Unilateral Keratosis Lichenoides Chronica

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Keratosis lichenoides chronica is rare chronic dermatosis characterized by progressive development of licheniod papulonodules especially on the extremities and trunk. A 15-year-old male patient had erythematous to violaceous scaly patches and plaques on the left side of trunk and lower extremity along Blaschko's lines. Clinical and histologic findings were compatible with keratosis lichenoides chronica showing unilateral distribution.
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Key Word: Unilateral keratosis lichenoides chronica

Keratosis lichenoides chronica is a rare and chronic skin disease characterized by symmetric, erythematous to violaceous papular and nodular lesions typically arranged in a linear and reticulate pattern, mostly marked on the extremities and accompanied by a seborrheic dermatitis-like eruption on the face.[1] Histologic features are often nonspecific and include irregular thickening of the epidermis and a rather heavy superficial dermal infiltrate of lymphocytes, histiocytes, and plasma cells.[2] The authors report a case of keratosis lichenoides chronica that showed unilateral distribution.

REPORT OF A CASE

A 15-year-old male patient visited our clinic for the evaluation of asymmetric skin rashes on the left side of trunk and lower extremity which he had for 6 months. His skin lesion began to appear at his left flank and spread gradually to his left thigh with scales. There were no subjective symptoms. There was no history of drug intake, trauma or prior eruption in that area. No other family member was affected. Physical examination showed violaceous to erythematous lichenoid and scaly papular eruptions and infiltrated plaques over the left side of flank, groin, and lower extremity along Blaschko's lines(Fig. 1, 2). There were no nail changes and mucosal involvement, Laboratory studies, including complete blood count with differential cell count, urinalysis, liver function test, VDRL, TPHA, ASO titer, CRP, RA factor, ANA, LE cell, chest PA and KOH smear for fungus were all negative or within normal limit.

Biposy was performed on the left thigh. A scalpel biopsy specimen revealed hyperkeratosis, focal parakeratosis, hypergranulosis, irregular acanthosis, follicular plugging and dyskeratosis in the epidermis. In several areas, the acanthotic epidermis showed hydropic degeneration of basal cells, and melanin incontinence. The upper dermis showed hydropic degeneration of basal cells, and melanin incontinence. The upper dermis showed zones of increased vascularity and band-like lymphocytic infiltration. Cytoid bodies were not seen(Fig. 3). Direct immunofluorescent study revealed streaky deposition of fibrinogen along the dermoepidermal junction and papillary tips and IgG, IgA, C3 were not seen. On the basis of clinical and pathological findings, a diagnosis of keratosis lichenoides chronica was made.

This patient was treated by topical corticosteroid therapy and intralesional injection of triamcinolone acetonide for 3 months but his skin lesion persisted except for the disappearance of scales.

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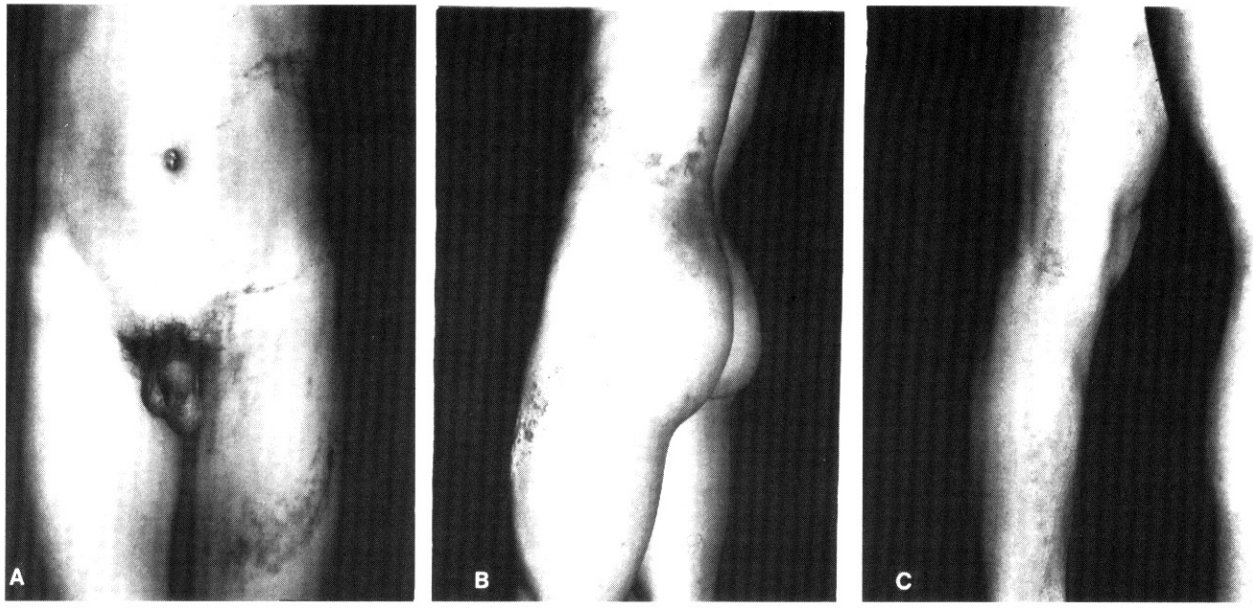

