

Acral Arteriovenous Hemangioma: A Case Report and the Utility of Ultra-High Frequency Ultrasound (UHFUS) in Diagnosis

Stefania Guida^{1,2}, Antonio Podo Brunetti¹, Gianmarco Diego Bigotto¹,
Giorgio Stabile¹, Franco Rongioletti^{1,2}

¹ Vita-Salute San Raffaele University, Milan, Italy

² Dermatology Clinic, IRCCS San Raffaele Scientific Institute, Milan, Italy

Key words: acral arteriovenous hemangioma, arteriovenous tumor, ultra-high frequency ultrasound, vascular tumor

Citation: Guida S, Podo Brunetti A, Bigotto GD, Stabile G, Rongioletti F. Acral Arteriovenous Hemangioma: A Case Report and the Utility of Ultra-High Frequency Ultrasound (UHFUS) in Diagnosis. *Dermatol Pract Concept*. 2024;14(2):e2024089. DOI: <https://doi.org/10.5826/dpc.1402a89>

Accepted: October 10, 2023; **Published:** April 2024

Copyright: ©2024 Guida et al. This is an open-access article distributed under the terms of the Creative Commons Attribution-NonCommercial License (BY-NC-4.0), <https://creativecommons.org/licenses/by-nc/4.0/>, which permits unrestricted noncommercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.

Funding: None.

Competing Interests: None.

Authorship: All authors have contributed significantly to this publication.

Corresponding Author: Antonio Podo Brunetti, MD, Vita-Salute San Raffaele University, Via Olgettina, 58 20132, Milan, Italy. E-mail: a.podobrunetti@gmail.com

Introduction

Acral arteriovenous hemangioma is a rare vascular lesion with an uncertain etiology predominantly affecting middle-aged adults. The lesion typically manifests as a painless 0.5–1 cm erythematous-violaceous papule, predominantly affecting the face and extremities [1]. We present a case study of a middle-aged female diagnosed with acral arteriovenous hemangioma of the lower lip. The clinical, histological, dermoscopic, and immunohistochemical features of this condition are discussed, highlighting the importance of a multidimensional diagnostic approach. Additionally, the application of ultra-high frequency

ultrasound (UHFUS) with color Doppler as a diagnostic tool is introduced.

Case Presentation

A 53-year-old female presented with an asymptomatic persistent lower lip edema that had been present for four months. Previous episodes of transient and self-resolving lower lip swelling were referred. Clinical evaluation revealed swelling of the lower lip (Figure 1A). Dermoscopy showed non-specific vascular congestion with non-arborising telangiectasia in the absence of lacunae (Figure 1B). Several differential diagnoses, including Miescher's granulomatous cheilitis,

