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Extragenital unilateral lichen sclerosus et atrophicus in a child: a case report

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Abstract

Lichen sclerosus et atrophicus (LSA) is a rare chronic inflammatory dermatosis with anogenital and extragenital presentations. Extragenital lichen sclerosus is most common on the neck, shoulders, and upper portion of the trunk. Linear lesions are uncommon in LSA and very few such cases are reported.

We describe a 5-year-old female child with unilateral linear extra-genital lichen sclerosus. The lesions were confined to the left lower extremity linearly along the left foot, left knee, thigh and a lesion on the left side of her abdomen. Histological findings obtained from the thigh lesion were those of typical LSA.

Introduction

Lichen sclerosus (LS) is a chronic dermatitis predominantly found in the anogenital area. It can be found in patients of any age group, sex, or race. Linear extragenital lichen sclerosus represents an exceptionally rare form of lichen sclerosus. We report a case of extra-genital linear lichen sclerosus et atrophicus in a child. This case suggests that there is a linear form in LSA as already recognized in localized scleroderma and it can occur in children also.

Case report

A 5-year-old female child was brought with asymptomatic linear skin lesions involving her left leg and the left side of her abdomen that had appeared, at the age of 4. Initially, his parents noted a group of small, shiny white flat lesions located over the dorsum of the left foot. Over the following few months, the similar lesions also appeared over leg, near the knee, thigh and abdomen, coalesced into patches, assuming the form of a linear lesion. Some of them were depressed, wrinkled and slightly atrophic. There was no history of any genital lesions or complaints. Parents denied history of any past treatment.

On examination, lesions consisted of sharply demarcated, hypo-pigmented, atrophic, depressed, wrinkled, confluent, and isolated patches in a linear configuration, along the left lower extremity and left side of abdomen. (**Fig 1**)



Fig 1: Multiple hypopigmented patches over left lower extremity and left side of abdomen running linearly.

On the left abdominal wall, there was a parchment like patch. **(Fig 2)** There was no underlying bony atrophy. Rest of her cutaneous examination was normal including genital examination.



Fig 2: Close-up view of multiple atrophic, whitish patches over left foot, knee and thigh and abdomen.

A provisional diagnosis of linear lichen sclerosus, lichen striatus or linear nevus depigmentosus was thought clinically.

Skin biopsy specimen taken from thigh lesion, showed features consistent with lichen sclerosus et atrophicus including epidermal atrophy, vacuolar alteration of dermo-epidermal junction, hyalinized papillary dermal collagen and few melanophages with sparse superficial perivascular lymphocytic infiltrate. (**Fig 3, 4**)

