Angiokeratoma Circumscriptum Naeviforme Presenting as a Dark Warty Plaque on the Leg

Dear Editor,

A 45-year-old man presented with a large, dark, keratotic, warty, and friable plaque on the distal posterior aspect of the left leg (Figure 1, a).

The patient reported that the lesion was not present at birth but had appeared approximately at the age of three as an erythematous patch that progressively grew over the time.

During adolescence, the surface of the lesion became rough and warty and was easily traumatized due to its location, resulting in recurrent bleeding episodes over a period of years. For this reason, the patient requested lesion removal.

The patient did not report any other significant comorbidity, and physical examination revealed no other abnormalities.

A shaving biopsy of the lesion was performed, and histopathology highlighted ectatic vascular spaces with some luminal red blood cell beneath a papillomatous and hyperkeratotic epidermis (Figure 1, b).

Based on clinical and histopathological features, a diagnosis of angiokeratoma circumscriptum naeviforme (ACN) was established.

ACN is one of the five disorders belonging to the group of angiokeratomas (AKs) that also include AK of Mibelli, AK of Fordyce, solitary or multiple AK, and AK corporis diffusum.

Among these variants, ACN is the rarest and is seldom studied (1).

AKs are benign vascular anomalies of the superficial vascular plexus that appear as dark red papules and plaques arranged either discretely or in clusters.

ACN lesions are typically situated unilaterally on the lower limbs, especially on the legs and feet, but can occasionally occur elsewhere.

Lesions are generally noted in early childhood. The early lesions are flat and reddish in color, while older lesions become increasingly studded and acquire a verrucous or warty surface. There is no tendency of

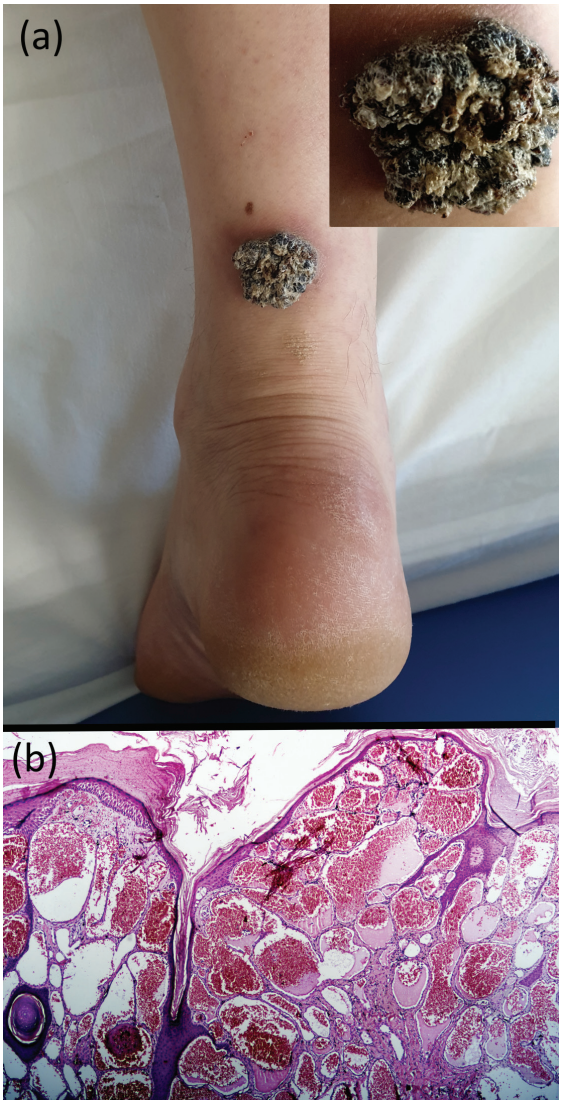

Figure 1. (a) Clinical appearance of the lesion and close-up view of the plaque on the leg showing a verrucous and hyperkeratotic surface (inset). (b) Light microscopy highlighted ectatic vascular spaces with some luminal red blood cell beneath a papillomatous and hyperkeratotic epidermis (hematoxylin and eosin, original magnification x40).

spontaneous improvement, and minor traumas can easily cause beading and infection (2).

While the plaques were linear in disposition in most of the cases reported in the literature, a peculiar feature of our case was the isolated, round, and giant appearance of the plaque.

Even though ACN is not typically associated with other abnormalities, coexistence with other vascular malformations has been reported in some cases, including AK of Fordyce, Cobb syndrome, Klippel-Trenaunay syndrome, nevus flammeus, infantile hemangioma, and traumatic arteriovenous fistula (3).

From a histological point of view, ACN appears as dilated dermal papillary capillaries drained by dilated venules. The overlying epidermis shows a variable degree of acanthosis, papillomatosis, and compact hyperkeratosis. Typically, the deep dermis and hypodermis are not involved, helping distinguish them from verrucous hemangioma (4).

Lichen simplex chronicus, verrucous carcinoma, and verrucous melanoma must also be considered in the differential diagnosis (5).

Recurrent bleeding or cosmetic reasons are common indications for treatment. Surgical excision represents the most effective option. Other possibilities include diathermy, electrocautery, cryosurgery, or laser (6).

In our case, there was no macroscopic residual disease after the shaving biopsy. A collagen dressing was applied, and the wound underwent second intention healing in three weeks. There was no evidence of local recurrence after 18 months.

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Diego Abbenante[1], Beatrice Raone[1], Carlotta Baraldi[1], Miriam Anna Carpanese[1], Annalisa Patrizi [1,2]

[1]Dermatology Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Bologna, Italy

[2]Dipartimento di Medicina Specialistica, Diagnostica e Sperimentale (DIMES) Alma Mater Studiorum – Università di Bologna, Bologna, Italy

Corresponding author:

Diego Abbenante, MD

Dermatology Unit, IRCCS Azienda Ospedaliero-Universitaria di Bologna, Bologna, Bologna, Via Massarenti 1, 40100, Bologna, Italy

diego.abbenante@studio.unibo.it

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