DOI: 10.3315/jdcr.2010.1048

Journal of Dermatological Case Reports

Multiple eruptive clear cell acanthoma

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Key words:

clear cell acanthoma, cryotherapy, multiple, tumor

Abstract

Background: Clear cell acanthoma is a rare solitary benign epidermal tumor of unknown etiology. The disease arises in the middle-age, with no sex predominance. It appears as a single reddish papule or papule-nodule and a peripheral scaling collarette is characteristic. Although solitary lesions are the rule, less than 30 cases of multiple Clear cell acanthoma have been described in the literature to date.

Main observations: We report an unusual case of a healthy 74-year-old male with multiple clear cell acanthoma on the lower extremities treated successfully with cryotherapy.

Conclusion: Despite significant progress in treatment of clear cell acanthoma, cryotherapy, based on liquid nitrogen, remains an important alternative in treating multiple clear cell acanthomas.

Introduction

Clear cell acanthoma (CCA) is a usually solitary benign epidermal tumor of clear glycogen-containing epithelial cell, first described in 1962 by Degos *et al*.[1] The etiology remains unknown. Clinically, it occurs frequently on the lower limbs, although lesions on the inguinal region or scrotum, face, vermilion mucosa, scalp, far palm, trunk, nipple, buttock, forearm, head and toe should be present.[2,3] The disease arises in the middle-age, with no sex predominance. It appears as a single reddish papule or papule-nodule and a peripheral scaling collarette is characteristic, but not always present;[4] some nodules will be covered with a thin crust rather than keratinous layer and often exuding a slight moisture.[1,3] Other features that may be present include a stuckon appearance and a vascular blush.[5] They usually reach a diameter of 1-4 mm until few centimetres, but a giant form has been descripted by Duperrat *et al*.[1] Although solitary lesions are the rule, less than 30 cases of multiple CCA (from 2 up to 400 lesions) have been described in the literature to date.[6] The clinical diagnosis frequently needs to be supported using dermoscopy and, in some cases, histology is also required. Dermoscopically dotted or globular vessels, similar to those seen in psoriasis and Bowen disease, are found.[7,8] Thus, in lesions with this pattern, located on an extremity, unequivocal differentiation from Bowen's disease may be difficult based on clinical and dermoscopic features alone, necessitating biopsy for histological examination. Reflectance Confocal Microscopy has been used for the evaluation of non-melanocytic neoplasms, showing microscopic findings similar to optical histology.

Case report

A 74-year-old man was admitted to our clinic for 60 asymptomatic rounded papules and papule-nodules on the lower extremities (Fig. 1), appeared more than 20 years and gradually increased in number. On physical examination, the papules were flattened, bright red and sharply circumscribed, with a diameter of 2-12 mm, sometimes with a peripheral scaling collarette. Routine blood examination did not reveal abnormality. His medical history was unremarkable and no others abnormal physical signs were found. There was no history of drug ingestion. An incisional skin biopsy was performed, and the specimens revealed marked acanthosis, epidermal cells with clear cytoplasm and abundant glycogen, as demonstrated by positive period acid-Schiff (PAS)

staining and removal of the staining after diastase digestion (Fig. 2). A diagnosis of multiple CCA therefore was made. The lesions were treated by cryotherapy, with liquid nitrogen and a cotton wool swab, applied for 15 seconds and repeated after 1 minute. All lesions resolved with minimal residual scarring after four sessions with no relapse at 6 months clinical follow-up.

Figure 1

Rounded papules and papule-nodules on the lower extremities.

