CENTRIFUGAL NECROTIC KERATOACANTHOMA

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Abstract

Centrifugal necrotic keratoacanthoma is a quite rare variant of keratoacanthoma, with a locally aggressive behavior and no tendency to spontaneous involution. We report a typical case of the nasal-orbital region. Based on its clinical, histological and behavioral picture, centrifugal necrotic keratoacanthoma appears as variant of giant keratoacanthoma and can be regarded as an intermediate form in a spectrum where keratoacanthoma and overt squamous cell carcinoma represent the most benign and the most malignant end, respectively, as also highlighted by analogies between the two lesions recently reported in the field of molecular biology.

Key Words: *Centrifugal necrotic keratoacanthoma, keratoacanthoma, low-grade squamous cell carcinoma*

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Introduction

Keratoacanthoma is a rapidly growing tumor with no potential for metastatic spread and a well-defined cycle of growth eventually ending in spontaneous involution.[1,2] Besides the common solitary form, a few less frequent variants have been recognized, including centrifugal necrotic keratoacanthoma. This was first described four decades ago by Lapiere and, it has been ignored, for many years, by the international literature.[3,4] We report a case of centrifugal necrotic keratoacanthoma whose clinical and histological features are characteristic and obviously different from the typical appearance of the classic solitary form.

Case Report

A 60-year-old man, living in Sicily, showed a large, painful, tumor involving the left lateral aspect of the nose and the corresponding orbital region. The lesion had arisen as a small-domed nodule with a smooth, reddish surface, which had rapidly enlarged in the course of five months, developing a crateriform configuration with a central necrosis. Physical examination revealed a 6 × 5 cm, cup-shaped lesion, which showed an elevated and sharply outlined edge with a smooth, flesh-colored surface and an irregular central crater filled with gray-black necrotic material (Fig. 1).

A radial biopsy was taken from the lesion under local anesthesia and processed for histological observation, while the rest of the lesion was thoroughly electrodesiccated and curetted. The specimen, which extended from the margin of the lesion to its necrotic center, showed conspicuous epithelial proliferation with large endo-exophytic, hornfilled isles and strands of squamous cells projecting in the dermis up to its deepest third (Fig. 2). On the external side of the lesion, the

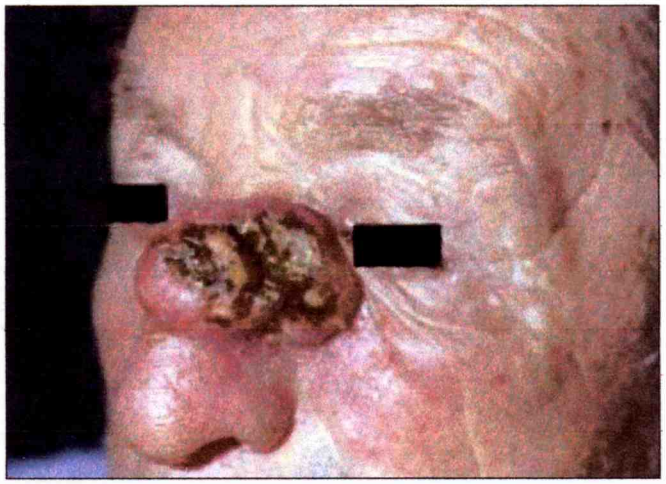

Fig. 1: Centrifugal Necrotic Keratoacanthoma of the nasal orbital region. (Reprinted with permission from Antologia Dermatologica: Tosti A, Fazzini M.L, Fiorella S.; Edizioni Minerva medica, Torino 2001)

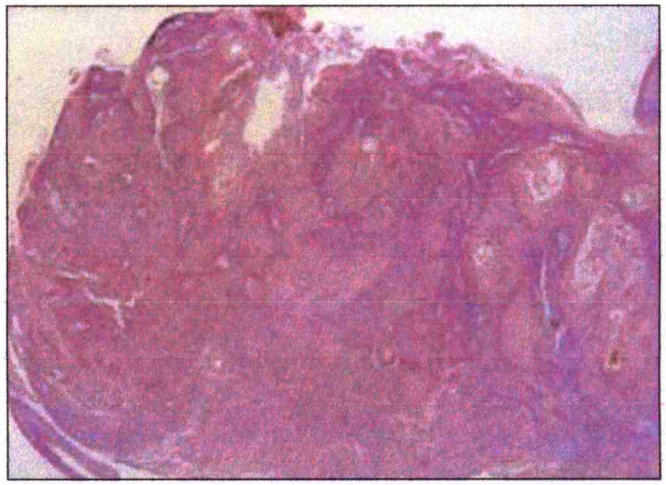

Fig. 2: Centrifugal Necrotic Keratoacanthoma external portion of the lesion conspicuous, peripherally lipped, endo-exophytic proliferation of keratinocytes with an arciform inferior profile. Hematoxylin and eosin stain, magnification 25×

