Acantholytic dyskeratotic acanthoma: A rare and underappreciated entity

A 64-year-old man presented with a slightly painful plaque on the neck for 6–7 months [Figure 1a]. He had no other underlying disease except osteoarthritis and denied any family history of skin disease. Clinical examination revealed a 1.4 cm-sized erythematous-to-brownish crusted plaque on the neck. Dermoscopy findings showed a central mass of keratin surrounded by linear-irregular vessels [Figure 1b].

An excisional biopsy was performed for treatment and diagnosis under suspicion of inflamed seborrheic keratosis, squamous cell carcinoma and basal cell carcinoma. Histopathological findings showed acanthosis, parakeratosis, dyskeratosis and acantholysis involving a full epidermal layer. Superficial perivascular lymphocytic infiltration in the dermis was present without follicular involvement [Figures 2a and 2b]. After excision, the lesion healed without a sign of recurrence.

Question

What is your Diagnosis?

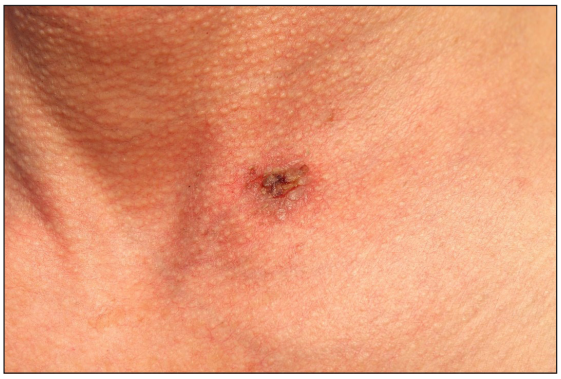

Figure 1a: A solitary, 1.4 cm-sized, erythematous to brownish plaque with crusts on the neck

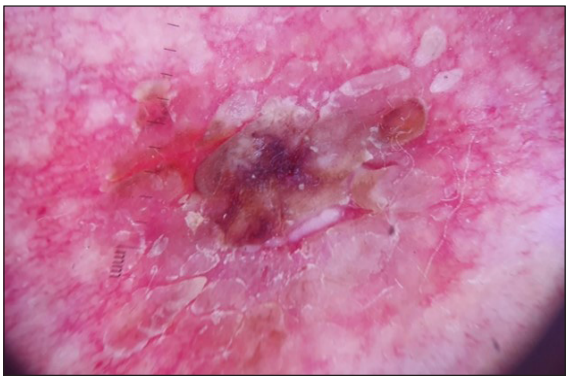

Figure 1b: Dermoscopy showed a central mass of keratin surrounded by linear-irregular vessels

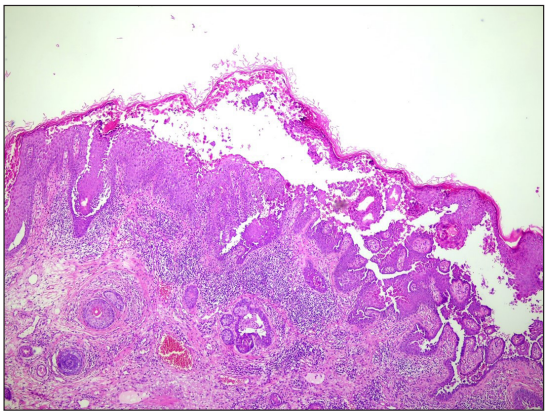

Figure 2a: Histopathological findings showed acanthosis and dyskeratosis with full epidermal acantholysis in the epidermis. Superficial perivascular lymphocytic infiltration in the dermis is also observed (H & E, original magnification x40)

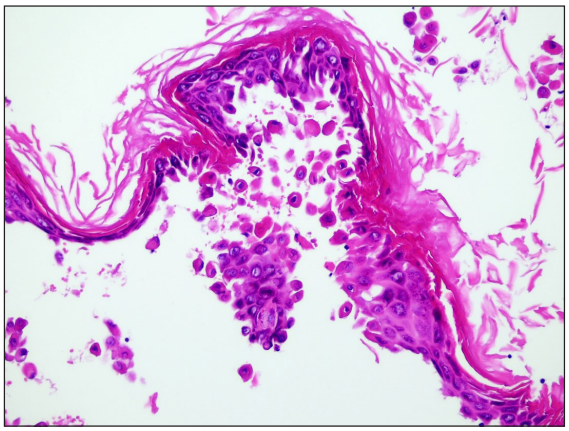

Figure 2b: Rounded eosinophilic dyskeratotic cells (corps ronds) and hyperkeratotic and flattened parakeratotic cells (grains) in the cornified layer are also observed (H & E, x200)

How to cite this article: Doh JY, Lee JH, Bang CH. Acantholytic dyskeratotic acanthoma: A rare and underappreciated entity. Indian J Dermatol Venereol Leprol 2023;89:904-7.

