A Case of Acantholytic Dyskeratotic Epidermal Nevus

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We report a case of acantholytic dyskeratotic epidermal nevus in a 28-year-old female. The patient showed unilateral, asymptomatic, grouped, 2 mm sized, brownish, follicular and non-follicular, keratotic papules disposed along Blaschko's lines on the right side of the back, flank and abdomen. She had had this condition for 4 years. The biopsy specimen obtained from the abdomen showed hyperkeratosis, parakeratosis, acanthosis, acantholytic suprabasal clefts, corps ronds, and grains. Although cryotherapy with liquid nitrogen was performed on half of the lesions, the whole skin lesions healed with focal hypertrophic scars 4 weeks later. There was no recurrence after a 12 months follow-up period. To our knowledge, this is the first reported case of acantholytic dyskeratotic epidermal nevus in Korean dermatologic literature. (Ann Dermatol 9:(1)8~11, 1997).

Key Words : Acantholytic Dyskeratotic Epidermal Nevus, Cryotherapy

Linear epidermal nevus(LEN) is a developmental disorder characterized by hyperplasia of the surface or adnexal epithelium. It consists of closely-set, verrucous papules arranged in a linear distribution on the body, which may be either localized or systematized[1]. Usually it is present at birth, and may enlarge slowly during childhood. By adolescence, the lesions reach a stable size and further extension is unlikely. Nearly all cases of the localized type of LEN show considerable hyperkeratosis, papillomatosis, and acanthosis. However, in some instances, histological examination reveals features of acantholytic dyskeratosis as seen in Darier's disease. Darier's disease(keratosis follicularis) is characterized by extensive, persistent, progressive, hyperkeratotic or crusted, papules on seborrheic areas. About 10% of all cases of Darier's disease skin lesions may show unilateral, linear distributions like a linear epidermal nevus[2].

Several authors[3-7] reported that patients with hyperkeratotic papules distributed in a unilateral, linear or zosteriform pattern disclosed histological features of dyskeratotic acantholysis. Although classification of these cases as a variant of Darier's disease or a linear epidermal nevus is still a point of discussion, these cases may be considered as an acantholytic dyskeratotic epidermal nevus (AD EN). Herein, we report a case of linear epidermal nevus with acantholytic dyskeratosis. This is probably the first case reported in Korean dermatologic literature.

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REPORT

A 28-year-old woman had had asymptomatic band-like papular eruptions on the right side of her body for 4 years. A physical examination revealed numerous, small brown papules, which were found unilaterally over the right side of her back, flank and abdomen along Blaschko lines. The papules ranged from 1 to 3 mm in diameter; they were both follicular and nonfollicular. There were no other abnormalities of the nail, skin or oral mucosa. Sunlight, hot weather and perspiration did not aggravate the condition. This patient denied having a family history of a similar condition. Her past medical history was insignificant.

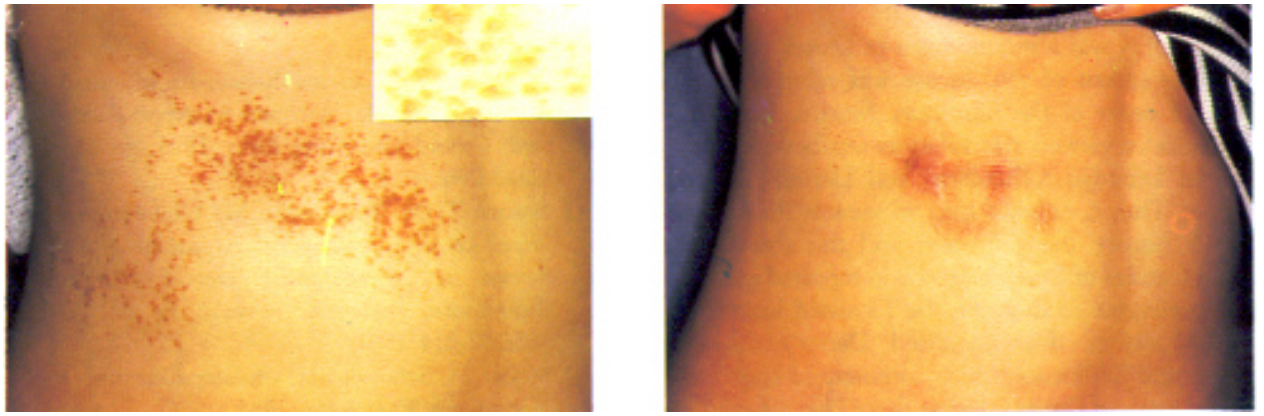

Fig. 1. (A) Multiple pinhead-sized brownish papules following the lines of Blaschko on the right abdomen (B) Heal with focal hypertrophic scars at 12 months follow-up.

