

# A Variant of Epidermal Nevus

## — Acrosyringal Epidermal Nevus ? —

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A 17-year-old male was presented with pruritic grouped erythematous papules on the face, chest, upper back, both axillae, and genital area.

The histologic picture of a benign papilloma and spongiosis around the acrosyringium were prominent findings.

The term acrosyringal epidermal nevus is proposed for this histological picture, and some explanations for its pathogenesis are suggested. (*Ann Dermatol* 3:(1) 45–48, 1991)

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*Key Words:* Acrosyringal epidermal nevus

Epidermal nevi are organoid nevi arising from the pluripotential germinative cells in the basal layer of the embryonic epidermis. These nevi have often been classified according to their predominant component. Consequently the terms nevus verrucosus (keratinocyte), nevus sebaceous (sebaceous glands), nevus comedonicus (hair follicles), and nevus syringocystadenosus papilliferus (apocrine glands).<sup>1-3</sup>

In 1980, Abell and Read<sup>4</sup> described an epidermal nevus which pathologically demonstrated gross examples of cornoid lamellae associated exclusively with the eccrine duct and ostia. However, the studies on the relationship between epidermal nevi and eccrine glands have not attracted much attention in the literature.

This paper describes a variant of epidermal nevus with a marked localization of histological changes to the acrosyringium.

## REPORT OF A CASE

A 17-year-old male student visited our dermatologic clinic complaining of pruritic grouped, erythematous papules on the face, chest, upper back, both axillae, and genital area. In his early childhood, his parents found discrete erythematous papules on his pubic area. The same lesions appeared on both axillary fossae at age of 10. Thereafter, the skin lesion extended to the face, chest, and upper back, in that order. The skin lesion provoked mild to moderate itching sensation during the course. Various dermatologists made diagnoses such as Darier's disease, Fox-Fordyce disease, inflammatory linear verrucous epidermal nevus, and so forth. Therefore, he was treated with oral antihistamines, topical corticosteroid, keratolytics, systemic etretinate and cryotherapy without any satisfactory response.

On physical examination, grouped erythematous papular eruptions were seen on the face, chest, upper back, both axillae, and genital area (Fig. 1-5). Every lesion was distributed in symmetrical fashion. Laboratory studies, including complete blood cell count, urinalysis, liver function test, 17-hydroxycorticosteroid and 17-keto-

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**Fig. 1.** Discrete erythematous papules on the face



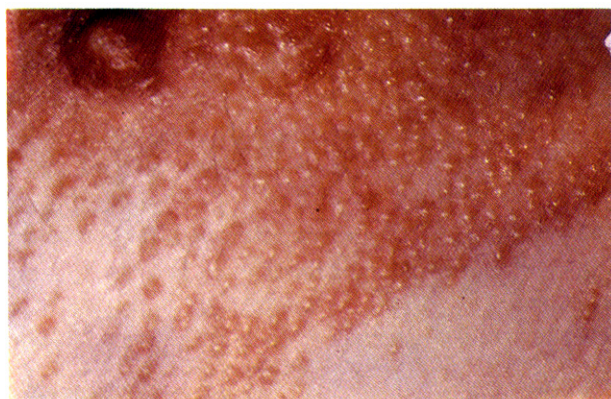
**Fig. 4.** Same lesions on the genital area



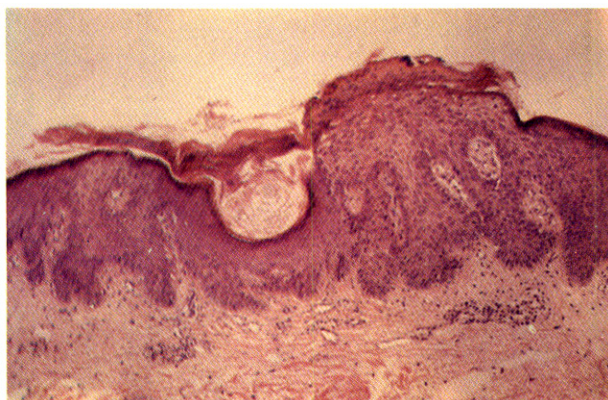
**Fig. 2.** Numerous, Grouped erythematous papules on the chest and both axillae



**Fig. 5.** Close-up view of lesions of the axilla



**Fig. 3.** Close-up view of lesions of the chest

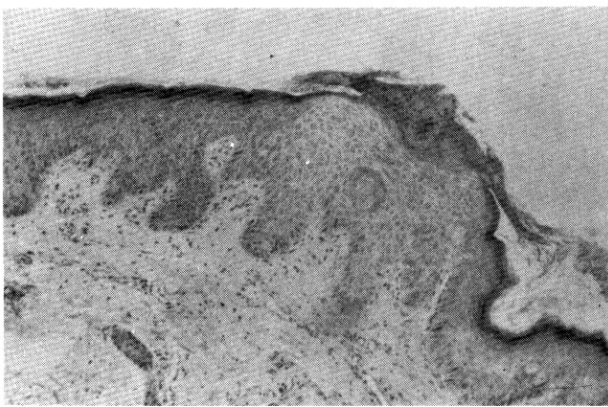


**Fig. 8.** Epidermal changes, same as Fig. 7, is absent in the infundibular portion of vellus hair follicle (H & E stain,  $\times 100$ ).