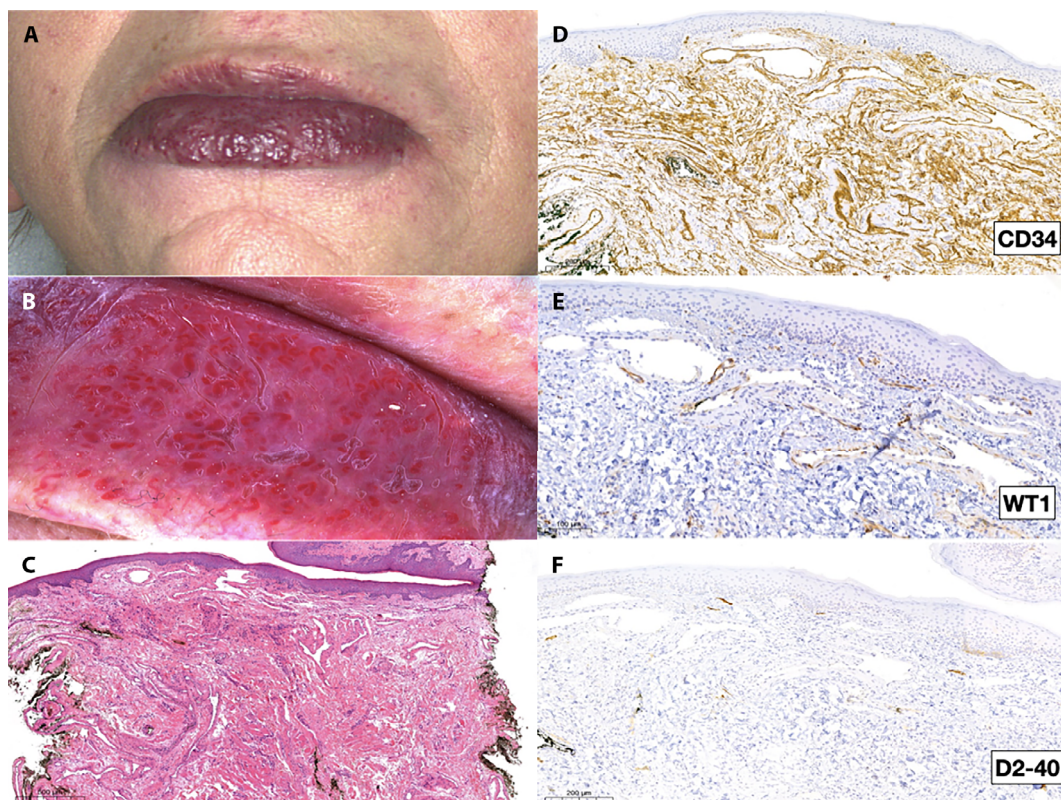


FIGURE 1. (A) 53-year-old patient with edema of the lower lip. (B) Dermoscopy; vascular congestion with non-arborising telangiectasia, without lacunae. (C) Histology: benign full-thickness dermal vascular proliferation (hematoxylin-eosin stain; 50x). (D,E,F) Immunohistochemistry: CD34 positive (D), WT1 focally positive (E) and D2-40 negative (F).

infectious causes, and neoplastic conditions, were considered. Skin biopsy revealed a benign dermal vascular proliferation characterized by thick-walled blood vessels containing muscle and lined with a single layer of endothelial cells intermingled with thin-walled dilated blood vessels (Figure 1C). Immunohistochemistry showed positive CD34 staining, further supporting the vascular nature of the lesion. Additionally, focal positivity for WT-1 and negativity for D2-40 aided in distinguishing this vascular entity from other vascular or lymphatic tumors and malformations (Figure 1D,E,F).

UHFUS Findings: UHFUS with color Doppler was employed to assess the lesion's vascularization. The imaging revealed abnormal arterial and venous vascular spaces, with increased flow observed in the lower lip region compared to the normal upper lip (Figure 2A,B). The UHFUS images were further analyzed using ImageJ software to quantify the observed red and blue pixels, as depicted in Figure 2 C-F [2]. Propranolol was initiated at a dose of 40 mg once daily [3]. However, the treatment was prematurely discontinued due to hypotensive crises. Subsequently, the

patient was referred to vascular surgery to explore alternative treatments.

Conclusion

The exact nature of arteriovenous tumors remains uncertain; however, the prevailing hypothesis proposes a multicentric hamartomatous origin from the suprapapillary vascular plexus with arteriovenous anastomoses. Its clinical diagnosis can be challenging, with accurate identification achieved in less than 5% of cases [4]. Dermoscopy has emerged as a valuable tool in the diagnosis of arteriovenous tumors exhibiting a pattern including non-arborising telangiectasia on a reddish background and the absence of lacunae in 72% of cases [5]. However, despite its utility, dermoscopy has inherent limitations that hinder the assessment of each structure or pattern's specificity in diagnosing arteriovenous tumors, and histopathological evaluation continues to serve as the definitive gold standard for diagnosis. This case highlights the rarity of acral arteriovenous hemangioma and emphasizes