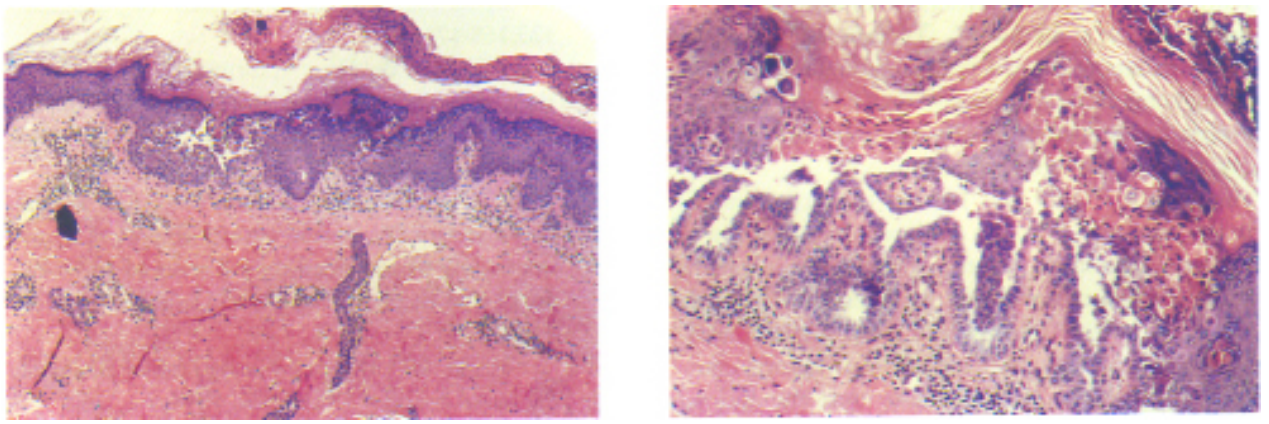

Fig. 2. (A) Biopsy specimen showing hyperkeratosis, irregular acanthosis and suprabasal acantholysis(H&E, ×40). (B) Suprabasal acantholysis, corps ronds and grains(H&E, ×200).

A biopsy specimen from the abdomen showed acanthosis, papillomatosis, hyperkeratosis with focal parakeratosis, hypergranulosis, acantholytic suprabasal clefts, dyskeratotic cells with corps ronds and grains in the epidermis, and a scanty perivascular lymphocytic infiltrate in the papillary dermis. The biopsy findings were consistent with Grover's disease, Darier's disease and ADEN. Differential diagnosis clinically included LEN, Darier's disease, and ADEN. We diagnosed this case as ADEN because of the different clinical finding from that of Darier's disease and the different histological finding from that of LEN. Steroid ointment was initially applied to the lesions without success. Cryotherapy with liquid nitrogen was performed on the half of the lesions. After 4 weeks, the whole skin lesions healed with focal hypertrophic scars. And there was no recurrence after a 12 months follow-up period.

DISCUSSION

Acantholytic dyskeratotic epidermal nevus (ADEN) was originally described as a 'forme fruste' of Darier's disease[8]. These linear lesions may be present since birth or infancy, but in most cases, they arise at the age of 20 or above. It is an uncommon disorder with a delayed onset, which is rather late for epidermal nevi as well as for Darier's disease. Sunlight, PUVA and hot moist weather may provoke the lesions[3,9]. Furthermore it may improve considerably with topical vitamin A acid[10].

Munro and Cox[3] described a patient with unilateral nevoid lesions typical of Darier's disease, accompanied by other features of this disorder on the same side of the body. They suggested that at least some ADEN are indeed a localized form of keratosis follicularis. Cambiaghi et al[6] thought

that ADEN was a mosaic form of Darier's disease, and the extent of the disease was related to the time of onset of the mutation. Starink and Woerdeman[3] reviewed the relationship between 'localized keratosis follicularis' and Darier's disease proper. They concluded that, in view of the lack of family history, delayed onset and the absence of other features of Darier's disease in reported cases, it was preferable to describe the localized lesions as ADEN rather than localized Darier's disease. Burge and Wilkinson[11] also commented that unilateral forms of Darier's disease should be classified as ADEN rather than localized Darier's disease.

Retinoids are known to be beneficial in the treatment of several disorders of keratinization inducing Darier's disease[12-14]. Topical tretinoin, as previously reported in a few cases, is beneficial for localized keratosis follicularis and may induce complete remissions. However, its effect usually lasts only as long as the treatment is maintained[4].

Generally ADEN was known to persist indefinitely. However, Thomas et al[7] reported that linear keratosis follicularis treated with tretinoin cream healed completely without recurrence. Although only half of the lesions had been treated with cryotherapy in this case, all the skin lesions completely healed. So, we speculate that the lesions might heal spontaneously. Therefore, we suggested that ADEN is probably a separate clinical entity from Darier's disease or LEN. ADEN has different clinical findings from Darier's disease; the delayed onset of the lesions, distribution of lesions, spontaneous recovery and absence of inheritance. ADEN, unlike LEN proper which shows hyperkeratosis, papillomatosis, acanthosis, and granular degeneration of epidermis, reveals features of acanthotic dyskeratosis as seen in Darier's disease. Therefore, we think this case is not Darier's disease and LEN proper but ADEN.

This is the first reported case of ADEN in Korean dermatologic literature.

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