



CASE REPORT

Cutaneous leiomyosarcoma mimicking arteriovenous malformation

Noureddine Litaïem^{1,2}  | Mariam Tabka^{1,2} | Mouadh Nefiss^{2,3} | Maroua Slouma^{2,4}  | Sana Ben Slama^{2,5} | Faten Zeglaoui^{1,2}

¹Department of Dermatology, Charles Nicolle Hospital, Tunis, Tunisia

²Faculty of Medicine of Tunis, University of Tunis El Manar, Tunis, Tunisia

³Department of Orthopedics, Mongi Slim Hospital, Tunis, Tunisia

⁴Department of Internal Medicine, Military Hospital, Tunis, Tunisia

⁵Department of Pathology, Mongi Slim Hospital, Tunis, Tunisia

Correspondence

Noureddine Litaïem, Department of Dermatology, Charles Nicolle Hospital, Tunis, Tunisia.

Email: Noureddine.litaïem@gmail.com

Abstract

There is a clinical overlap between hypervascularized sarcomas and arteriovenous malformations (AVM). MRI imaging should be interpreted with caution keeping in mind that some cancers could mimic AVM. A biopsy is mandatory in doubtful cases.

KEYWORDS

arteriovenous malformation, leiomyosarcoma, skin cancer

1 | INTRODUCTION

Arteriovenous malformations (AVM) are disfiguring and potentially life-threatening fast-flow abnormal communications between the arteries and veins creating an arteriovenous nidus. The diagnosis of AVM is usually made based on clinical and imaging examinations. Given the risk of bleeding and trauma associated worsening, biopsies are rarely performed.¹ Therefore, some hypervascularized tumors could initially be misdiagnosed as AVM.¹ Cutaneous sarcomas mimicking AVM were rarely reported.

2 | CASE REPORT

2.1 | Case history/Examination

We report herein a case of a 21-year-old, otherwise healthy, woman who presented with a pulsatile mass on the dorsal

aspect of the right hallux evolving for 4 months. She stated that the pulsatile mass appeared after a moderate trauma, then rapidly increased in size with intermittent pain. Physical examination revealed a mildly tender 3 × 3 cm, pulsatile angiomatic mass on the hallux (Figure 1A). The mass was firm and its surface partly covered by hemorrhagic crusts and venous dilatation. Lymph nodes were not palpable. Routine laboratory tests were normal.

2.2 | Differential diagnosis and investigations

Based on the case history and physical examination, both AVM and soft-tissue sarcoma were suspected. Doppler ultrasonography showed a high-flow vascular lesion with arteriovenous shunting.

MRI revealed a 3 cm circumscribed vascular mass on the distal phalanx, with a tangled mesh of dilated arteries and

This is an open access article under the terms of the Creative Commons Attribution-NonCommercial-NoDerivs License, which permits use and distribution in any medium, provided the original work is properly cited, the use is non-commercial and no modifications or adaptations are made.

© 2020 The Authors. *Clinical Case Reports* published by John Wiley & Sons Ltd



FIGURE 1 A, A pulsatile angiomatous mass on the right hallux; (B) MR imaging showing a tangled mesh of dilated arteries and veins connected by focal shunts mimicking arteriovenous nidus

veins connected by focal shunts and with no soft-tissue signal (Figure 1B). *T2*-weighted gradient echo imaging showed hyperintense fast-flow vessel networks.

A deep biopsy under general anesthesia was performed. Histopathological examination showed a subepithelial proliferation of spindle-shaped cells with eosinophilic and fibrillary cytoplasm associated with cytologic atypia and marked pleomorphism. Spindle cells are arranged in fascicles (Figure 2A). Mitotic figures were numerous (5 mitotic figures per 10× field). Immunohistochemical studies revealed that the tumor stained positively for smooth muscle actin (SMA) and epithelial membrane antigen (EMA) and negatively for CD34 and S100 protein. Based on the histopathological and immunohistochemical findings, the diagnosis of grade 1 leiomyosarcoma was established. Total Body CT Scanning was unremarkable.

2.3 | Treatment and follow-up

A transmetatarsal amputation was performed leading to complete excision of the tumor (Figure 2B). No relapses or metastases were detected by clinical examination and CT imaging 12 months after surgery (Figure 2C).

