

Giant clear cell acanthoma with dermatoscopic white lines

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Introduction

Clear cell acanthoma (CCA) was first described by Degos in 1962 [1] and has been referred to as Degos' acanthoma. It is a benign epidermal tumor that commonly emerges as a solitary dome-shaped lesion on the legs with no gender predilection [1]. However, eruptive forms and involvements of other areas such as inguinal region, scrotum, vermilion mucosa, scalp, palm, nipple, and hallux have been reported [2,3].

Case Presentation

A 73-year-old Caucasian man presented with an asymptomatic strawberry-like plaque on his right ankle, reportedly present for several years. The plaque was approximately 4 cm in diameter, colored red and purple, and surrounded by a fine collarette of scale (Figure 1). There was no history of pigmented skin changes or melanoma among his family members. Past medical history was notable for ischemic heart disease, diabetes mellitus, and hypertension. His regular medications included aspirin, warfarin, metformin, and atorvastatin.



Figure 1. An asymptomatic erythematous vascular plaque on right ankle present for several years. [Copyright: ©2018 Zargari et al.]

Dermatoscopic examination revealed a variegated red/purple-colored lesion with curved white lines and with dot vessels in a serpiginous ("string of pearls") pattern (Figure 2). There were no colors to indicate melanin (black,

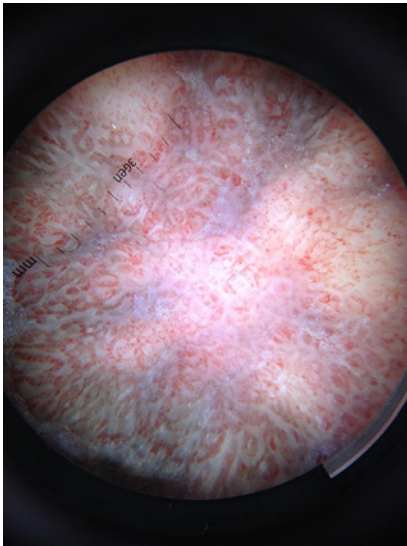


Figure 2. Dermoscopy of the lesion shown in Figure 1 showing a variegated red/purple-colored lesion with curved white lines and with dot vessels in a serpiginous pattern. [Copyright: ©2018 Zargari et al.]

brown, gray, or blue). A biopsy was performed. Histopathological examination showed an acanthotic epidermis with pale-appearing keratinocytes full of a glycogen-rich cytoplasm positive to periodic-acid-Schiff (PAS) staining, consistent with CCA (Figure 3). The lesion was treated with cryotherapy and curettage. In follow-up, the patient had not developed new CCAs after 1 year.

Discussion

CCA is frequently presented as a solitary, slowly growing nodule or plaque on the legs of elderly persons. Its size usually varies from 3 to 20 mm [4]. Perhaps, the original description of Fine and Chernosky is still the most comprehensive one: “CCA has the stuck on appearance of seborrheic keratosis, the vascular look of pyogenic granuloma, the scale and exudation of an eczematous process and advancing rounded border of an epithelioma” [4].

The pathogenesis of CCA remains unknown. There is not significant evidence to support a traumatic or drug-induced origin. Although it was primarily considered to be a benign epidermal

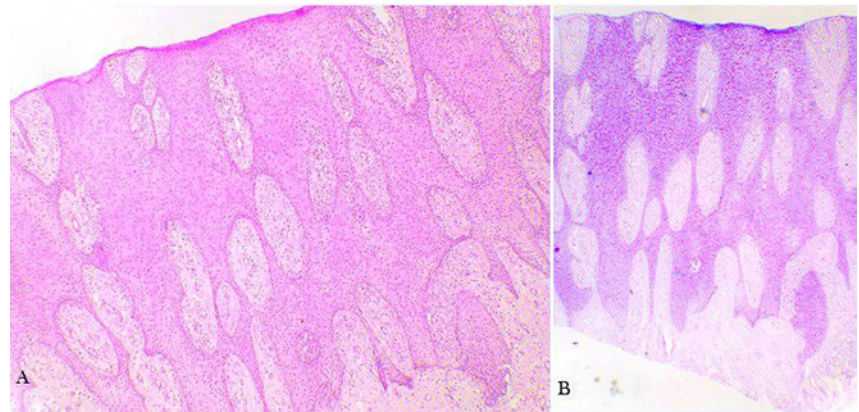


Figure 3. Dermatomicrographs of the lesion in Figures 1 and 2. (A) Hematoxylin and eosin (H&E) stain demonstrating pale-appearing cytoplasm of plump keratinocytes with elongated rete ridges. (B) Pale keratinocytes full of a glycogen-rich cytoplasm positive to periodic-acid-Schiff (PAS) staining. (Original magnification: H&E stain $\times 100$, PAS $\times 40$). [Copyright: ©2018 Zargari et al.]

tumor, a more recent hypothesis suggests that CCA is a reactive inflammatory dermatosis [5]. The higher incidence of CCA in lower limbs of elderly patients favors a reactive nature, probably induced by stasis dermatitis [6].

Shalin et al demonstrated that the concurrence of CCA with syringofibroadenomatous changes also suggests that both of these entities may share derivation from the eccrine apparatus [7]. The clinical variants of CCA include giant, polypoid or pedunculated, pigmented, eruptive, atypical, and cystic patterns [8]. The giant type of CCA measures more than 40 mm. Until now, there are only 5 reports of giant CCA in the English literature, these being located on the foot, buttock, and perineum. Due to the variable clinical presentations of CCA, it frequently is mistaken for several other lesions, including Bowen’s disease, squamous cell carcinoma, malignant melanoma, Kaposi sarcoma, and angiosarcoma [9,10].

Indeed, the diagnosis is rarely made before skin biopsy, and frequently it can be misdiagnosed. Dermoscopy can improve the accuracy in diagnosing CCA [9,11]. The dermatoscopic pattern of CCA was first described by Blum in 2001 as partly homogenous, symmetrically or bunch-like arranged pinpoint-like capillaries [12]. CCA usually has a

unique appearance on dermoscopy, characterized by red dots, globules, and, sometimes, coiled (glomerular) vessels, arranged in a serpiginous pattern. When fully developed, these serpiginous arrangements are strikingly symmetric. However, in some cases the serpiginous vascular pattern is incomplete or partly developed, either showing a forme fruste or a compression artifact but is still clearly recognizable.

Dot or coiled (glomerular) vessels can also be a characteristic of inflammatory dermatoses, such as psoriasis, pityriasis lichenoides, and discoid eczema. Nevertheless, in these diseases, the red dots or coiled vessels are uniformly distributed and do not join together to form serpiginous vascular arrays [12-16]. Other uncommon dermatoscopic features of CCA include the presence of areas of hemorrhage, orange crusts, and a peripheral collarette of translucent scales. A new finding recently described in the literature is the frequent presence of crystalline structures when polarized dermoscopy is used for the evaluation of CCA [17].

CCA is a benign tumor, and when the diagnosis can be made with confidence due to the characteristic serpiginous vascular pattern, treatment is not indicated for small asymptomatic lesions. If treatment is desired, surgical

excision may be the treatment of choice for single lesions. However, cryosurgery remains a very useful alternative, especially for multiple lesions [10].

Conclusions

Giant forms of CCA are extremely rare and confident clinical diagnosis is not always possible. Despite the nonspecific clinical presentation, the giant CCA reported herein had the known specific dermatoscopic feature of dot vessels distributed in a serpiginous arrangement as well as a new feature of curved white lines, not previously reported.

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