurrence, metastasis, and/or mortality. No association with a recurrence has been noted with tumor width and mitotic index [10,14]. In contrast, local recurrence is dependent on the proper excision of clean margins rather than on tumor depth [19].

## Conclusion

We present a rare case of primary cutaneous PCL arising out of a leiomyoma. We excised the presenting tumor with Mohs and referred to oncology due to subcutaneous involvement. Our case adds to the increasing recognition that Mohs is an attractive alternative to wide local excision in the management of patients with PCL for its potential to ensure negative margins, which reduces rates of recurrence and improves patient satisfaction.

## Acknowledgement

We thank Dr. Darin Trelka, M.D., Ph.D. (Florida Atlantic University Charles E. Schmidt College of Medicine) with obtaining high-resolution pathology images.

## References

1. Sondak V (2000) Sarcomas of bone and soft tissue surgery: Scientific principles and practice. Lippincott Williams Wilkins 2000: 2258-2280.
2. Hung GY, Horng JL, Chen PC (2019) Incidence of soft tissue sarcoma in Taiwan: A nationwide population-based study. Cancer Epidemiol 60: 185-192.

3. Mayerhofer K, Obermair A, Windbichler G (1999) Leiomyosarcoma of the uterus: A clinicopathologic multicenter study of 71 cases. *Gynecol Oncol* 74: 196-201.
4. Di Luigi G, D'Alfonso A, Patacchiola F, Stefano DL, Palermo P, et al. (2015) Leiomyosarcoma: A rare malignant transformation of a uterine leiomyoma. *Eur J Gynaecol Oncol* 36: 84-87.
5. Bharambe BM, Deshpande KA, Surase SG, Ajmera AP (2014) Malignant transformation of leiomyoma of uterus to leiomyosarcoma with metastasis to ovary. *J Obstet Gynaecol India* 64: 68-69.
6. Rotmensch J, Bosnyak S, Montag A (1993) Malignant transition of uterine leiomyomata. *Int J Gynaecol Obstet* 42: 47-49.
7. Yanai H, Wani Y, Notohara K, Takada S, Yoshino T (2010) Uterine leiomyosarcoma arising in leiomyoma: clinicopathological study of four cases and literature review. *Pathol Int* 60: 506-509.
8. Kohlmeyer J, Steimle SA, Hein R (2017) Cutaneous sarcomas. *J Dtsch Dermatol Ges* 15: 630-648.
9. Winchester DS, Hocker TL, Brewer JD (2014) Leiomyosarcoma of the skin: Clinical, histopathologic, and prognostic factors that influence outcomes. *J Am Acad Dermatol* 71: 919-925.
10. Fields JP, Helwig EB (1981) Leiomyosarcoma of the skin and subcutaneous tissue. *Cancer* 47: 156-169.
11. Bernstein SC, Roenigk RK (1996) Leiomyosarcoma of the skin, treatment of 34 cases. *Dermatol Surg* 22: 631-635.
12. Choi JH, Ro JY (2018) Cutaneous spindle cell neoplasms: Pattern-based diagnostic approach. *Arch Pathol Lab Med* 142: 958-972.
13. Starr GF, Dockerty MB (1955) Leiomyomas and leiomyosarcomas of the small intestine. *Cancer* 8: 101-111.
14. Humphreys TR, Finkelstein DH, Lee JB (2004) Superficial leiomyosarcoma treated with Mohs micrographic surgery. *Dermatol Surg* 30: 108-112.
15. Rao UN, Finkelstein SD, Jones MW (1999) Comparative immunohistochemical and molecular analysis of uterine and extrauterine leiomyosarcomas. *Mod Pathol* 12: 1001-1009.
16. Starling J, Coldiron BM (2011) Mohs micrographic surgery for the treatment of cutaneous leiomyosarcoma. *J Am Acad Dermatol* 64: 1119-1122.
17. Liao WC, Wang YC, Ma H (2017) Cutaneous leiomyosarcoma: The clinical experience of taipei veterans general hospital revisited. *Ann Plast Surg* 78: 47-51.
18. Zacher M, Heppt MV, Brinker TJ, Hayani KM, Flaig MJ, et al. (2018) Primary leiomyosarcoma of the skin: A comprehensive review on diagnosis and treatment. *Med Oncol* 35: 135.
19. Svarvar C, Bohling T, Berlin O (2007) Clinical course of nonvisceral soft tissue leiomyosarcoma in 225 patients from the Scandinavian Sarcoma Group. *Cancer* 109: 282-291.

### Author Affiliations

[Top](#)

<sup>1</sup>Florida Atlantic University, Charles E. Schmidt College of Medicine, Boca Raton, FL, U.S.A

<sup>2</sup>Clearly Derm Center for Dermatology, Boca Raton, FL, U.S.A

### Submit your next manuscript and get advantages of SciTechnol submissions

- ❖ 80 Journals
- ❖ 21 Day rapid review process
- ❖ 3000 Editorial team
- ❖ 5 Million readers
- ❖ More than 5000 
- ❖ Quality and quick review processing through Editorial Manager System

Submit your next manuscript at • [www.scitechnol.com/submission](http://www.scitechnol.com/submission)