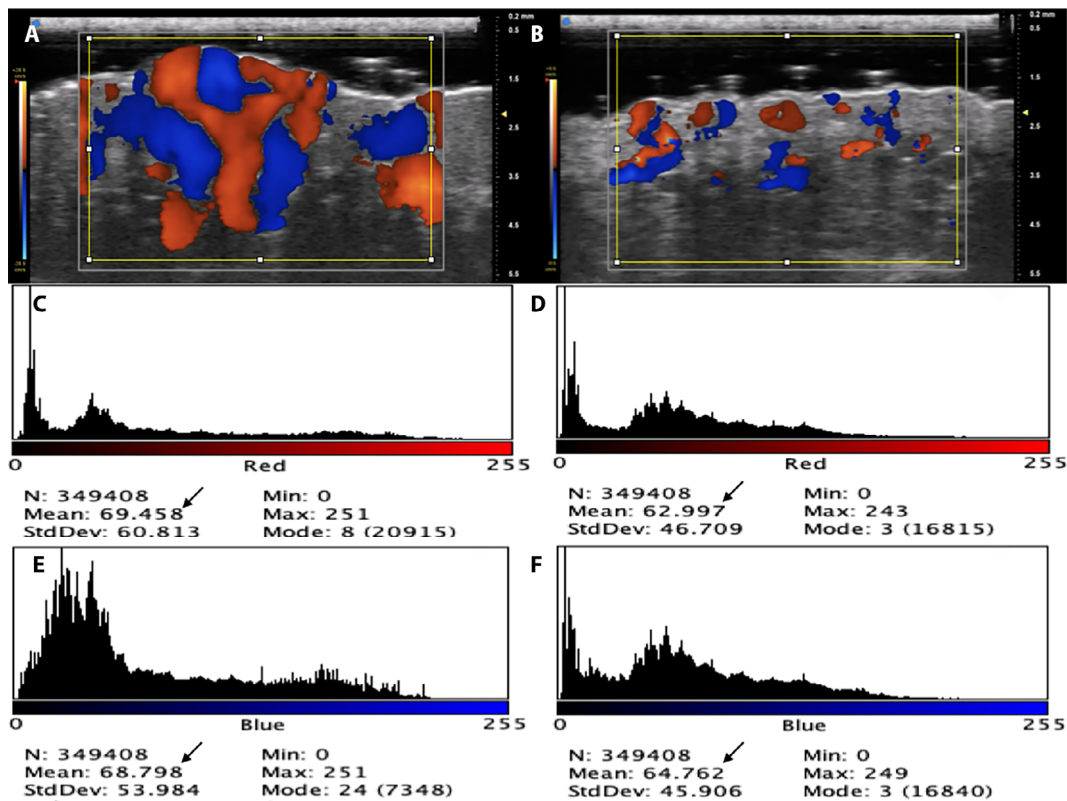


FIGURE 2. (A,B) Color Doppler evaluation with the 70 MHz probe of the lower lip, showing high vascular flow (A) and of the upper lip showing low vascular flow (B). (C,D) Red color histograms of the lower lip showing the average number of red pixels (69458, arrow) (C) and of the upper lip showing the average number of red pixels (62997, arrow) (D). (E,F) Blue color histograms of the lower lip showing the average number of blue pixels (68798, arrow) (E) and of the upper lip showing the average number of blue pixels (64762, arrow) (F).

the importance of integrating clinical, histological, immunohistochemical, and imaging data for an accurate diagnosis. Additionally, the potential application of UHFUS with color Doppler as a diagnostic tool for this vascular entity is proposed.

References

1. Requena L., Kutzner H. *Cutaneous Soft Tissue Tumors*. Lippincott Williams and Wilkins; 2014:485-486.
2. Rasband, W.S., ImageJ, U. S. National Institutes of Health, Bethesda, Maryland, USA, <https://imagej.nih.gov/ij/1997-2018>
3. Lu J, Anvari R, Wang J, et al. Propranolol as a potentially novel treatment of arteriovenous malformations. *JAAD Case Rep*. 2018;4(4):355-358. DOI: 10.1016/j.jdcr.2017.11.005. PMID: 29693069.
4. Connelly MG, Winkelmann RK. Acral arteriovenous tumor. A clinicopathologic review. *Am J Surg Pathol*. 1985;9(1): 15-21. DOI: 10.1097/00000478-198501000-00005. PMID: 3970296.
5. Zaballos P, Medina C, Del Pozo LJ, Gómez-Martín I, Bañuls J. Dermoscopy of arteriovenous tumour: A morphological study of 39 cases. *Australas J Dermatol*. 2018;59(4):e253-e257. DOI: 10.1111/ajd.12775. PMID: 29313900.