Angiokeratoma circumscriptum

Santiago Guillen-Climent MD, Alejandro García-Vázquez MD, Esmeralda Silva Díaz MD, Isabel Pinazo MD

Affiliations: Department of Dermatology, Clinical University Hospital of Valencia, Valencia, Spain

Corresponding Author: Santiago Guillen Climent, Clinical University Hospital of Valencia, Avenida de Blasco Ibañez, 17, 46010, Valencia, Spain, Tel: 34-654138127, Email: santigc85@gmail.com

Abstract

Angiokeratoma circumscriptum is the rarest variant of angiokeratoma. It usually affects females and it is characterized by dark-red to blue-black confluent papules or nodules on lower limbs in a segmental and unilateral distribution. We describe the clinical and histopathological findings in a patient with angiokeratoma circumscriptum and discuss the etiology, associations, diagnosis, differential diagnosis, and treatment.

Keywords: angiokeratoma, circumscriptum, verrucous haemangioma

Introduction

Angiokeratomas are ectasias of dermal capillaries with acanthotic and hyperkeratotic overlying epidermis. Based on the clinical presentation and evolution, five variants have been described: angiokeratoma of Mibelli, angiokeratoma of Fordyce, angiokeratoma circumscriptum, solitary or multiple angiokeratoma, and angiokeratoma corporis diffusum (Fabry disease), [1,2].

Case Synopsis

A 26-year-old woman presented with a congenital lesion on her right lower extremity in a grouped and linear pattern. Her medical history was unremarkable. There was no report of pruritus, pain, bleeding, trauma or family history of similar conditions. Physical examination revealed multiple, well-circumscribed, grouped and segmental, red-to-purple, hyperkeratotic papules on the right thigh (**Figure 1**). Papules were non-pulsatile and non-compressible. Histopathology showed epidermal hyperkeratosis, acanthosis, and ectasia of dermal thin-walled vessels surrounded by epidermal ridges (**Figure 2**). Systemic examination was within normal limits. Laboratory investigations including hemogram and biochemistry panel were normal.

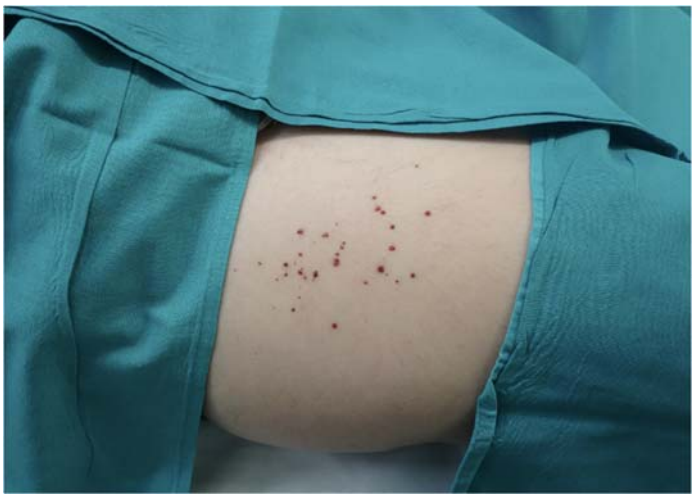

Figure 1. *Clinical image showing multiple, well-circumscribed, grouped and segmental, red-to-purple, hyperkeratotic papules.*

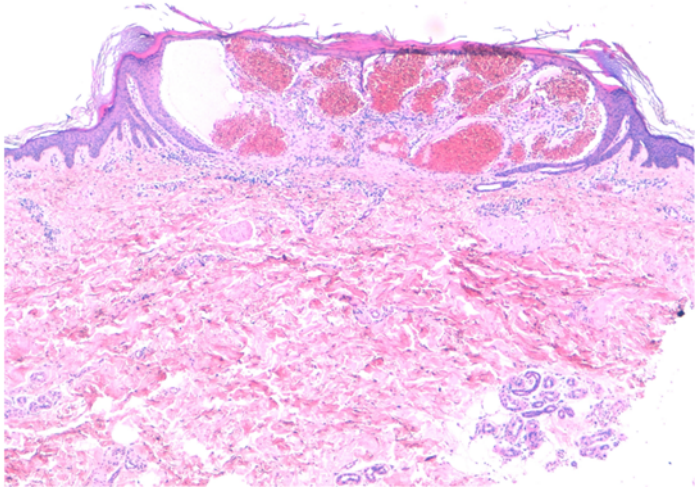

Figure 2. *Histologic image showing epidermal hyperkeratosis, acanthosis and ectasia of dermal vessels surrounded by epidermal ridges. H&E, 4×.*

Comparative measurements of the lower limbs showed no discrepancy. Magnetic resonance imaging of the lower limbs was normal. A diagnosis of angiokeratoma circumscriptum (AKC) was made. The patient chose clinical observation without treatment.