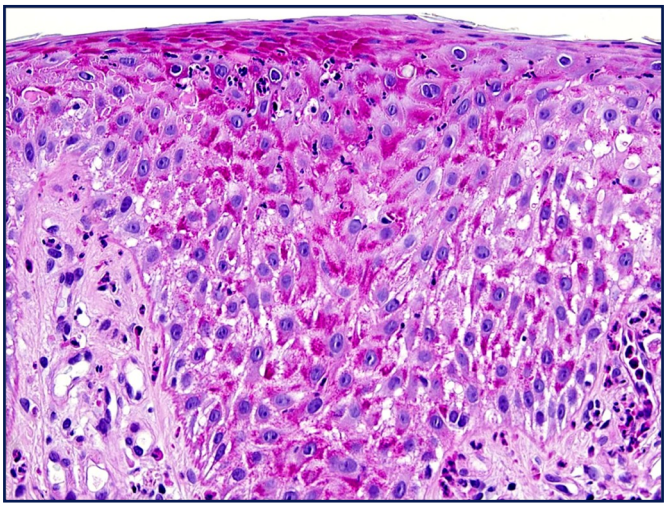

Figure 2

The epidermal cells showed clear cytoplasm and abundant glycogen, as demonstrated by positive period acid-Schiff (PAS) staining and removal of the staining after diastase digestion (original magnification x 10).

Discussion

Clear cell acanthoma (CCA) is a usually solitary benign epidermal tumor, first described in 1962 by Degos *et al*.[1] It occurs frequently on the lower extremities of middle-age patients of both sexes. The etiology is not well understood. There is no evidence that toxic substances, trauma or drugs lead to the onset of lesions. In CCAs seems to exist a metabolic enzyme defect. This enzyme performs an important role in the synthesis of keratine.[6] Some Authors support the existence of an interrelation with the output of melanocytes and interaction between melanocytic-keratinocytic. Someone suggests that lesions are benign epithelial neoplasm, while others support the hypothesis of inflammatory origin.[7] Neither spontaneous regression or malignant transformation have been reported. It is often confused with a lot of other diseases such as: seborrhoeic keratosis, eczematous dermatitis, pyogenic granuloma, hemangioma, psoriasis, guttate parapsoriasis, histiocytoma, basal or squamous cell carcinoma, common wart, amelanotic melanoma, dry skin and ichthyosis.[2,3] Dermoscopy could be performed but histological confermation is required for a certain diagnosis. There is an association with other skin diseases: xerosis, hyttiosis and seborrhoic keratosis.[8] The most distinctive histopathologic features of CCA are well-demarcated: acanthotic epidermis composed of glycogen-rich clear cells that can be revealed by positive PAS (Periodic acid-Schiff) staining and removal of the staining after diastase digestion. The epidermis is frequently infiltrated by abundant numbers of neutrophils, which may form microabscesses in the parakeratotic stratum corneum. The Malpighian layer shows slight spongiosis and several pyknotic neutrophilis in the intercellular spaces.[4] In the papillary dermal there are ectasia and a mixed cellular infiltrate. The acanthoma is limited by a definite line of demarcation produced by an increase in size of the epidermal cells except those in the basal layer. The histochemical examination underlines the absence of phosphorylase, a constitutive enzyme, able to demote glycogen.[5] Electron microscopy studies have revealed glycogen granules.[11,12] Videodermatoscopy has been recently introduced for the diagnosis of CCA, showing homogeneous pinpoint-like vascular structures, that at an higher magnification have a bush-like aspect.[13] At present two main forms of CCA are recognized: solitary CCA and multiple CCA. Although solitary lesions are the rule, less than 30 cases of multiple CCA (from 2 up to 400 lesions) have been described in the English literature to date. Multiple CCA, first described by Delacretaz in 1964,[14] represents a rare variant of acanthoma subdivided into a discrete variant with less than 12 lesions and a rare eruptive form with more than 30 lesions;[4] our case with 60 lesions could be classified as multiple eruptive CCA. Treatment consists in electrofulguration, curretage, surgical excision. In case of multiple lesions, cryotherapy or topical 5-fluoruracil are used.[15,16] The carbon dioxide laser can also be used with good outcome.[17] In our case the lesions were multiple, so a surgical approach was found to be unworkable. Therefore we opted for cryotherapy, methodical of simple execution, cheap and painless. After four sessions lesions were completely

healed without relapse and with an acceptable aesthetic result. In conclusion we can say that cryotherapy is still an optimal method for treating forms of Multiple eruptive clear cell acanthoma.

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