Our Dermatology Online

Case Report

Clear cell acanthoma: An atypical localization

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ABSTRACT

Clear cell acanthoma (CCA) is a rare, benign epidermal tumor first described in 1962. Typically, it manifests as a solitary lesion on the lower extremities, posing diagnostic challenges. Dermoscopy and histology are key for accurate diagnosis. Herein, we report an unusual case of CCA in the axilla. A 52-year-old patient presented with a persistent axillary lesion persistent for two years. Dermoscopy revealed a pearl-necklace vascular pattern, raising the suspicion of CCA. Histological examination confirmed CCA with subacute hidradenitis. This case underscores the need to consider CCA even in atypical locations and highlights the diagnostic significance of clinical, dermoscopic, and histological findings.

Key words: Clear cell acanthoma, Axilla, Atypical localization, Benign epidermal tumor

INTRODUCTION

Clear cell acanthoma (CCA) is a rare benign epidermal tumor described for the first time by Degos and Civatte in 1962, usually presenting as a solitary lesion affecting the lower extremities.

Diagnosis is difficult clinically, guided by dermoscopy, and confirmed by histology. Herein, we report the case of a patient with an atypical localization of CCA in the axilla straightened by dermoscopy.

CASE REPORT

A 52-year-old patient with no previous medical history consulted for an asymptomatic axillary lesion that had been evolving for two years and for which the patient had applied several healing creams without disappearance. She never presented associated systemic symptoms. Dermatological examination revealed a well-limited, scaly, erythematous plaque measuring 3 cm in the left axilla (Fig. 1). Dermoscopy showed glomerular vascularization of linear distribution describing a pearl necklace (Fig. 2). The diagnosis of CCA was suspected, and biopsy excision confirmed CCA associated with subacute hidradenitis. A histological examination showed hyperplasia of the epidermis with an acanthosis composed of enlarged clarified keratinocytes, topped by ortho and parakeratotic keratosis.

The dermis was the site of a dense focal inflammatory infiltrate of neutrophils, lymphocytes, and histiocytes, which also infiltrated the eccrine and apocrine glands (Figs. 3a and 3b).

PAS staining was positive for clarified keratinocytes (Fig. 3c).

The patient's follow-up did not reveal any recurrence after one year of evolution.

DISCUSSION

CCA is considered a rare tumor, affecting men more frequently than women and more commonly appearing in individuals aged between the fifth and seventh decades of life [1].

The precise cause of CCA remains uncertain [2], as it is unclear whether CCA is a result of a neoplastic

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or inflammatory process [3]. Despite CCA often appearing as a single lesion, it shares significant histopathological, cytochemical, immunohistological, and dermoscopic similarities with psoriasis [4,5]. Additionally, the presence of EMA positivity in CCA is also observed in reactive, inflammatory conditions. Furthermore, the cytokine expression pattern in CCA mirrors that seen in other types of inflammatory dermatoses. These findings collectively suggest that CCA is more likely a reactive response rather than a neoplastic one [6].

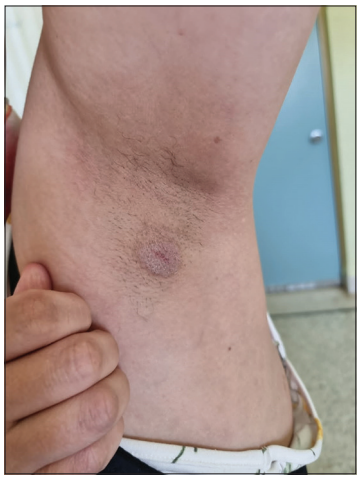

Figure 1: Rounded, scaly erythematous patch in the armpit.

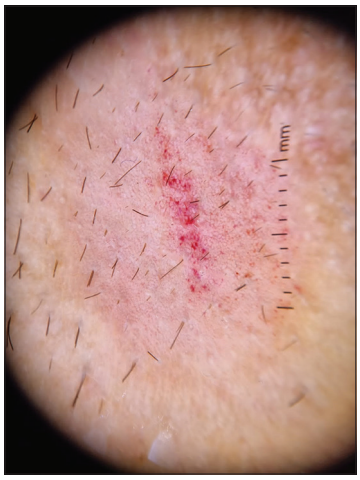

Figure 2: Dermoscopy showing dotted vessels describing a pearl-necklace appearance.

