"Nevoid Eruptive Keratoacanthoma" - Yet Another Atypical Manifestation of Generalized Keratoacanthoma

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Sir,

Keratoacanthomas are dome-shaped lesions with umbilicated center filled with keratin plug. They are known to be solitary but may be multiple. The generalized variety can occur over prurigo nodularis and sites of trauma. Linear Darier's disease like lesions of this disorder may present a diagnostic dilemma. We report a rare case of multiple keratoacanthomas occurring in a nevoid distribution resembling the nevoid Darier's disease.

A 26-year-old youth presented with papular, nodular, and noduloulcerative lesions on the left side of his body since birth. These were pruritic and few were painful. Sites affected were the axilla, scapular region, left lower quadrant of the abdomen and the corresponding lumbar region, and inner side of the left lower extremity up to the medial side of the left foot, often following Blaschko's lines [Figures 1 and 2]. Some papules had coalesced to form plaques showing central crater. Nails were normal. A few lesions were follicular. No lymphadenopathy was present. Family history was not contributory, and there was no complaint of exacerbation with sun exposure. Routine blood tests and X-ray of the chest were normal as was ultrasonography of the whole abdomen. Person was otherwise healthy and so, extensive investigations of the gastrointestinal tract or the bone marrow were not carried out. Papular lesion on histopathology showed hyperkeratosis, parakeratosis, and acanthosis without any dyskeratotic cells. The dermis was unremarkable without any dysplastic cells, with only inflammatory cells in the upper part [Figure 3]. The crateriform lesion displayed buttressing of the invagination, which was filled with keratin [Figure 4]. Squamous dysplasia or dyskeratosis was not discernible. Horn pearls were absent as was any basaloid cells. Histopathology proved the lesions to be keratoacanthomas.

Generalized eruptive keratoacanthomas are of the Grzybowski type, the familial Ferguson Smith type, and the Witten and Zak type, which is a mixture of the two earlier mentioned types.[1] Eruptive keratoacanthomas have been known to occur with diverse conditions such as after immunosuppressive therapy, with Muir–Tore syndrome, and also with hypertrophic lichen planus.[2] Human papillomavirus serotypes, exposure to chemical carcinogens, such as coal tar and oils, and preexisting diseases such

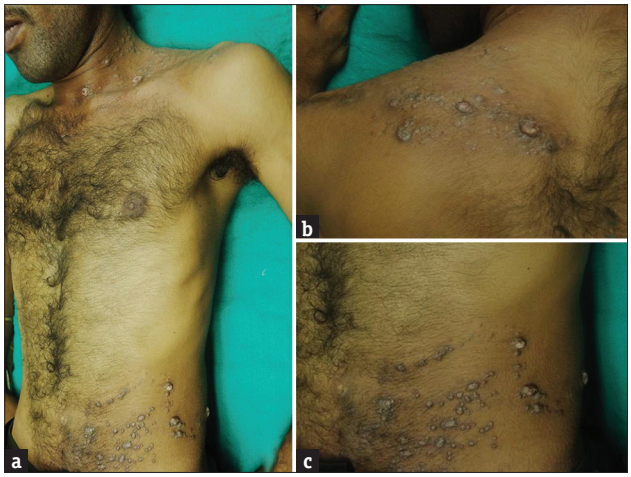

Figure 1: (a) Hyperkeratotic papules and plaques on left side of the body; (b) papules and plaques on the back; (c) lesions along Blaschko's lines on left lower abdomen