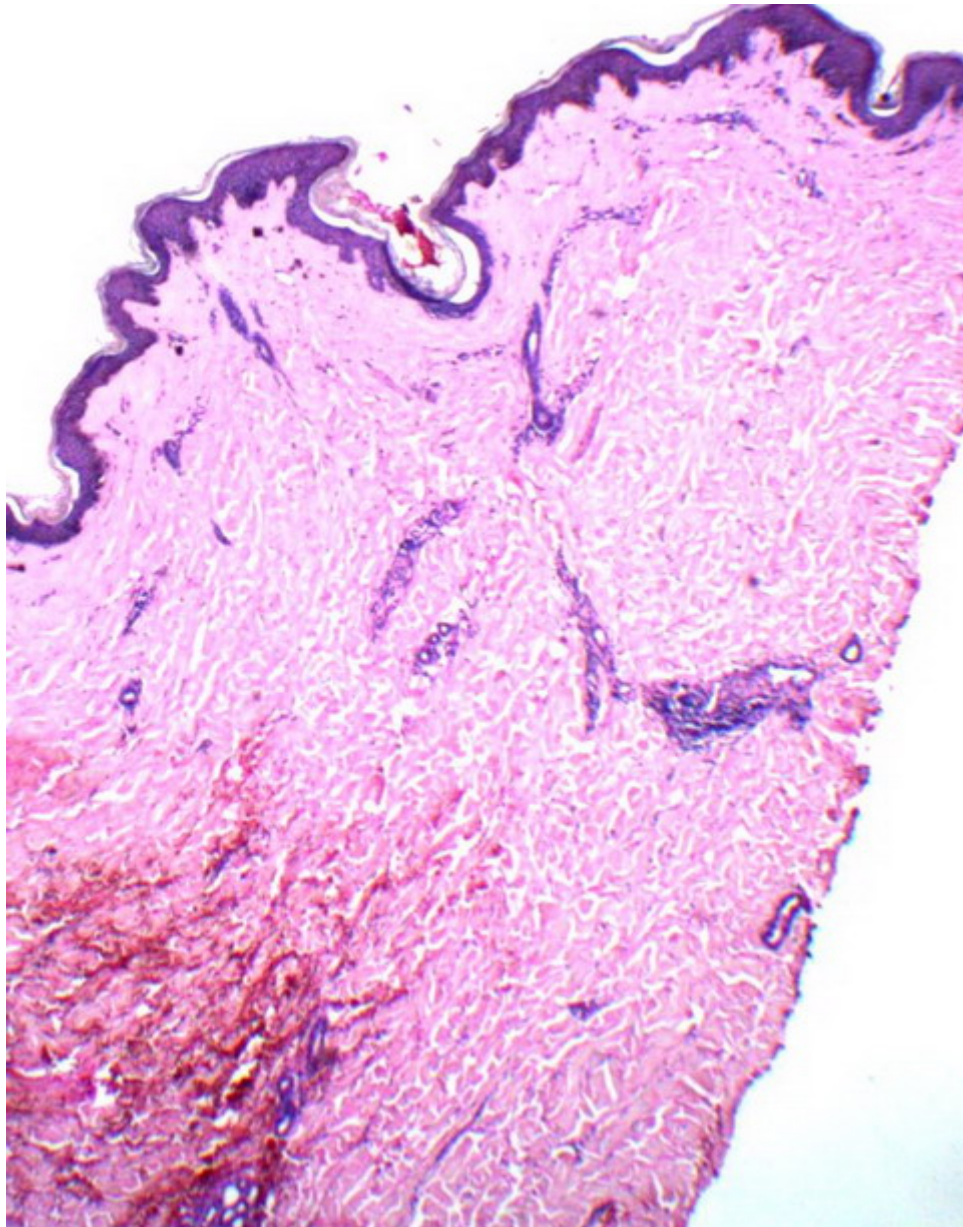


Fig 3: Skin biopsy from thigh lesion showing atrophic epidermis, vacuolar interface and hyalinized papillary dermis. (H&E, 40X)