Received: October, 2022 **Accepted:** January, 2023 **EPub Ahead of Print:** April, 2023 **Published:** October, 2023

DOI: 10.25259/IJDVL_970_2022 **PMID:** 37317765

Answer

Diagnosis: Acantholytic dyskeratotic acanthoma

Discussion

Acanthoma is a benign tumor of epidermal keratinocytes, showing a broad range of histological patterns.[1] Acantholytic dyskeratotic acanthoma is a relatively uncommon variant of acanthoma, classified within the past decade. The term 'focal acantholytic dyskeratosis' was first used by Ackerman to describe incidental lesions showing acantholysis and dyskeratosis.[2] In contrast, genital lesions with similar characteristics had been described as 'papular acantholytic dyskeratoma' or 'papular acantholytic dyskeratosis'.[3,4] However, non-genital lesions showing acantholysis and dyskeratosis were not adequately identified until Omulecki et al. defined acantholytic dyskeratotic acanthoma for the first time in 2007.[1] Acantholytic dyskeratotic acanthoma refers to a solitary, non-genital lesion with prominent acantholysis and dyskeratosis without cup-shaped architecture or follicular involvement.[4,6,7] However, whether it is an actual distinct diagnostic entity is debatable. There is no statement from the World Health Organization, and the description of acantholytic dyskeratotic acanthoma differs even within textbooks. Lever's histopathology and McKee's pathology textbooks describe acantholytic dyskeratotic acanthoma as a phenomenon rather than a separate entity,[8,9] while Weedon's textbook considers acantholytic dyskeratotic acanthoma a separate entity.[10]

About 43 cases of acantholytic dyskeratotic acanthoma have been reported in the English literature to date [Table 1]. Patient ages ranged from 12 to 97 years, with 38 (88.4%) older than 40 years. The patients were often clinically suspected of having basal cell carcinoma, squamous cell carcinoma, and/or actinic keratosis. Most lesions were located on the trunk and often presented as a small papule of <1 cm. Among the 43 reported cases, three were in subungual

Table 1: Clinical characteristics of acantholytic dyskeratotic acanthoma cases reported in the English literature

Year	Author	Age	Gender	Number/size (mm)	Site	Clinical diagnosis	Duration	Medical history
2007	Omulecki *et al*.[1]	64	Male	1/20 x 50	Back	(a)	7 years	DM
2008	Ko *et al*.[4]	48	Female	1/7	Ankle	BCC	(b)	(a)
		40	Female	1/5	Forearm	AK/SCC	(b)	(a)
		45	Female	1/4	Back	(a)	(b)	(a)
		64	Female	1/4	Back	BCC	(b)	(a)
		48	Female	1/5	Abdomen	BCC/SCC	(b)	(a)
		49	Female	1/6	Chest	BCC/SCC	(b)	(a)
		43	Female	1/3	Chest	BCC	(b)	(a)
		63	Male	1/7	Back	BCC/BD	(b)	(a)
		50	Female	1/4	Chest	SK	(b)	(a)
		39	Female	1/7	Back	Wart	(b)	(a)
		52	Female	1/6	Back	SCC	(b)	(a)
		43	Male	1/5	Abdomen	BCC	(b)	(a)
		45	Female	1/4	Abdomen	SK/AK	(b)	(a)
		49	Female	1/5	Nevus	BCC	(b)	(a)
		57	Male	1/2	Chest	BCC/AK	(b)	(a)
		63	Male	1/5	Clavicle	AK/BCC/SCC	(b)	(a)
		84	Male	1/9	Thigh	BD	(b)	(a)
		48	Female	1/4	Flank	SK/AK	(b)	(a)
		58	Male	1/7	Sternum	BCC	(b)	(a)
		60	Male	1/3	Back	BCC	(b)	(a)
		68	Male	1/6	Flank	SCC	(b)	(a)
		57	Female	1/6	Chest	Papilloma	(b)	(a)
		51	Female	1/5	Shoulder	BCC	(b)	(a)
		64	Male	1/3	Chest	Nevus	(b)	(a)
		75	Male	1/5	Lower leg	BCC	(b)	(a)

(Contd...)