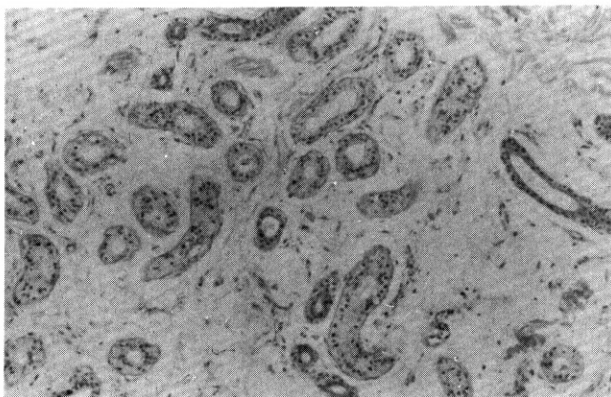




**Fig. 6.** Biopsy specimen taken from the lesion of chest shows papillomatous changes of the epidermis and proliferation of the dermal eccrine ducts (H & E stain,  $\times 40$ ).



**Fig. 7.** High-power view of Fig. 6 shows an irregular acanthosis, hyperkeratosis, parakeratosis, and spongiosis. These findings are distinct around acrosyringium (H & E stain,  $\times 100$ ).



**Fig. 9.** Dermal ducts are dilated and hyperplastic, and wind at the upper or mid dermis (H & E stain,  $\times 400$ ).

steroid of 24 hour-urine collection, were within normal limits.

Biopsy specimens were taken from lesions on the chest, axillae, upper back, and genital area. Histologically, all the skin sections showed considerable hyperkeratosis, parakeratosis, spongiosis, and acanthosis with elongation of the rete ridges. The changes were distinct around the acrosyringium (Fig. 6, 7). These findings were absent in the infundibular portion of vellus hair follicle; in other words, they had skipped over the hair follicles (Fig. 8). In some sections, dilated and hyperplastic eccrine dermal ducts were noted (Fig. 6, 9). Direct immunofluorescence testing showed deposits of IgG at the intercellular space of the spongiotic area.

The patient was treated with oral administration of antihistaminics and 5% urea ointment, but the lesions have persisted.

## DISCUSSION

Epidermal nevi are organoid nevi arising from the basal layer of the embryonic epidermis. They may involve any part of the body surface. Some patients had widespread nevi.<sup>5</sup> It is well known that, although epidermal nevi are often present at birth, they also may develop during childhood, as in this case. Epidermal nevi may extend beyond their original distribution. However, once lesions has reached stability in late adolescence, further progression is unlikely.<sup>5</sup> The lesions in this case also appeared not to have extended.

Epidermal nevi, composed of keratinocytes as a prominent component, have a distinctive histologic picture consisting of hyperkeratosis, papillomatosis, and acanthosis with elongation of the rete ridges.<sup>6</sup> This case had in common hyperkeratosis, parakeratosis, spongiosis, and acanthosis with elongation of the rete ridges, and it was localized exclusively to the acrosyringium. The term "acrosyringeal epidermal nevus" summarizes this histologic picture. There were previous reports which characterized it by localization of histological changes around the acrosyringium.<sup>4, 7, 8</sup> However, the precise mechanism of this phenomenon has not been fully understood.

When speculating upon the pathogenesis of

acrosyringal epidermal nevus, several explanations could be proposed. Eccrine glands may serve as a predominant component in developing epidermal nevi, because epidermal nevi arise from the pluripotential germinative cells which give rise to keratinocytes and to skin appendages. This case also showed eccrine malformations in the form of dilated and hyperplastic eccrine ducts (Fig. 6, 9). Furthermore, our observation was not discovered in any other adnexal structure of the skin (hair infundibulum, apocrine, and sebaceous duct) (Fig. 8). Thus, it appears that acrosyringal epidermal nevus is a variant of epidermal nevus combined with eccrine malformation.

In epidermal nevi, considerable intercellular edema (spongiosis) is rare. In this case, direct immunofluorescence testing showed deposits of IgG at the intercellular space of the spongiotic lesion, but unfortunately, it is not valuable. This observation agrees with those of Wilkinson<sup>9</sup>, who found that the antisweat precipitins (IgG) were present in healthy adults. It is likely that spongiosis is a secondary change from sweating. We did not perform a sweating test in the lesion, but the function of eccrine apparatus in this lesion appeared active.

The predominant tissue of epidermal nevi may vary with the evolution of the lesion, and different areas of the same lesion may show a varie-

ty of components at the same time. Thus, epidermal nevi may show various clinical and histopathologic varieties. More examples of this type of epidermal nevus are needed to make a distinctive clinical entity.

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