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Incidental Focal Acantholytic Dyskeratosis in a Patient with Discoid Lupus Erythematosus: A Possible Role for SPCA1 in the Pathogenesis of the Disease

Hyeong-Rae Kim, Seul-Ki Lim, Hae-Eul Lee, Myung Im, Young Lee, Young-Joon Seo, Jeung-Hoon Lee

Department of Dermatology, Chungnam National University School of Medicine, Daejeon, Korea

Dear Editor:

Incidental focal acantholytic dyskeratosis (IFAD) is a rare incidental histopathological condition characterized by suprabasilar clefts, acantholytic and dyskeratotic epidermal cells, hyperkeratosis, and parakeratosis. Various etiological factors including ultraviolet radiation, hormones, and viral infection have been suggested[1]. However, the precise etiology of the condition remains unknown. We present the first case of IFAD associated with discoid lupus erythematosus (DLE) and suggest a possible role for the secretory pathway Ca^{2+} ATPase1 (SPCA1) in the disease.

A 38-year-old female presented with a 1-month history of an enlarging, erythematous, pruritic facial plaque (Fig. 1). Laboratory test data including a complete blood count, a liver function test, and measurements of fluorescent anti-nuclear and anti-double-strand DNA antibodies were all normal. Histological examination revealed epidermal atrophy, follicular plugging, loss of the rete ridge pattern, vacuolar degeneration of basal keratinocytes, slight thickening of basement membrane and perivascular and perifollicular infiltration of lymphohistiocytes, compatible with a diagnosis of DLE (Fig. 2A). In addition, a suprabasilar cleft and acantholytic and dyskeratotic cells were also noted (Fig. 2B). The patient was diagnosed with DLE with IFAD and prescribed 0.1% tacrolimus ointment. The lesion had improved moderately at the 1-month follow-up. Patient tissue samples were subjected to further immunohistochemical study, using normal skin from another patient as a control; the tissues were stained with antibodies against the SPCA1 (bs-2434R; Bioss, Woburn, MA, USA) and the sarco/endoplasmic reticulum Ca^{2+} ATPAase2 (SERCA2) (sc-30110; Santa Cruz Biotech, Dallas, TX, USA). SERCA2 staining was of similar intensity in both tissue samples (Fig. 2C, D). SPCA1 staining of IFAD tissue was less intense than that of normal tissue (Fig. 2E, F).

In 1972, Ackerman[2] first described the distinctive histological features of FAD. Changes which had been considered unique to Darier's disease (DD) were also found in a variety of other conditions. FAD presented as either single or multiple lesions. The single lesions were divided into

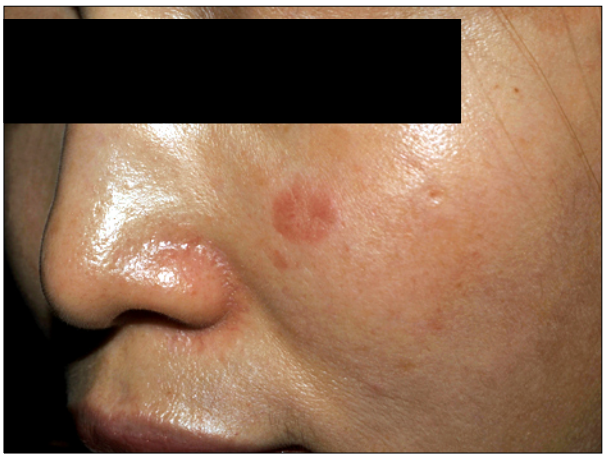

Fig. 1. A slightly elevated, well-demarcated, erythematous round plaque approximately 1 cm in diameter.