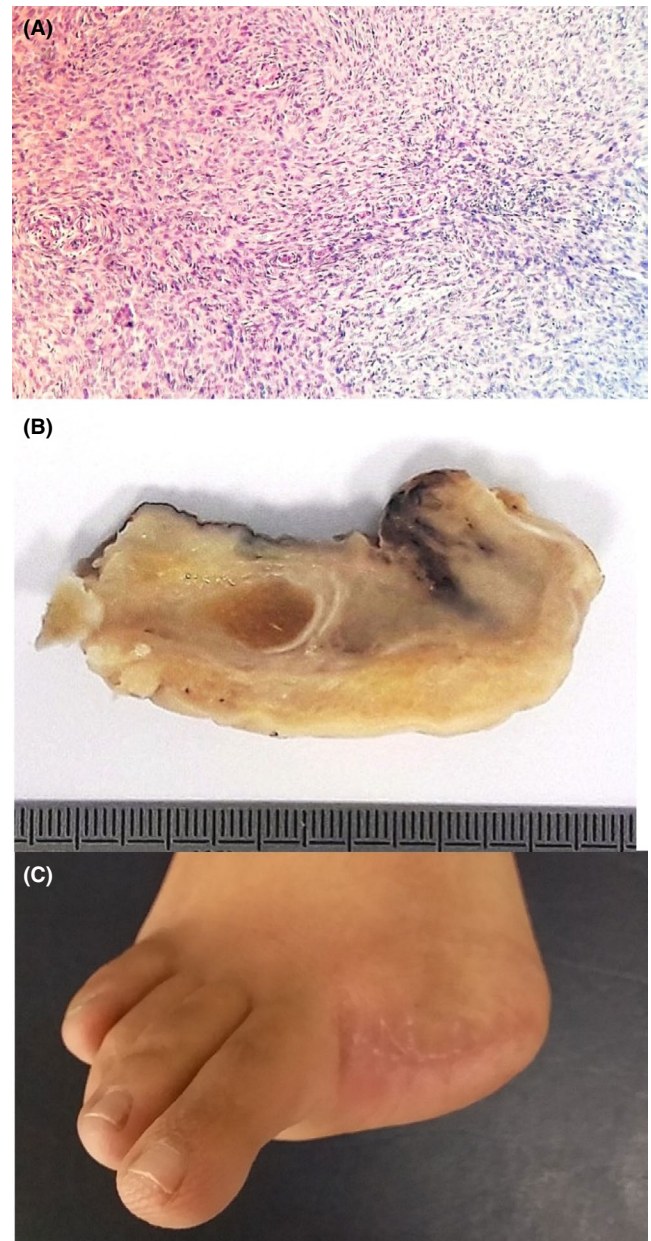


FIGURE 2 A, Leiomyosarcoma. (Hematoxylin-eosin ×200) fascicles of spindle cells with storiform and palisaded arrangement. Mitotic figures are present. B, excised specimen: subcutaneous gray-white firm mass with infiltration of the deep soft tissues and bone; (C) no recurrence was seen 12 mo after surgery

3 | DISCUSSION

In our patient, the diagnosis of AVM was suspected based on the pulsatile nature of the lesion, and the history of trauma-induced worsening. AVM may not be present at birth and worsening is commonly related to puberty, pregnancy, and trauma.² The pulsatile nature of the lesion is not specific to AVM and was described in vascular sarcomas, metastases of hypernephroma or thyroid carcinoma, and subcutaneous tumors of multiple myeloma.³

The diagnosis of AVM is based on imaging techniques especially MRI. MRI is very sensitive but could be misleading. On rare occasions, AVM and sarcomas could demonstrate similar findings on imaging.^{1,4,5} Lee et al reported 2 cases of soft-tissue sarcomas mimicking AVM where MRI did not detect any soft-tissue lesions. These findings were consistent with our patient's MRI results. It is possible that the arteriovenous peritumoral shunt masked the tumor or that it was too small to be detected.¹

In our patient, MRI failed to detect soft-tissue signal. The diagnosis of leiomyosarcoma was based on histopathological findings. Three groups of leiomyosarcomas are described: cutaneous, subcutaneous, and secondary metastatic forms.⁶ Cutaneous leiomyosarcoma arises from the arrector pili muscle.⁶ Complete surgical excision is mandatory. The prognosis of cutaneous leiomyosarcoma is generally good but recurrences after incomplete excision are possible.⁶

There are lessons to learn based on this case report and literature review. First, there is a clinical overlap between sarcomas and AVM. AVMs mimicking Kaposi sarcomas are known as Stewart-Bluefarb syndrome.⁷ MRI imaging should be interpreted with caution keeping in mind that some cancers could mimic AVM. A biopsy is mandatory in doubtful cases.

CONFLICT OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

NL, MT, and MS: contributed to the first draft of the manuscript. SBS: wrote parts of the manuscript related to the histopathological aspects of the disease. MN, MT, and MS and FZ: contributed to the literature search, analysis, and interpretation of the data. FZ: critically revised the manuscript and gave final approval. All authors read and approved the

final manuscript and agree to be fully accountable for ensuring the integrity and accuracy of the work.

ORCID

Noureddine Litaïem  <https://orcid.org/0000-0002-9640-5613>

Maroua Slouma  <https://orcid.org/0000-0002-6889-9687>

REFERENCES

1. Lee HN, Hyun D, Do YS, Park KB, Bae SH. Soft-Tissue Tumors Mimicking Arteriovenous Malformations. *J Vasc Interv Radiol*. 2019;30(10):1660-1664.e7.
2. Enjolras O, Logeart I, Gelbert F, et al. Malformations artérioveineuses: étude de 200 cas [Arteriovenous malformations: a study of 200 cases]. *Ann Dermatol Venereol*. 2000;127(1):17-22.
3. Selmanowitz VJ. Pulsatile cutaneous tumors. *Dermatologica*. 1969;138(1):59-63.
4. Suh JS, Cho J, Lee SH, et al. Alveolar soft part sarcoma: MR and angiographic findings. *Skeletal Radiol*. 2000;29(12):680-689.
5. Chow S, Chu R, Ho S, Chan A, Burd A. Paraganglioma presenting as a vascular malformation: case report. *J Plast Reconstr Aesthet Surg*. 2006;59(1):90-93.
6. Rodríguez-Lomba E, Molina-López I, Parra-Blanco V, Suárez-Fernández R, Pulido-Pérez A. Clinical and histopathologic findings of cutaneous leiomyosarcoma: Correlation with prognosis in 12 patients. *Leiomiosarcoma cutáneo: características clínicas, histopatológicas y correlación pronóstica en 12 pacientes. Actas Dermosifiliogr*. 2018;109(2):140-147.
7. Verma SB, Vasani R. Stewart-Bluefarb syndrome revisited. *Clin Exp Dermatol*. 2016;41(5):522-525.

How to cite this article: Litaïem N, Tabka M, Nefiss M, Slouma M, Ben Slama S, Zegloufi F. Cutaneous leiomyosarcoma mimicking arteriovenous malformation. *Clin Case Rep*. 2020;8:2538–2540. <https://doi.org/10.1002/ccr3.3222>