The lesion is usually single and mainly affects the distal lower limbs. Other localizations have also been reported, such as the hands, wrists, buttocks, abdomen, and nipples [2,7]. However, to our knowledge, no axillary site has ever been described before.

The size of CCA generally varies from 5 to 20 mm in diameter. Nevertheless, a giant variant has been reported measuring 45 × 40 mm, known as the giant form of Degos acanthoma [5]. Other clinical forms have been described, including the polypoid, pigmented, verrucous, and other forms [8].

CCA appears as a bright-red to dark-red papule that slowly develops into a hemispherical, conical, or plaque-like shape. The surface is generally smooth yet may be mamelinated with a scaly border.

On dermoscopy, the lesion has a highly characteristic and specific appearance, characterized by red dots, globules and, in some cases, glomerular vessels, on a pale or pink, homogeneous background. The vessels are linear, creating a pearl-necklace pattern [9]. Therefore, this arrangement of vessels is not pathognomonic of CCA; it may also be observed in pigmentated seborrheic keratosis.

The differential diagnosis may be made with benign epithelial lesions such as seborrheic keratosis and actinic keratosis, or malignant lesions such as Bowen,

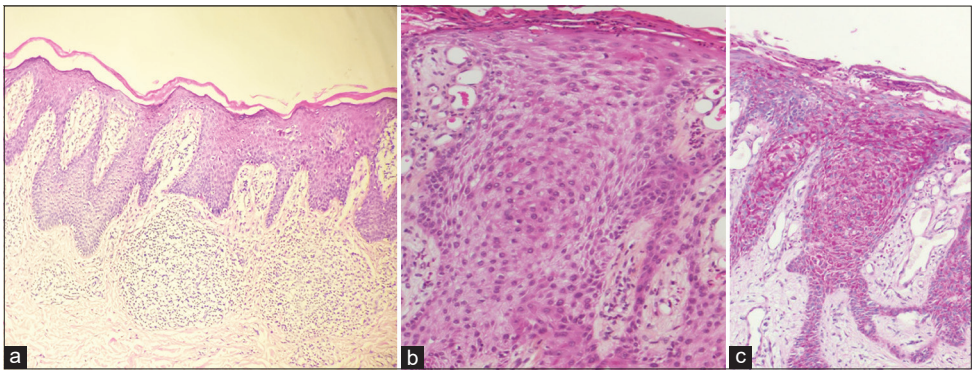

Figure 3: (a) Histological section of skin tissue bordered by acanthotic epidermis with psoriasiform hyperplasia (H&E; 100x). (b) Keratinocytes clarifi ed (HES x 400). (c) Clarifi ed Keratinocytes are PAS+ (x 200).

basocellular carcinoma, and squamous cell carcinoma. On the other hand, inflammatory dermatoses, including psoriasis vulgaris, eczema and lichen planus, as well as prurigo and folliculitis, may also be confused with CCA [1,2].

Histology provides diagnostic certainty. CCA presents a compact acanthosis, clearly separated from the surrounding epidermis. Focal papillomatosis and parakeratosis may be observed, often with reduced or absent stratum granulosum [10]. Keratinocytes, known as clear cells, appear enlarged with pale, PAS-positive cytoplasm, containing glycogen and small, centrally located nuclei. In the tumor zone, the epidermis becomes edematous and infiltrated by isolated polymorphonuclear leukocytes, sometimes resulting in microabscesses in the stratum corneum.

Moreover, the histopathological features underlying the dermoscopic phenomenon of the string of pearls structure have not been definitively elucidated. It is hypothesized that the dots and clods visible in dermoscopy correspond to the dilated vessels at the tips of the papillae [11]. In tangential sections through the CCA, the cut tips of the papillae with their dilated capillaries present as round islands tightly packed together in the area of acanthosis [12].

The recommended treatment for CCA is surgical excision. In some cases, and especially when lesions are multiple, curettage, electrocoagulation, cryotherapy, or CO_2 laser treatment may be proposed [2,13]. Topical treatments such as 5-fluorouracil and calcipotriol are also therapeutic alternatives [4].

The clinical evolution of untreated ACC is benign. Only one case of spontaneous resolution has been reported [14]. No cases of malignant transformation have been reported [15].

CONCLUSION

The pertinence of our observation lies in the importance of considering the possibility of CCA even when lesions occur outside the lower limbs. The clinical and dermoscopic features may provide valuable clues to the diagnosis. However, it is imperative to confirm this by histology.

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