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Corresponding author: Jeung-Hoon Lee, Department of Dermatology, Chungnam National University School of Medicine, 266 Munhwa-ro, Jung-gu, Daejeon 35015, Korea. Tel: 82-42-280-7707, Fax: 82-42-280-8459, E-mail: Jhoon@cnu.ac.kr

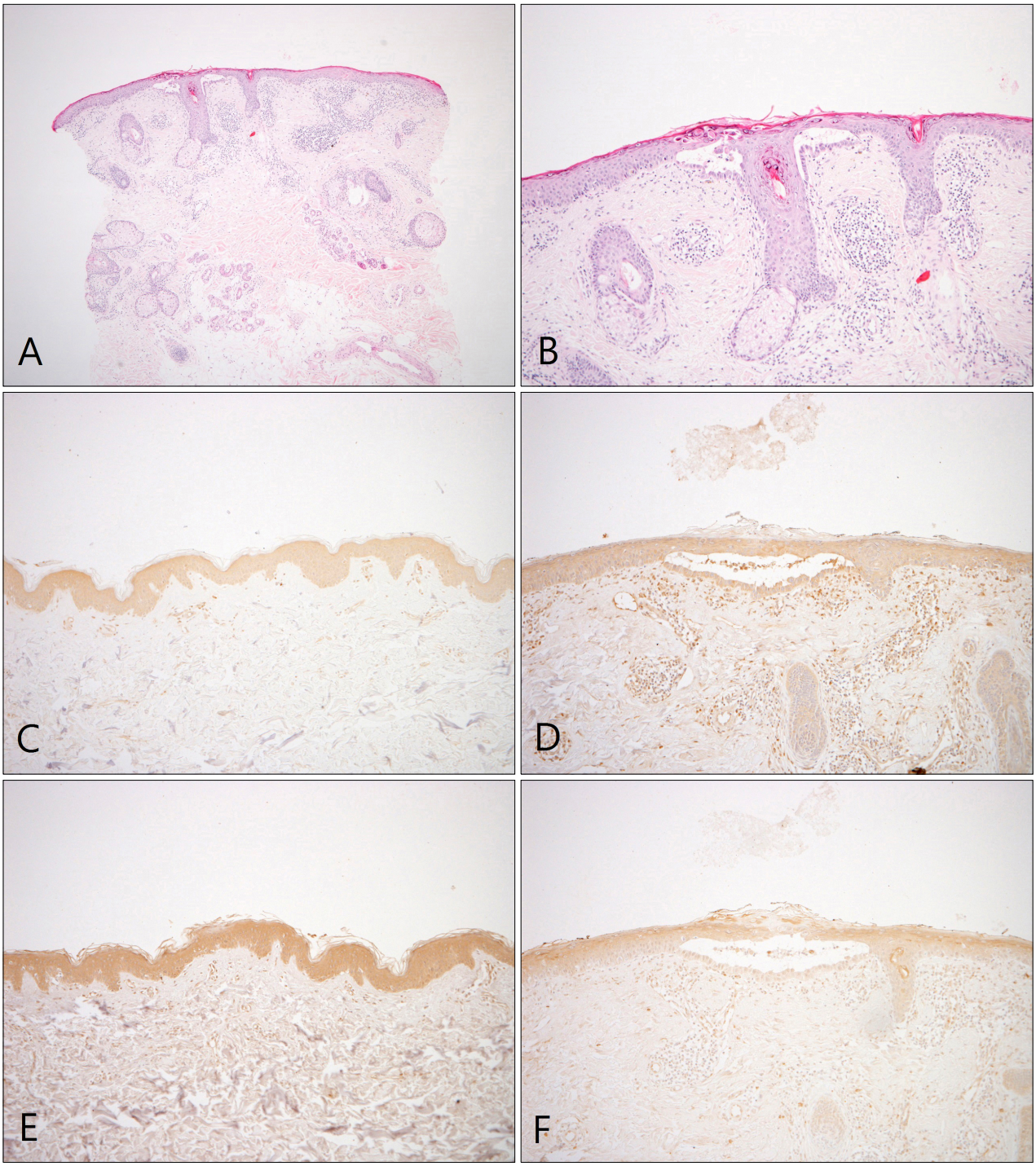

Fig. 2. (A) Epidermal atrophy, loss of the rete ridge pattern, vacuolar degeneration of basal keratinocytes, and perivascular and perifollicular lymphohistiocyte infiltration, were evident (H&E, ×40). (B) A suprabasilar cleft consisting of acantholytic and dyskeratotic cells was observed (H&E, ×100). The intensity of SERCA2 staining was similar in normal (C) and incidental focal acantholytic dyskeratosis (IFAD) tissue (D). The SPCA1 staining intensity was reduced in IFAD epidermal tissue (F) compared to normal tissue (E) (C, E: normal tissue, D, F: IFAD tissue; ×100).

three types: incidental, papular, and nodular[1,2]. Various lesions are associated with IFAD; these include epthelial tumors, fibrohistocytic, inflammatory and melanocytic lesions[1].

Mutation in *ATP2A2* encoding SERCA2 and in *ATP2C1* encoding SPCA1 were found in DD and Hailey-Hailey

disease (HHD), respectively[3]. SERCA2 and SPCA1 regulate the Ca^{2+} levels of the endoplasmic reticulum and Golgi apparatus. Translation, translocation, folding, and processing of adhesional molecule of keratinocyte are closely related with Ca^{2+} levels; mutations affecting the Ca^{2+} pumps induce histologically acantholysis and dyskeratosis[3]. An earlier study revealed that SERCA2 and SPCA1 staining was less intense than normal in both DD and HHD skin, but SERCA2 was more affected in DD patients and SPCA1 in those with HHD[4,5]. We found that the SERCA2 staining intensity was similar in both tissues, but SPCA1 expression level was much reduced in IFAD compared to normal tissue. Thus, IFAD seems to be more closely associated with low-level SPCA1 expression.

