

A Case of Verrucous Epidermal Naevus Successfully Treated with Acitretin

Sir,

Verrucous epidermal naevus (linear epidermal naevus), characterized by closely set verrucous papules that may form large plaques, can be localized or systematized (unilateral or bilateral). Therapeutic modalities, including excision, laser, electrofulguration, cryotherapy, dermabrasion and chemical peels, have been tried with some success. There have been few cases of verrucous epidermal naevus treated with systemic retinoids.

Here we present a case of systematized unilateral verrucous epidermal naevus responsive to acitretin and stress the importance of the histopathologic pattern of naevi in the management of oral retinoid therapy.

CASE REPORT

A 20-year-old otherwise healthy patient presented with verrucous, papular lesions arranged in a linear and curved fashion since birth. General physical examination was normal. Dermatologic examination revealed unilaterally localized brown, hyperkeratotic, verrucous, papular lesions on the right side of the trunk, buttock and thigh. The lesions had a linear configuration and followed skin tension lines (Blaschko's lines) (Fig. 1). No pathologic finding was detected in routine biochemical analyses. Skin biopsy specimen showed hyperkeratosis, acanthosis, papillomatosis and elongated rete ridges.

The patient was given acitretin capsules (Neotigason, Roche) 75 mg/daily (approximately 1.1 mg/kg/day). At the end of the third week of oral retinoid therapy, the lesions disappeared but slight hyperpigmentation remained (Fig. 2). The dose was reduced to 50 mg/daily and discontinued 3 months later. Lesions began to reappear following a complete remission period of 6 weeks.



Fig. 1. Unilaterally localized brown, hyperkeratotic, papular lesions.

Oral retinoid therapy was well tolerated by the patient. The only distressing side effect was palmoplantar desquamation, which disappeared upon discontinuation of the drug.

DISCUSSION

Oral retinoid therapy in cases of verrucous epidermal naevus is a matter of debate. Its effectiveness is usually limited by possible side effects, which will usually appear in the high dose and long-term treatment period.

Verrucous epidermal naevi are hamartomas that originate in embryonic ectoderm. Most cases show the histologic picture of a benign papilloma characterized by hyperkeratosis, papillomatosis and acanthosis with elongation of rete ridges. There are also other histopathologic varieties, namely epidermolytic hyperkeratosis (epidermolytic type of epidermal naevus), Darier's disease-like type (acantholytic dyskeratotic epidermal naevus) and linear lichen planus (lichenoid epidermal naevus).

Even if it usually produces a temporary response, systemic retinoid therapy can be a reasonable alternative, especially for systematized unilateral/bilateral cases of verrucous epidermal naevi. In the literature there are few cases of verrucous epidermal naevus treated with etretinate with good temporary effect (1–4).

It is proposed that the histopathologic pattern of verrucous epidermal naevi may be of help in the management of oral retinoid therapy. For example, the initial dosage should be fairly low in "epidermolytic epidermal naevus" in order to avoid



Fig. 2. Appearance of the lesions at the end of the third week of oral retinoid therapy.

increased blister formation (5). In addition, some authors suggest that all cases of verrucous epidermal naevus should be examined for epidermolytic hyperkeratosis, because gonadal mosaicism (arising from a gene mutation in early embryogenesis) in these patients may be responsible for transmission of the abnormality to the offspring. So, it is reasonable to inform patients with epidermolytic epidermal naevus of the risk of transmission of epidermolytic hyperkeratosis with a generalized cutaneous involvement to the next generation and the possibility of prenatal diagnosis of generalized epidermolytic hyperkeratosis (6).

In conclusion, oral retinoid therapy is a suitable therapeutic approach for cases of systematized verrucous epidermal naevus, as in our patient. Long remission periods may be obtained by reducing oral retinoid dosage to a minimum level at which no new lesion appears. It should also be emphasized that histopathologic pattern is also one of the most important factors in the management of oral retinoid therapy.

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