Fig. 1. Violaceous to erythematous lichendoid eruptions and plaques on the left side of trunk, thing(A), flank, lateral side of left buttock, upper thigh (B), and medial side of thigh and leg(C).

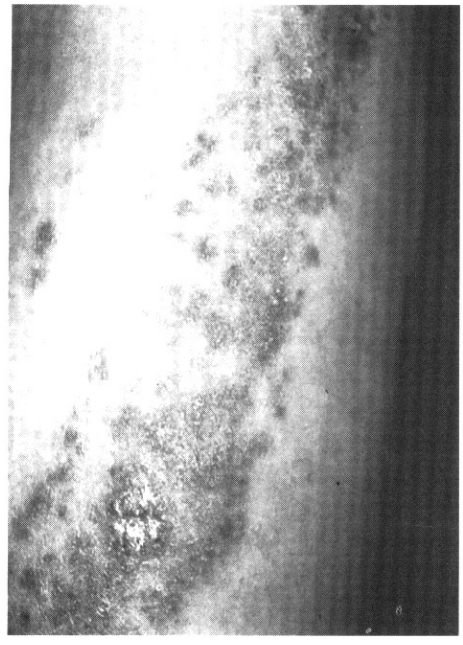

Fig. 2. Close up view: Violaceous to erythematous lichenoid scaly confluented papules and infiltrative plaques on the thigh.

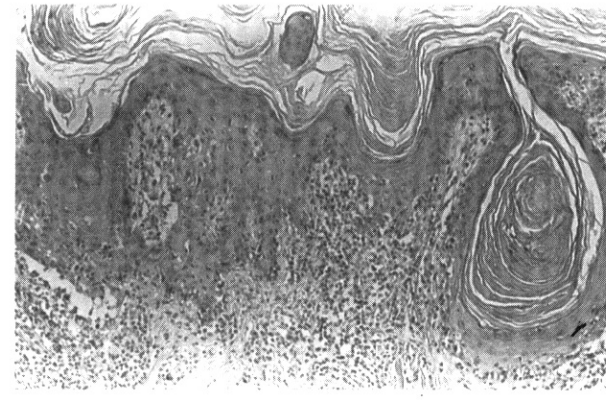

Fig. 3. Specimen from skin lesion of thigh showed hyperkeratosis, focal parakeratosis, hypergranulosis, irregular acanthosis, follicular plugging and dyskeratosis in the epidermis. The acanthotic epidermis showed hydropic degeneration of basal cells and melanin incontinence. The epidermis showed zones of increased vascularity and lymphocytic infiltration (H & E stain, × 100).