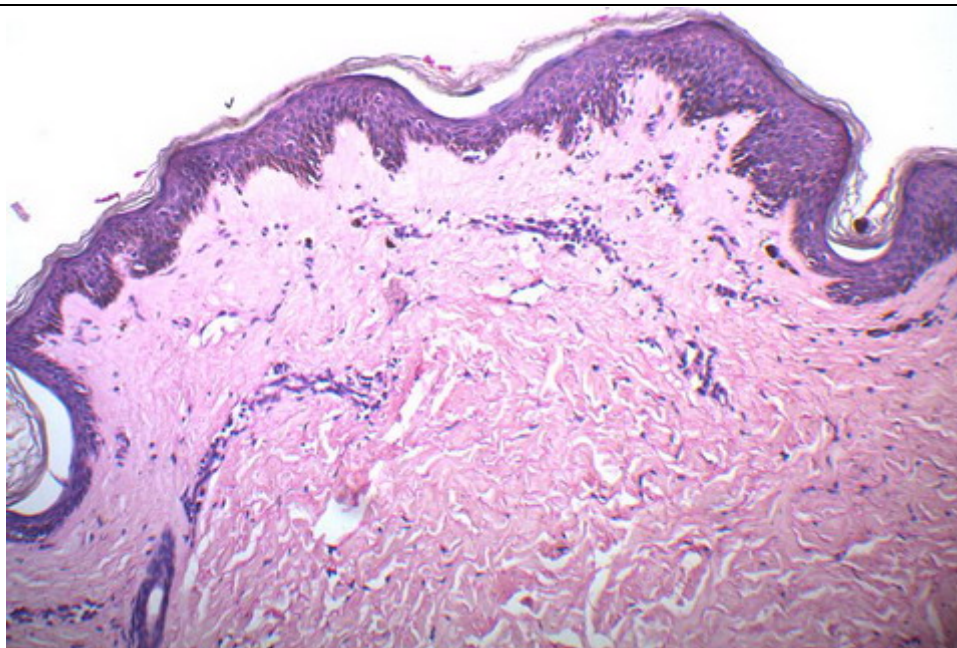


Fig 4: High power view showing hyalinized collagen, lymphocytic infiltrate, melanophages and slightly atrophic epidermis with interface. (H&E, 100X)

Hemogram, autoantibody screen, liver, kidney, and thyroid function tests were normal. *Borrelia burgdorferi* serology was not done due to cost constraints of patient.

Based on clinico-pathological correlation a diagnosis of linear extragenital lichen sclerosis was made. Patient was advised clobetasol propionate 0.05% cream along with moisturizers and is under follow-up.

Discussion

Lichen sclerosus (LS) is a chronic dermatitis affecting predominantly, the anogenital area. It can be found in patients of any age group, sex, or race, but is most commonly present in peri- or postmenopausal women. Although the etiology of LS remains uncertain, an autoimmune process is believed to underlie this condition. [1]

Extragenital lichen sclerosus without accompanying genital lesions was recorded in 805 of 4280 cases reviewed by Meffert et al.[2] Extragenital lichen sclerosus is most common on the neck, shoulders, and upper portion of the trunk. It is generally asymptomatic, but occasionally pruritic. Most lesions of extragenital lichen sclerosus present as flat, white, polygonal papules, and slight atrophic white plaques. [3]

Extragenital lichen sclerosus with linear lesions or following the lines of Blaschko represents rare presentation of lichen sclerosus.

In 1995, Izumi et al. [4] were the first to describe a linear form of lichen sclerosus extending from the left upper back and along the left arm, probably following the lines of Blaschko. Okamoto et al added another case of linear lichen sclerosus in a 23-year-old woman who developed initial lesions at the age of 18. [2,4]

Location of lesions preferentially on left side of body in most of the reported cases has been attributed to stronger cell-mediated immune hypersensitivity in the left side of the body than the right in healthy young subjects and it is speculated that the cellular immune responsiveness might influence the confinement of the Blaschko-linear lichen sclerosus to the left side of the body. [3,5] In our case also, lesions were located on left lower extremity and left side of abdomen and probably were following lines of Blaschko supporting above speculation. The lines of Blaschko were described and drawn in 1901 by Alfred Blaschko (1858-1922), a private practitioner of dermatology in Berlin. In disorders that affect skin areas corresponding to Blaschko's lines, it is believed that two distinct cell clones arise early in embryogenesis, often produced by genetic mosaicism.[2,6]

Various acquired conditions that can follow Blaschko's lines include lichen striatus, linear psoriasis, linear lichen planus, linear scleroderma, linear atrophoderma etc. [6]

Our case thus presents a rare presentation of lichen sclerosus with linear and unilateral extragenital lesions in a child.

References

1. Funaro, D. Lichen sclerosus: a review and practical approach. *Dermatologic Therapy*, 2004;17: 28-37
2. Choi SW, Yang JE, Park HJ, Kim CW. A case of extragenital lichen sclerosus following Blaschko's lines. *J Am Acad Dermatol* 2000;43:903-4
3. Pavlovic MD. Linear lichen sclerosus with underlying bony atrophy. *J Am Acad Dermatol* 2004;50(3):E4
4. Izumi T, Tajima S. A case of linear type of lichen sclerosus et atrophicus? *J Dermatol* 1995; 22: 279- 82.
5. Dane S, Erdem T, Gumustekin K. Cell-mediated immune hypersensitivity is stronger in the left side of the body than the right in healthy young subjects. *Percept Mot Skills* 2001; 93: 329- 32.
6. Tagra S, Talwar AK, Walia RS. Lines of Blaschko. *Indian J Dermatol Venereol Leprol* 2005; 71: 57- 9