CASE REPORT

Cutaneous acantholytic dyskeratotic acanthoma: A case report

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ABSTRACT

Acantholytic dyskeratotic acanthoma (ADA) is a benign disorder marked by limited epidermal growth with acantholysis and dyskeratosis. Basal cell carcinoma is the most common clinical diagnosis, which leads to excision and histopathologic evaluation.[1] Ackerman was the first to describe the incidental finding of focal acantholytic dyskeratosis.[2] Acantholytic acanthoma was first characterized as a solitary lesion with acantholysis-like histology but no dyskeratosis.[3] Acanthomas with acantholysis but no substantial dyskeratosis or dyskeratotic acanthomas with acantholysis are uncommon.[1] In this paper, we describe a case of ADA in a 52-year-old man presenting with scaly, reddish, brownish papule on auricle of right ear.

CASE REPORT

A 52-year-old male patient presented with a solitary scaly reddish brownish papule of auricle of right ear of one-year duration. The condition started with gradual onset and followed slowly progressive course. The lesion was painless and non-tender. There was no family history or personal history of the same condition. The patient did not have any systemic disease like diabetes or hypertension. Laboratory investigations such as CBC, hepatic and renal profile were within normal range. Markers for hepatitis B & C, and HIV were also negative. Skin examination revealed solitary, scaly, reddish brownish skin colored papule on auricle of right ear (Fig. 1). Our differential diagnosis including chondrodermatitis nodularis helices, basal cell carcinoma, squamous cell carcinoma, and wart. The lesion was completely excised under local anesthesia. Histopathological examination showed hyperkeratosis, parakeratosis and epidermal acanthosis

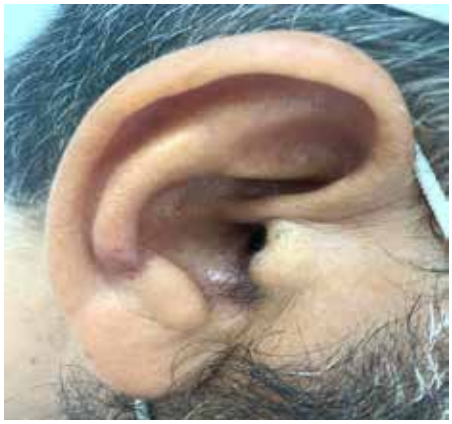

Fig. 1 A solitary scaly Reddish brownish papule on auricle of right ear.

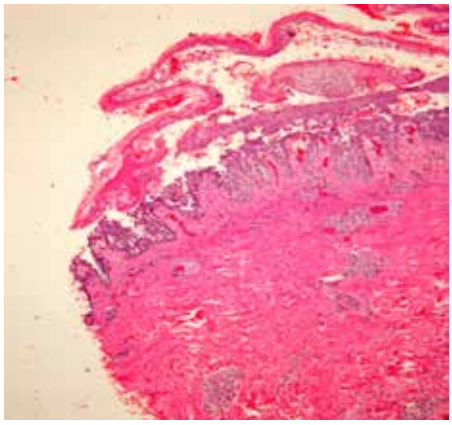

Fig. 2 Hyperkeratosis, parakeratosis, and acanthosis with interaepidermal clefts with elongation of rete ridges.

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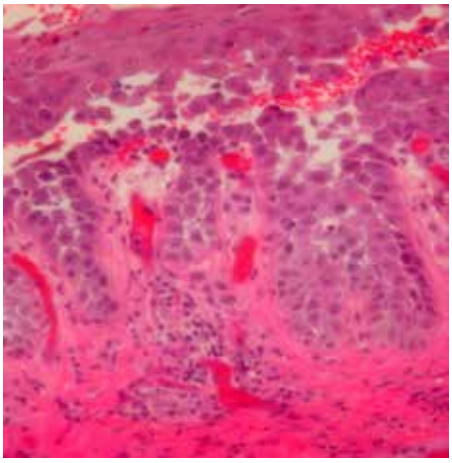

Fig. 3 Suprabasal and interaepidermal clefts with diffuse acantholysis and few areas of dyskeratosis.

with suprabasal and interaepidermal clefts. Numerous acantholytic cells and few dyskeratotic cells. The dermis showed superficial perivascular inflammatory infilterate formed of lymphohistiocytic admixed with melanophages. (Fig.2,3) The patient was followed up for 8 months with no recurrence.

