Clear Cell Acanthoma: A Clinical, Dermoscopic and Histological Review

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Abstract
Clear cell acanthoma (CCA) is an uncommon, benign epidermal tumor that may be easily misdiagnosed on a clinical basis alone. Although biopsy is commonly performed for diagnosis, perceptive clinicians may suspect a CCA with the use of clinical and dermoscopic findings. We present a case of a suspected clear cell acanthoma confirmed by biopsy along with a clinical, dermoscopic and histological review of the condition.

Introduction
CCA was first described in 1962 and was also known as “Degos acanthoma” and “acanthome cellules claires of Degos and Civatte.”1 There are currently no known risk factors, and the etiology is unknown. It is theorized that the cause may be an inflammatory reaction secondary to an unknown trigger.2 Yet further investigation is necessary to conclude the actual cause. CCA typically presents as an erythematous, solitary papule with a peripheral scale, usually on the lower extremities. Because this description clinically coincides with a multitude of other lesions, our aim is to describe how dermoscopy can distinguish CCA from its differentials, making diagnosis biopsy-free.

Case Report
A 68-year-old white female presented to our outpatient clinic for a full-body skin exam. Her past medical history was significant only for chronic obstructive pulmonary disease. She denied personal or family history of skin cancer. Physical exam revealed a sharply demarcated, 0.3 cm x 0.3 cm, shiny, pink, moist, blanchable papule with a collarette scale located on the left anterior distal shin in conjunction with varicose veins (Figure 1). Dermatoscopic evaluation showed dotted vessels arranged in a linear “string of pearls” distribution, revealing the characteristic dermoscopic vascular pattern seen in clear cell acanthoma (CCA) (Figure 2).3,4

Discussion
CCA is a rare, benign lesion that is oftentimes difficult to diagnose with clinical observation alone. CCA shares clinical features that overlap with a variety of other lesions, making the differential diagnosis extremely broad. Dermoscopically, however, this lesion has unique and specific features, which greatly improves diagnostic accuracy. The dermoscopic features show a stereotypical vascular pattern composed of dotted vessels distributed linearly in a “string of pearls” configuration.

Clinical Findings
CCAs are generally solitary, asymptomatic, red or brown, dome-shaped papules or nodules. They may be covered by scaled edges or appear moist. The size of lesions can range from approximately 3 mm to 20 mm, and they can slowly grow for up to 10 years. When closely examining the surface of the lesion, vascular puncta are present, which easily bleed following minor trauma. These lesions are usually found on the lower extremities in middle-aged to elderly adults, with both sexes affected equally.5,6

Although this is the most common presentation, there are a variety of clear cell acanthoma types, creating a large list of differential diagnoses.
types include giant, polypoid, pigmented, eruptive, atypical and cystic.  

In addition, there have been three recent literature reports detailing "atypical CCA," which some authors argue is a malignant counterpart of CCA. These cases were clinically described as erythematous, moist nodules, all of which were located on the face. Dermooscopically, these lesions portrayed a dot-like pattern of globular capillary vasculature, similar to benign CCA. The literature is still pointing toward calling them benign lesions, secondary to the lack of recurrence. Further research is required for atypical CCA.  

**Differential Diagnosis**
CCA has a vast differential diagnosis that includes actinic keratosis, lichenoid keratosis, pyogenic granuloma, dermatofibroma, basal cell carcinoma, squamous cell carcinoma, inflamed seborheic keratosis, eccrine poroma, clear cell hidradenoma, amelanotic melanoma, and psoriasis. When considering non-pigmented skin lesions such as these, dermoscopic vascular structures are often helpful in making a correct diagnosis. Among this wide differential base, clear cell acanthomas are unique in their dermoscopic distribution of dotted or globular vessels, arranged in a curvilinear pattern.

**Diagnosis**
CCA may be suspected on physical exam, especially when combined with the clues and patterns visualized with a dermatoscope. Confirmatory diagnosis of clear cell acanthoma requires a skin biopsy. Dermooscopically, these lesions are set apart from their differentials by the pattern of their vasculature, rendering a skin biopsy practically unnecessary. Under a dermatoscope, clear cell acanthomas portray pinpoint vessels in a linear pattern, described as pearls on a string.

**Histopathology**
Typically, CCAs are characterized by well-demarcated epidermal hyperplasia made up of large keratinocytes and basal cells full of a glycogen-rich cytoplasm positive to periodic-acid–Schiff staining. An abundance of densely packed, dilated capillaries is seen in a well-demarcated distribution that correlates with the dermoscopic vascular features or red dots and globules outlined above. Parakeratosis, neutrophilic exocytosis and mild spongiosis are also present (Figures 3 and 4).

In the atypical variant of CCA, histological findings consist of cytological atypia of tumor cells with enlarged nuclei, some of which show mitotic figures. In one study, these tumor cells were positive for p63.

**Management and Therapy**
Management of a solitary CCA lesion is by excisional removal. This can be done through a variety of methods including, but not limited to, standard surgical excision, Mohs micrographic surgery, cryotherapy, electrofulguration, curettage and carbon dioxide laser. For cases of multiple or larger lesions, cryotherapy and carbon dioxide laser have been successfully used. In addition, in line with a theorized inflammatory reactive cause, one case report showed regression of CCA after a two-month trial of calcipotriol. In the case of our patient, shave excision combined with electrofulguration was used for diagnosis and treatment.

**Conclusion**
CCAs have a large differential including many lesions that are less benign and occur with much higher frequencies in the population. Under these conditions, the diagnosis of CCA is usually made histologically, after a biopsy has been performed. Since the features of this lesion are dermoscopically distinct, this may afford the clinician more diagnostic confidence. The use of routine dermoscopy may therefore reduce the number of biopsies performed on this benign dermatologic entity.

**References**