DISCUSSION

Keratosis lichenoides chronica originally described by Kaposi as lichen ruber verrucosus et reticularis received its present name in 1972 by Margolis et al[3]. There are many synonyms such as Nekam's disease,[4] porokeratosis striata lichenoides[5], lichen rubra monilliformis, and lichen verrucousus et reticularis.[6]

The main clinical manifestations of this rare dermatosis are lichenoid hyperkeratotic papules which may develop directly as goups, or in a linear or reticular pattern and, in addition. seborrhea-like dermatitis of the face. Palmoplantar involvement and nail changes have been reported in about one third of the patients.[7] Extracutaneous signs, such as oral ulcers,[8,9] ocular pemphigoid-like lesions, and hoarseness[3] ar also reported. The nail changes in keratosis lichenoides chronica may superficially resemble psoriasis, but pitting and pustulosis do not occur. Hyperkeratotic hypertrophy of the periungal tissue is a distinctive feature.[10] The cause of the keratosis lichenoides chronica is unknown. authors suggest that the disorder is a manifestation of acquired toxoplasmosis[4] and immunodeficiency.[2] Whether keratosis lichenoides chronica is a distinct entity or not is unclear,[4] and there are some suggestions that it is a variant of lichen planus.[8-11] Histologic feature of our case(Fig. 3) resemble that of lichen planus, but it showed parakeratosis, alternating areas of atrophy, acanthosis, and heavier infiltration than usually seen in lichen planus. Histologic features of our case resemble inflammatroy linear verrucous epidermal nevus and lupus erythematosus. But inflammatory linear verrucous epidermal nevus is usually present at birth or appears during the few years of life and is characteristically pruritic. We could rule out lupus erythematosus by direct immunofluorescent study. Typically, keratosis lichenoides chronica is recalcitrant to all forms of treatments. Corticosteroid, levamisole,[2] and chloroquine[12] are all unsatisfactory. There are some reports of successful treatment with etretinate[7] and PUVA.[1,9] The coumbination of PUVA and etretinate have improved some cases of nail changes in keratosis lichenoides chronica.[10] In most cases there ae no subjective symptoms except one[7] and the skin lesions are distributed symmetrically. In our case, there were no subjective symptoms but the distrubution of the skin lesion was unilateral.

REFERENCES

1. Ryatt KS, Greenwood R, Cotterill JA:Keratosis lichenoides chronica. Br J dermatol 106:223-225, 1982.
2. Nabai H, Mehregan AH:Keratosis lichenoides chronica. J Am Acad Dermatol 2:217-220, 1980.
3. Margolis MH, Cooper GA, Johnson SAM: Keratosis lichenoides chronica. Arch Dermatol 105:739-743, 1972.
4. Menter MA, Morrison JG:Lichen verrucosus et reticularis of Kaposi(porokeratosis striata of Nekam): a manifestation of acquired adult toxoplasmosis. Br J Dermatol 94:645-654, 1976
5. Miller HE:Lichen verrucosus et reticularis (Kaposi):porokeratosis striate (Nekam) morbus mobiliformis lichenoides (Wise and Rein). Arch Dermatol Syph 52:129-130, 1945.
6. Mehregan AH, Heath LE, Pinkus H:Lichen ruber moniliformis and lichen ruber verrucosus et reticulosis of Kaposis. J Cutan Pathol 11:2-11, 1984.
7. David M, Filhaber A, Rotem A, Weisman VK, sandbank M:Keratosis lichenoides chronica with prominent telangiectasia: reponse to etretinate. J Am Acad Dermatol 21:1112-1114, 1989.
8. Kersey P, lve FA: Keratosis lichenoides chronica is synonymous with lichen planus. Clin Exp Dermatol 7:49-54, 1992.
9. Lang PG:Keratosis lichenoides chronica: successful treatment with psoralen-ultraviolet-A therapy. Arch Dermatol 117:105-108, 1981.
10. Baran R, Panizzon R, Goldberg L: The nail in keratosis lichenoides chronica: characteristics and response to treatment. Arch Dermatol 120:1471-1474, 1984.
11. Petrozzi JW: Keratosis lichenoides chronica: possible variant of lichen planus. Arch Dermatol 112: 709-711, 1976.
12. MacDonald DM, Williams DT: Lichen verrucosus et reticulosis of Kaposi(porokeratosis of Nekam). Br J Dermatol 91 (suppl):39, 1974.