Case Discussion

Angiokeratoma circumscriptum is the rarest variant of angiokeratoma. It is more frequent in females than males. Angiokeratoma circumscriptum is predominantly a congenital lesion. However, there are reports of development later in life [2-6]. The mechanism for development of AKC is unknown. Several etiologic factors have been proposed, such as congenital causes, chronic trauma, tissue asphyxia, pregnancy, and subcutaneous hematomas [7].

Clinically, AKC presents as dark-red to blue-black papules or nodules that often become confluent [7,8]. Commonly it is present on the lower limbs since birth in a segmental and unilateral distribution [1,7]. The initial presentation may be of multiple red macules like a capillary malformation. Then, within a few years, they evolve into papules and nodules; the surface of the lesion becomes hyperkeratotic and verrucous [7,8]. The lesions do not show propensity towards spontaneous regression. Patients may report bleeding and pain with minimal trauma [2].

The entity is benign and it is not frequently associated with systemic disease. There are reports of coexistence of AKC with Klippel-Trenaunay, Cobb syndrome, nevus flammeus, cavernous hemangioma, traumatic arteriovenous fistula, and angiokeratoma of Fordyce [3,7].

The diagnosis of AKC can be usually suspected clinically and confirmed by histopathological study, which identifies characteristic ectatic thin-walled vascular channels in the papillary dermis with acanthosis and hyperkeratosis of the epidermis. The deep dermis and subcutaneous tissue are not affected and if they are involved the probable diagnosis is a verrucous hemangioma [1,2,9]. The differential diagnosis includes verrucous haemangioma, melanocytic nevus, melanoma, capillary aneurysm, pigmented basal cell carcinoma, and Spitz nevus [2]. Small lesions may be eradicated with diathermy, curettage, electrocautery, and cryosurgery. Larger lesions require surgery and laser ablation (carbon dioxide or argon laser), [2,7,8].

Conclusion

In summary, the diagnosis of AKC is more frequent in females and should be considered for grouped and linear papules and nodules present since birth. It is important to diagnose this cutaneous lesion because angiokeratomas of similar appearance may be related to other diseases with notable comorbidity.

Potential conflicts of interest

The authors declare no conflicts of interests

References

1. Sadana D, Sharma YK, Dash K, et al. Angiokeratoma circumscriptum in a young male. *Indian J Dermatol*. 2014;59:85-7. [PMID: 24470669].
2. Ivy H, Julian CA. Angiokeratoma Circumscriptum. StatPearls [Internet]. In: *Treasure Island (FL): StatPearls [Internet]*; 2020, July. [PMID: 31747176].
3. Kassem Youssef H, Backobi E, Michel C. Angiokeratoma circumscriptum neviforme. *Ann Dermatol Venereol*. 2018;145:552-53. [PMID: 30041823].
4. Bouaddi M, Colonna C, Cavalli R, Gelmetti C-M. A New Location of Angiokeratoma Circumscriptum. *Ann Dermatol Venereol*. 2012;139:732-35. [PMID: 23199770].
5. Grewal AR, Chambers CJ, Silverstein M. A case report of adult-onset multiple angiokeratomas with zosteriform distribution. *Dermatol Online J*. 2018;24(10). [PMID: 30677813].
6. Jindal SR, Chalvade P, Jerajani HR. Late Onset Palmar Angiokeratoma Circumscriptum: An Unusual Presentation. *Indian Dermatol Online J*. 2014;5:320-22. [PMID: 25165655].
7. Das A, Mondal AK, Saha A, Chowdhury SN, Gharami RC. Angiokeratoma circumscriptum neviforme: An entity, few and far between. *Indian Dermatol Online J*. 2014;5:472-74. [PMID: 25396132].
8. Del Pozo J, Fonseca E. Angiokeratoma Circumscriptum Naeviforme: Successful Treatment with Carbon-Dioxide Laser Vaporization. *Dermatol Surg*. 2005;31:232-36. [PMID: 15762221].
9. Oppermann K, Boff AL, Bonamigo RR. Verrucous hemangioma and histopathological differential diagnosis with angiokeratoma circumscriptum neviforme. *An Bras Dermatol*. 2018;93:712-15. [PMID: 30156622].