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epidermis stretched like a lip over the proliferation, while on the opposite side, corresponding to its center, a multilayered mound of keratin mixed with necrotic debris, neutrophils and red blood cells was present. The inferior profile of the tumor, considered on the whole, drew an arciform line giving the impression of an expansile growth, although the single epithelial isles and strands showed jagged contours. These masses were mostly composed of well-differentiated keratinocytes with typical nuclei and large eosinophilic glassy cytoplasm. The concentric keratinization was progressively more pronounced toward the center of the lesion and the horny whorls, together with increasingly larger admixtures of red cells and necrotic material, eventually merged into the central necrotic mass. A few foci of individual keratinization, with atypia and mitotic figures, were seen. The stroma and the underlying dermis showed a heavy, predominantly lympho-histiocytic infiltrate with a few plasma cells and neutrophils and were focally necrotic in the center of the lesion. Based on these architectural and cytological features, the diagnosis of centrifugal necrotic keratoacanthoma was issued.

In the subsequent follow-up there were two instances of recurrence, five months and one year after the electrodessication, respectively. The lesions were surgically excised and the histological examination showed in both a relatively well-circumscribed epithelial proliferation with concentric keratinization and areas of atypia and mitoses, ascribable to low-grade squamous cell carcinoma. Until the present day, five years after the first appearance of the tumor, no signs of further recurrence have been observed and the patient is in good health, although a scar with cosmetic defect has ensued from the lesion.

Discussion

Centrifugal necrotic keratoacanthoma was first named in 1965 by Lapiere, who described its distinctive features and recognized it as a precise clinico-pathologic entity.[3,4] According to the author's observations, this form shares with common keratoacanthomas the localization to photoexposed areas, the rapid development, the sharp delimitation with mainly exophytic growth, the hemispherical shape with a central crater and the symmetrical arciform inferior contour observable histologically. However, it deviates from the classic form, for its still faster expansion, its huge size, the central necrosis and the peripheral inflammation, as well as for the pain that it causes. Moreover, at a difference from the classic form, it does not tend to heal spontaneously, but is bound to grow indefinitely, eventually resulting in considerable destruction of the tissue and sometimes evolving in metastasizing squamous cell carcinoma. For these reasons Lapiere considered this tumor as an intermediate form between keratoacanthoma and squamous cell carcinoma and gave the name of kerato-acantho-spinalioma to his cases with malignant evolution.[3-4]

Our case, which thoroughly fits Lapiere's description, is a significant reminder of this unusual form of keratoacanthoma, which, after the original report, has featured quite rarely in the literature and, to our knowledge, has not been reported in recent years. Based on its clinical, histological and evolutionary picture and in accordance with Lapiere's opinion, this entity should be located in intermediate position in a spectrum whose most benign and most malignant ends are common keratoacanthoma and high-grade squamous cell carcinoma, respectively. On the other hand, the hypothesis of a continuum, with progressively more aggressive stages finally merging with overt malignancy, is supported by the demonstration of only subtle differences between keratoacanthoma and squamous cell carcinoma in regards to P53 and Ki-67 expression, nucleolar organizer enumeration, as well as cyclin, cyclin-dependent Kinases and H-ras activity.[5,6] In this context, centrifugal necrotic keratoacanthoma could be reasonably considered either as a premalignant form or as a low-grade carcinoma sharing with common keratoacanthoma the follicular histogenesis. There are in both cases striking biological and behavioral analogies between this form and the group of verrucous carcinoma, currently including epithelioma cuniculatum oral florid papillomatosis and Buschke -Lowenstein's giant condylomata.[7]

Centrifugal necrotic keratoacanthoma could well represent a third variant of giant keratoacanthoma, which it overlaps in the gigantic size, the rapid development and the locally aggressive behavior, although differing from this for its distinguishing huge central necrotic-hemorrhagic mass.

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