DISCUSSION

Acantholytic dyskeratotic acanthoma (ADA) is a rare benign skin disorder and it requires histopatholgical examination to confirm its diagnosis.[1] Acantholysis and dyskeratosis are microscopically present in ADA, which is an uncommon variation of epidermal acanthoma.[3] Suprabasilar clefting with acantholytic and dyskeratotic cells at all layers of the epidermis characterizes acantholytic dyskeratosis, a histological response pattern. It's also a subset of the vesiculobullous tissue reaction, however the vesiculation isn't the same as the vesiculobullous tissue reaction. However, clinically, the vesiculation is rarely visible. The tonofilament desmosome complex is defective, resulting in disorganized epidermal maturation as the major defect. Darier's disease, Grover's disease, warty dyskeratoma, acantholytic solar keratosis, and vulval and anal acantholytic dyskeratosis are some of the disorders where acantholytic dyskeratosis can be seen. Focal acantholytic dyskeratosis is the medical term for it when it occurs incidentaly.[4]

A solitary lesion with cup-shaped, cystic, or nodular architecture with symptoms indicative of a follicular adnexal lesion is referred to as a warty dyskeratoma.[5] A separate histologic entity has been defined for solitary, nongenital lesions with acantholysis and dyskeratosis but no cup-shaped architecture or follicular involvement: ADA.[3,6]

Multiple lesions on the vulva, perianal area, or penis are referred to as "papular acantholytic dyskeratoma" and the lip has also been observed to have a clinically visible lesion. A case described as "congenital acantholytic dyskeratotic dermatosis" appears to be a papular acantholytic dyskeratosis variation. Since birth, this patient has had several erosive papules and plaques on the left leg, left ankle, and right neck. There was no history of Darier's disease in the patient's family. There were no genetic investigations carried out. Atypical melanocytic lesions had a higher incidence of focal acantholytic dyskeratosis.[4]

Nests of cells with acantholytic dyskeratosis are seen on histopathology of the lesion. In comparison to Darier's disease, hyperkeratosis is less visible in incidental lesions. The more pronounced villi, clefting, and corps ronds distinguish warty dyskeratomas from focal acantholytic dyskeratomas. With considerable acantholysis and minor dyskeratosis, several of the genital and crural cases discussed above exhibit a histological resemblance to Hailey–Hailey disease. They are part of the acantholytic subset of acantholytic dyskeratosis,

while another subset only has dyskeratosis. Immunofluorescence revealed intercellular IgG and C3 within the epidermis in one case of papular acantholytic dyskeratosis in the anogenital region.[4] Our case showed marked acantholysis with few dyskeratotic cells.

Acantholysis with little or no dyskeratosis can be noticed as a coincidental occurrence or as a solitary skin tumor called acantholytic acanthoma. With significant acantholysis and little or no dyskeratosis, a large proportion of the rare vaginal, crural, and perineal cases referred to as papular acantholytic dyskeratosis had a histological resemblance to Hailey–Hailey disease. The term "acantholytic dermatosis of the genitocrural/perineal region" seems to be acceptable for these patients.[4]

In a limited percentage of seborrheic keratosis cases, acantholysis has been found. In seborrheic keratosis, degenerative alterations combined with spongiosis cause acantholysis, which is more common in the irritated variety. The acantholysis is seen between and around the horn cysts in the upper region of the epidermis. Seborrheic keratosis does not have prominent dyskeratosis.[7] Papillomatosis and irregular acanthosis are not as evident in ADA as they are in seborrheic keratosis. These characteristics aid in distinguishing acantholytic seborrheic keratosis from ADA.[8,9]

ADA can present with different clinical features including keratotic yellow plaque on medial malleolus, erythematous pruritic papules and vesicles on face, white brownish scaly papules in calf region, erosive plaque under breast, red to purpuric plaque on face, umblicated papules on chest wall, reddish brown scaly papules on lower back, blackish verrucous plaques on thigh, subungual hyperkeratosis, erythronychia and onycholysis.[1,2,5,10-15]

The trunk (50%) was the most common site, followed by the extremities (25%) and the face (5%). The lesions were either asymptomatic or itchy. The lesions appeared as a red-brown, blackish, yellow plaque or papule in most cases. The lifetime of the lesions ranged from one week to sixty years. Three of the eight patients had received immunosuppressive drugs following organ transplant surgery, and one of the publications postulated that the immunosuppression required for organ transplant recipients may have prevented the eradication of the cell clones that cause this unusual benign lesion.[1] They were either asymptomatic or had serous exudation and discomfort. Onycholysis, hyperkeratosis, and erythronychia were the most common clinical signs.[5,15] our patient presented with asymptomatic reddish brownish lesion in auricle of his right ear.

Up to now, there is no World Health Organization statement or agreement in explicative dermatopathology that ADA is a separate entity. Indeed, acantholytic dyskeratosis is presented as a phenomenon rather than a distinct entity in Lever's histology textbook.[7] The authors of the most recent edition of Weedon's Skin Pathology, on the other hand, believe cutaneous ADA to be a distinct entity that frequently presents clinically with basal cell carcinoma-like characteristics.[4] Many publications suggested that the (ADA) should be managed as a separate entity.[2]

In conclusion, ADA is uncommon benign lesion that usually appears as a papule or plaque. Onycholysis and erythronychia are signs of subungual instances. Our case is a 52-year-old man who was complaining of a solitary redish

brownish papule on the auricle of his right ear. We observed that ADA affects people of all ages, and the predominant histopathological findings are diffuse intraepidermal acantholysis and few dyskeratosis, as well as minor epidermal characteristics. It is regarded as a clinically and pathologically separate entity that requires further studies to explain the exact etiopathogenesis of ADA.

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