

Recurrent Linear Inflammatory Rash with Background Leukoderma

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Clinical Findings

An 8-year old boy presented with persistent linear leukoderma over his left leg that followed an inflammatory dermatosis four years prior to consultation. The same rash subsequently recurred within the area of leukoderma.

Clinical examination revealed confluent hypopigmented, non scaly patches in a linear distribution over the medial aspect of the left knee, extending down the anterior aspect of the leg. There was a superimposed scaly and crusted linear rash (Figure 1). The hypopigmented patches were accentuated with wood's light. There were no patches representing vitiligo at other sites and there was no family history of vitiligo.

Histopathological Findings

Skin biopsy taken from the hypopigmented area showed an epidermis with laminated stratum corneum, hypopigmented basal keratinocytes and scant lymphocytic inflammation around superficial dermal vessels. Schmorl's stain showed decreased pigment in the basal layer (Figure 2).

A second biopsy, taken from the scaly rash, showed an epidermis covered by focally compact and parakeratotic stratum corneum. Prominent lymphocytic inflammation with liquefaction degeneration of the junctional zone of the epidermis and formation of cytoid bodies were apparent. The lymphocytes extended around superficial, mid and deep dermal vessels and around the appendages (Figure 3).

What is Your Diagnosis?

Answer: Lichen striatus with associated leukoderma.

Discussion

Lichen Striatus (LS) is an uncommon inflammatory dermatosis that can affect any age but is typically more frequent in children aged from 5-15 years [1,2].



Figure 1: Linear hypopigmented patches on the left leg, with superimposed erythematous, scaly and crusted rash.

The aetiology and pathogenesis of this disease have not been identified. It is suggested to be a cell mediated immune reaction towards mutant keratinocyte clone that have developed in early foetal life and migrated along the Blaschko's lines. This hypothesis is supported by the striking linearity of lichen striatus that usually follows Blaschko's lines indicating a somatic mosaicism. In addition, the majority of lymphocytes in the epidermis are CD8 positive reflecting a cytotoxic immune reaction and these may form clusters around dyskeratotic keratinocytes [1-3]. The triggers of this condition are unknown but the early onset in childhood and seasonal variations have suggested the possible presence of viral infection [1]. Clinically LS usually involves a single limb followed by the trunk and face. It has an inflammatory phase of discrete and coalescing flat topped papules that are scaly or smooth, pink, erythematous or skin coloured. The rash usually resolves spontaneously in up to several months [1-3]. It can be followed in 30-50% of cases by single or parallel streaks of post inflammatory hypopigmentation resulting in lichen striatus associated leukoderma [2,3]. The latter usually resolve in one to three years [1]. Hyperpigmentation can also follow LS but much less frequently [3]. In dark skinned individuals the rash may initially manifest as hypopigmented linear streaks but careful clinical examination can often reveal the primary flat topped papules [1,4].

The typical histopathological findings of lichen striatus are those of

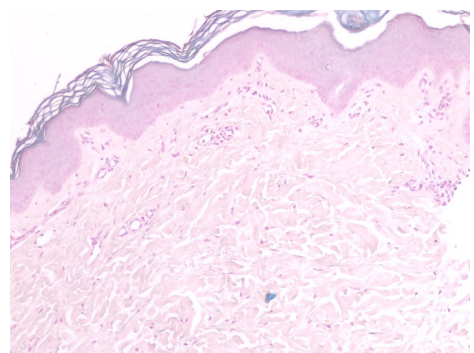


Figure 2: Laminated stratum corneum, hypopigmented basal keratinocytes and scant perivascular lymphocytic inflammation in the upper dermis. Schmorl's stain, original magnification x100.

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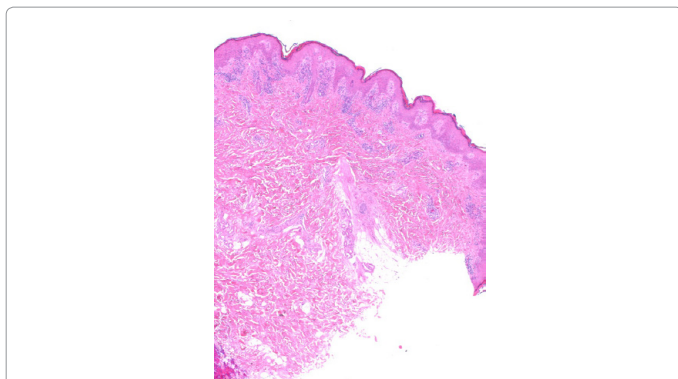


Figure 3: Prominent lichenoid lymphocytic inflammations, extending around mid and deep blood vessels and around appendages. Haematoxylin and eosin, original magnification $\times 25$.

a lichenoid reaction pattern with lymphohistiocytic inflammation along the dermoepidermal junction and lymphocytes in a perivascular and peridnexal distribution. Extension of lymphocytes around the sweat glands is a helpful clue to diagnosis. The epidermis can be associated with hyperkeratosis, focal parakeratosis, acanthosis, mild spongiosis and lymphocytic exocytosis. Dyskeratotic keratinocytes can usually be found at any level in the epidermis [1-3]. Lichen striatus is considered a self-limiting disorder and no treatment is usually required. Relapses are unusual but can occur either in the same site or in a new location [1,3].

Segmental Vitiligo (SV) is a rare subtype of vitiligo that usually has an onset in childhood. SV presents clinically as one or more depigmented macules in a linear distribution involving a unilateral segment of the body, with an abrupt discontinuation at the midline [5]. The process favours the face particularly the trigeminal dermatome [4,5]. The linearity of this condition is neither strictly dermatomal nor Blaschkolinear [5]. It usually evolves over one or two years, then stabilizes or regresses. Some cases may be associated with poliosis [4]. The biopsy in vitiligo usually does not show significant inflammation

but there may be a focus of subtle lymphocytes at the boundary of the advancing edge. A rare variant of vitiligo can be inflammatory, presenting as an erythematous raised border outlining the depigmented patches [3].

Our patient had typical LS four years prior to presentation, which resolved and left behind an area of leukoderma. In contrast to this common sequel of LS, the leukoderma persisted and was the site of subsequent flare. In contrast to vitiligo the flare did not localise to the periphery of the leukoderma but occurred within the area of hypopigmentation. Although the biopsy of the hypopigmented area showed changes which were indistinguishable from vitiligo, this may represent the late pathology that occurs in the wake of lichen striatus.

Our patient's condition is best considered to be a lichen striatus induced leukoderma, for a diagnosis of vitiligo as an independent phenomenon, the presence of vitiligo at other site would be essential.

Learning Points

- Leukoderma is a common sequel of lichen striatus that can last for one to three years, and may be difficult to distinguish from segmental vitiligo.
- Relapses of Lichen striatus are uncommon and the localisation in the area of leukoderma allowed definite diagnosis as a sequale.

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