In summary, to the best of our knowledge, our present IFAD case is the first to be associated with DLE. Although further work is required, our study showed that SPCA1 expression was reduced in IFAD tissue; SPCA1 may thus play a role in disease pathogenesis.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

REFERENCES

1. Park SY, Lee HJ, Shin JY, Ahn SK. Incidental focal acantholytic dyskeratosis in the setting of rosacea. Ann Dermatol 2013;25:518-520.
2. Ackerman AB. Focal acantholytic dyskeratosis. Arch Dermatol 1972;106:702-706.
3. Dhitavat J, Fairclough RJ, Hovnanian A, Burge SM. Calcium pumps and keratinocytes: lessons from Darier's disease and Hailey-Hailey disease. Br J Dermatol 2004;150:821-828.
4. Porgpermdee S, Yu X, Takagi A, Mayuzumi N, Ogawa H, Ikeda S. Expression of SPCA1 (Hailey-Hailey disease gene product) in acantholytic dermatoses. J Dermatol Sci 2005;40:137-140.
5. Porgpermdee S, Takagi A, Mayuzumi N, Ogawa H, Ikeda S, Nakamura S. Expression of SERCA2 (Darier's disease gene product) in acantholytic dermatoses. J Dermatol Sci 2006;43:146-149.

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A Case of Dermatosis Neglecta Caused by an Inappropriate Habit of Applying a Moisturizer

Jungyoon Moon, Hyun-sun Park, Hyun-Sun Yoon, Soyun Cho

Department of Dermatology, SMG-SNU Boramae Medical Center, Seoul, Korea

Dear Editor:

Dermatosis neglecta (DN) is characterized by localized hyperkeratinization as a consequence of avoiding proper washing of the affected areas[1]. It is more common in patients with physical disability, neurological deficit, or psychiatric illness who are likely to lack cleanliness and also

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Corresponding author: Soyun Cho, Department of Dermatology, SMG-SNU Boramae Medical Center, 20 Boramae-ro 5-gil, Dongjak-gu, Seoul 07061, Korea. Tel: 82-2-870-2381, Fax: 82-2-870-3866, E-mail: sycho@snu.ac.kr