Table 1: *Contd...*

Year	Author	Age	Gender	Number/size (mm)	Site	Clinical diagnosis	Duration	Medical history
		76	Female	1/5	Neck	SCC	(b)	(a)
		47	Male	1/7	Back	Nevus	(b)	(a)
		56	Female	1/5	Chest	BCC	(b)	(a)
2009	Sass et al.[15]	53	Male	1/5	Right thumbnail	(a)	(b)	(a)
		15	Male	1/3	Right thumbnail	Onychopapilloma	6 months	(a)
		12	Female	(a)	Right thumbnail	Onychopapilloma	9 months	(a)
2013	Park et al.[16]	42	Female	Multiple/2-3	Face	(a)	Several years(b)	Associated with rosacea
2013	Pezzolo et al.[12]	49	Male	1/4 x 4	Lower leg	BCC/AK	3 years	Kidney allograft patient on immunosuppression
2014	Goldenberg et al.[3]	72	Female	1/10 x 4	Chest	(a)	3 months	No other illness
2017	Kim et al.[17]	38	Female	1/(a)	Face	(a)	1 month	Associated with DLE
2018	Burgler et al.[13]	60	Male	Multiple/(a)	Back and lateral chest wall	(a)	1 week	Heart transplant patient on immunosuppression
2019	Kanitakis et al.[5]	74	Male	Multiple/(a)	Back	(a)	(b)	Liver transplant patient on immunosuppression
2020	Nandakumar et al.[6]	75	Female	2/13 x 15, 10 x 12	Thigh	Chromoblastomycosis, SCC, tuberculosis verrucosa cutis, viral wart, lupus vulgaris	60 years	No other illness
2017	Komori et al.[14]	62	Female	1/(a)	Right shoulder	SCC	(b)	Metastatic melanoma on vemurafenib
2017	Ng et al.[18]	25	Male	1/(a)	Right thumbnail	Onychopapilloma, onychomatricoma or a verruca	1 year	No other illness
2021	Tanaka et al.[19]	97	Female	1/10	Right thigh	Verruca vulgaris	(b)	(a)
2022	Current case	64	Male	1/14	Neck	Inflamed SK, SCC, BCC	6–7 months	No other illness

a): Not available, b): exact duration, not given, AK: actinic keratosis, BCC: basal cell carcinoma, BD: Bowen's disease, DLE: discoid lupus erythematosus, DM: diabetes mellitus, SCC: squamous cell carcinoma, SK: seborrheic keratosis

areas,[13–15] three occurred in a transplant setting[5,12,13] and one after vemurafenib treatment.[14] Among the three patients with a history of transplants, two presented with multiple lesions. However, the association between immunosuppression and acantholytic dyskeratotic acanthoma remains inconclusive.

Pathogenesis of acantholytic dyskeratotic acanthoma is still elusive. However, genetic and immunological factors along with viral infections, physical stimuli, and sunlight exposure, might play a role.[3,10] Histopathologically, acantholytic dyskeratotic acanthoma is characterised by acantholysis and dyskeratosis.[3,6] Therefore, it should be differentiated from other acantholytic disorders such as keratosis follicularis (Darier's disease), transient acantholytic dermatosis (Grover's disease), Hailey-Hailey disease, and warty dyskeratoma. As Darier's, Grover's and Hailey-Hailey's diseases show distinct clinical features and immunofluorescence findings, they could be easily differentiated from acantholytic dyskeratotic acanthoma.[9] On the contrary, it could be challenging to distinguish acantholytic dyskeratotic acanthoma from warty dyskeratoma as they share many histological features. However, unlike acantholytic dyskeratotic acanthoma, warty dyskeratoma is associated with a cup-shaped invagination or follicular involvement.[1,3,4,6,11] Last, acantholytic dyskeratotic acanthoma can also be easily differentiated from acantholytic acnathoma by the presence of prominent dyskeratosis.

Clinically, patients with acantholytic dyskeratotic acanthoma typically present with an asymptomatic solitary keratotic papule or plaque showing predilection to the trunk,[4,5] and resemble non-melanotic skin cancers such as squamous cell carcinoma, basal cell carcinoma or actinic keratosis.[4,5] However, acantholytic dyskeratotic acanthoma is a benign disease that can be easily treated through excision. Therefore, it is necessary to rule out acantholytic dyskeratotic acanthoma from other conditions to avoid unnecessary procedures and treatments. It can be relatively easily differentiated from other non-melanotic skin cancers histopathologically as it lacks cellular atypia and specific morphological features of those conditions.[1,3]

