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CASE REPORT

A case of unilateral epidermal nevi complicated by atopic dermatitis treated with dupilumab

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ABSTRACT

Epidermal Nevus Syndrome (ENS) is a congenital disorder characterized by skin lesions called epidermal nevi. This condition typically appears at birth or in infancy. However, a case of adult-onset epidermal nevi was reported in a 36-year-old male with a history of atopic dermatitis (AD) and skin abnormalities on the right side of his body. He had been using topical steroids for AD but eventually started dupilumab treatment at age 41. The treatment led to significant improvements in his skin condition, including a reduction in erythema and scaling. Laboratory tests also showed improvement in serum Immunoglobulin E (IgE) levels and other markers. We report with a literature review.

Keywords: atopic dermatitis; epidermal nevi; dupilumab; unilateral

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1. Introduction

Epidermal nevus syndrome (ENS) is a congenital disorder characterized by extensive linear epidermal lesions consisting of epidermal nevi and extra-dermal involvement. Epidermal nevi are characterized by epidermal thickening, with linear brown patches or thin plaques often present along the Blaschko line. The disease usually presents at birth or within the first year of life, but rarely later in childhood. We have previously reported adult-onset epidermal nevi localized on the right side of the body which was also complicated by atopic dermatitis (AD)[1]. Here, we report a case of epidermal nevi improved by the use of dupilumab for AD.

2. Case synopsis

A 36-year-old male presented to our department with abnormal keratinization on the right side of his body. He had AD since childhood and had been using topical steroids but noticed a marked difference in skin manifestations from left to right around the age of 15. He had a history of childhood asthma and cataracts, and he had no family history of genetic disorders. Initial clinical manifestations revealed that ichthyosiform erythroderma-like skin symptoms were localized on the right side of the body and thick scaly nodules were found in some places. There were no other physical abnormalities.

He continued topical steroid treatment, but due to persistent generalized erythema and scaling, he started dupilumab treatment for AD at age 41 (**Figure 1a**). Pre-administration laboratory

investigation revealed high levels of serum total Immunoglobulin E (IgE) (6700 U/mL, normal < 170) and thymus and activation-regulated chemokine (TARC) (1230 pg/mL, normal < 450). The Eczema Area and Severity Index (EASI) was 38.5. After 8 weeks of treatment, erythema and lichenification of the entire body improved, as did the keratotic lesions on the right side of the body (**Figure 1b**). At nine months after the initiation of the treatment, laboratory tests showed marked improvement with serum total IgE (3000 U/mL) and TARC (180 pg/mL).

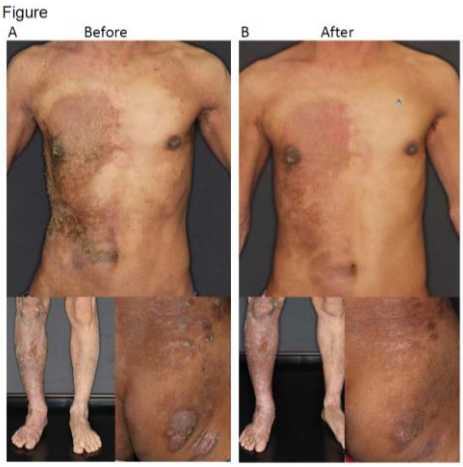

Figure 1. Clinical appearance before and after treatment with dupilumab; **(a)** clinical pictures of the entire body including ventral side of trunk, lower limbs and buttocks; **(b)** clinical pictures of the entire body 8 weeks after treatment with dupilumab.

3. Discussion

We experienced a case of epidermal nevi complicated by AD and administered dupilumab for AD. There is only one case of ENS associated with AD; a patient with a subset of ENS, Garcia-Hafner-Happle syndrome, was reported to be associated with AD. In the case report of Mizutani et al.[2], the details of AD treatment were not described. The pathogenesis of epidermal nevi is far from clear. For epidermal nevi with inflammatory linear verrucous epidermal nevi (ILVEN) and verrucous epidermal nevi (VEN) with inflammation, an analysis using proteomics has been reported. According to the article of Yuan et al.[3], the upregulated proteins in ILVEN lesions relative to VEN lesions are mainly involved in neutrophil activation. On the other hand, the downregulated proteins are mainly involved in the cellular response to cytokine stimulation, cell adhesion, and Th1 and Th2 differentiation[3]. Our case is complicated by AD, which is expected to be associated with increased Th2 cytokines such as IL-4 and IL-13. Given that IL-13 promotes fibrosis[4], we postulate that IL-13 may be involved in the pathogenesis of AD and nevus in this patient. Further research effort should be directed at exploring the role of Th2 cytokines in epidermal nevus; such effort may result in expanding the treatment options for epidermal nevus.

Conflict of interest

The authors declare no conflict of interest.

References

1. Kitayama N, Otsuka A, Kaku Y, et al. Case of unilateral epidermal nevi without extracutaneous anomalies. *The Journal of Dermatology* 2016; 43(10): 1241–1242. doi: 10.1111/1346-8138.13380
2. Mizutani Y, Nagai M, Iwata H, et al. Epidermal nevus syndrome associated with dwarfism and atopic dermatitis. *Children* 2021; 8(8): 267. doi: 10.3390/children8080697
3. Yuan T, Lu X, Tang B, et al. Differences in clinical characteristics and lesion proteomics between inflammatory linear verrucous epidermal nevus and local verrucous epidermal nevus. *Journal of Proteomics* 2022; 260: 104554. doi: 10.1016/j.jprot.2022.104554
4. Nguyen JK, Austin E, Huang A, et al. The IL-4/IL-13 axis in skin fibrosis and scarring: mechanistic concepts and therapeutic targets. *Archives of Dermatological Research* 2020; 312: 81–92. doi: 10.1007/s00403-019-01972-3