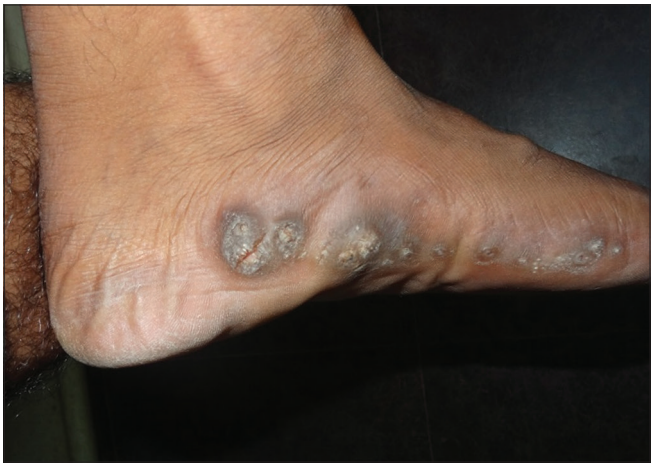

Figure 2: Plaques along the medial side of the left foot

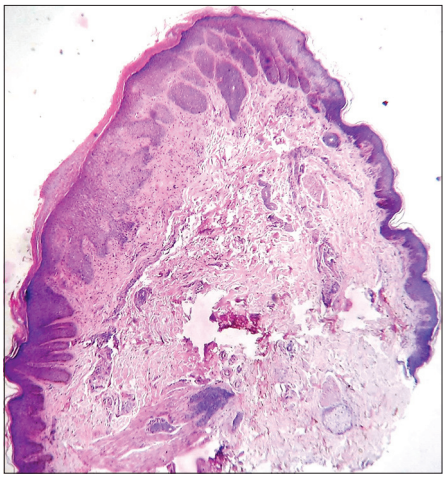

Figure 3: Histopathology of a papular lesion showing hyperkeratosis, parakeratosis, and acanthosis (H and E, × 10)

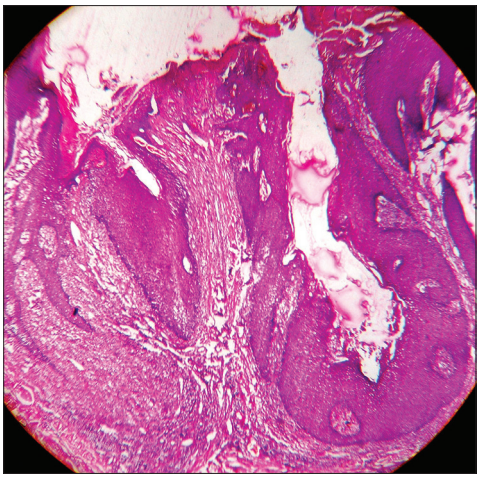

Figure 4: Histopathology of ulcerated plaque showing keratinized crater (H and E, × 40)

as eczema, psoriasis, and seborrheic dermatitis are also implicated among the etiologic factors of multiple keratoacanthomas.[3] Sun exposure has been discovered to be the most common triggering factor of keratoacanthomas. Such lesions have been reported to develop in linear epidermal nevus.[4] Associations with gastrointestinal malignancy and bone marrow disorders have been reported.

Affection of the oral mucosa is common in the Grzybowski type, which usually presents between the fifth and seventh decade of life and does not occur in the Ferguson Smith type. Our case did not present any oral lesions. Both the types are known to resolve spontaneously. The patient examined by us complained persistence of lesions.

In Grzybowski type, multiple lesions are also present on the face leading to mask-like facial appearance and ectropion. Our case did not present any of these features.[3]

A blaschkoid distribution of papules, which were somewhat hyperkeratotic initially, suggested the possibility of linear Darier's disease. Histopathology of the two lesions, however, failed to substantiate the disorder as any of the two types of linear Darier's disease. Grover's disease and epidermal nevus too could be ruled out by the same procedure. Squamous and basaloid progression have been reported in keratoacanthomas.[5,6] Eruptive keratoacanthomas have been described in Hodgkin's disease.[7] Acitretin given in adult dose failed to provoke any positive response in the case.

Genetic evaluation of our subject could not be performed. Localized form of Darier's disease has demonstrated it to be a genetic mosaic of the generalized form.[8] Exciting possibilities are open if such genetic studies can be done in all cases of atypical forms of keratoacanthomas. This unusual presentation of keratoacanthoma also presence diagnostic dilemma, so a histopathology and extensive literature search is necessary for correct diagnosis and treatment. Treatment of such keratoacanthomas is a challenge and should be probed further.

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