Our case reinforces the necessity of classifying acantholytic dyskeratotic acanthoma as a distinct clinical entity. Examining and ruling out acantholytic dyskeratotic acanthoma as a clinical entity will assist in avoiding unnecessary or invasive

procedures and treatments associated with another malignant disease with similar characteristics.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil

Conflict of interest

There is no conflict of interest

Jee Yun Doh, Ji Hyun Lee, Chul Hwan Bang

Department of Dermatology, College of Medicine, Seoul St. Mary's Hospital, The Catholic University of Korea, Seoul, South Korea

Corresponding author:
Prof. Chul Hwan Bang,
Department of Dermatology, College of Medicine, Seoul St. Mary's Hospital, The Catholic University of Korea, Seoul, South Korea,
mrbangga@gmail.com

References

1. Omulecki A, Lesiak A, Narbutt J, Woźniacka A, Piekarski J, Biernat W. Plaque form of warty dyskeratoma–acantholytic dyskeratotic acanthoma. J Cutan Pathol 2007;34:494–6.
2. Ackerman AB. Focal acantholytic dyskeratosis. Arch Dermatol 1972;106:702–6.
3. Goldenberg A, Lee RA, Cohen PR. Acantholytic dyskeratotic acanthoma: Case report and review of the literature. Dermatol Pract Concept 2014;4:25–30.
4. Ko CJ, Barr RJ, Subtil A, McNiff JM. Acantholytic dyskeratotic acanthoma: A variant of a benign keratosis. J Cutan Pathol 2008;35:298–1.
5. Kanitakis J, Gouillon L, Jullien D, Ducroux E. Multiple acantholytic dyskeratotic acanthomas in a liver-transplant recipient. Dermatol Online J 2019;25:4.
6. Nair NG, Athira B. Acantholytic dyskeratotic acanthoma: A rare clinicopathological entity–a case report and review of literature. J Skin Sex Transmit Dis 2020;2:115–8.
7. Brownstein MH. Acantholytic acanthoma. J Am Acad Dermatol 1988;19:783–6.
8. Elder DE. Lever's histopathology of the skin. 11th ed. Wolters Kluwer; 2015.
9. Calonje E, Brenn T, Lazar A, McKee PH. McKee's pathology of the skin: With clinical correlations:Elsevier/Saunders; 2012.
10. Patterson JW. Weedon's skin pathology. 5th ed. Elsevier; 2021.
11. Carlioz V, Balme B, Depaepe L, Thomas L. Dermoscopy of subungual acantholytic dyskeratotic acanthoma. Clin Exp Dermatol 2020;45:355–6.
12. Pezzolo E, Colato C, Del Giglio M, Tessari G, Girolomoni G. Acantholytic dyskeratotic acanthoma in an immunosuppressed patient: A case report with review of the literature. Clin Dermatol 2013;1:149–52.
13. Bürgler C, Schlapbach C, Feldmeyer L, Haneke E. Multiple acantholytic dyskeratotic acanthomas in an organ transplant recipient. JAAD Case Rep 2018;4:695–7.
14. Komori T, Otsuka A, Kaku Y, Honda T, Kabashima K. Acantholytic dyskeratotic acanthoma: A possible skin adverse event of vemurafenib treatment. J Eur Acad Dermatol Venereol 2017;31:e474–5.
15. Sass U, Kolivras A, Richert B, Moulonguet I, Goettmann-Bonvallot S, Anseeuw M, *et al*. Acantholytic tumor of the nail: Acantholytic dyskeratotic acanthoma. J Cutan Pathol 2009;36 :1308–11.
16. DiMaio DJ, Cohen PR. Incidental focal acantholytic dyskeratosis. J Am Acad Dermatol 1998;38:243–7.
17. Kim HR, Lim S-K, Lee H-E, Im M, Lee Y, Seo YJ, *et al*. Incidental Focal Acantholytic Dyskeratosis in a Patient with Discoid Lupus Erythematosus: A Possible Role for SPCA1 in the Pathogenesis of the Disease. Ann Dermatol 2017;29:655–7.
18. Ng J, Harvey NT, von Nida J, Wood BA. Subungual acantholytic dyskeratotic acanthoma: An unusual cause of longitudinal erythronychia. Pathology 2018;50:482–4.
19. Tanaka C, Kurabayashi A, Furihata K, Iwashita W, Naganuma S, Toi M, *et al*. Cutaneous acantholytic dyskeratotic acanthoma accompanying syringofibroadenomatous hyperplasia with proliferation of mature sebocytes: A case report. J Cutan Pathol 2021;48:451–4.