

## Simultaneous Occurrence of Lichen Nitidus and Morphea

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### ABSTRACT

Lichen nitidus and morphea are common diseases, but an associated localization of both lesions is rare. Here, we describe the first case of lesions distributed along Blaschko's lines. A 24-year-old Japanese woman was referred to our clinic for evaluation of band-like plaques of 18-months history on the right lateral side of her abdomen. In addition, multiple milky-white papules were seen within the plaques. Histopathological examination showed there was sclerosis in the lower half of the dermis and well-circumscribed, dense, papillary dermal lymphohistiocytic aggregations showing a so-called "claw clutching a ball." Immunohistochemical analysis revealed that the morphea and lichen nitidus had similar characteristics. We speculated that unique immunologic events led to the development of lichen nitidus and morphea in our patient.

**Key words** Blaschko's lines; lichen nitidus; linear scleroderma; localized scleroderma; morphea

Morphea, also known as localized scleroderma, is classified into some subtypes.<sup>1</sup> Clinically, in linear morphea which is one of subtypes in morphea, sclerosis is distributed on unilateral limbs or the face and head along Blaschko's lines. On the other hand, there are a few reports of linear nitidus along Blaschko's lines.<sup>2</sup> To our knowledge, there has been no report of simultaneous occurrence of lichen nitidus and morphea. We present the first case of lesions distributed along Blaschko's lines.

### PATIENT REPORT

A 24-year-old Japanese woman was referred to our clinic for evaluation of plaques on her abdomen. She had been aware of papules and plaques for 18 months. Physical examination revealed band-like plaques, light red-brown in color, along Blaschko's lines on the right lateral region of her abdomen (Fig. 1a). In addition, there were multiple milky-white papules within the

plaques (Fig. 1b). Dermoscopically, the papules were shown to be well-defined circular hypopigmented structures (Fig. 1c). Clinical differential diagnosis included atrophoderma, morphea and ashy dermatosis. Histopathologically, there was sclerosis in the lower half of the dermis (Fig. 1d) and perivascular lymphoplasmacytic infiltration near the eccrine glands (Fig. 1e). In addition, we observed well-circumscribed, dense, papillary dermal lymphohistiocytic aggregations showing a so-called "claw clutching a ball" pattern (Fig. 1f). Immunohistochemically, these aggregations were mainly composed of CD4<sup>+</sup> and CD8<sup>+</sup> lymphocytes and some CD68<sup>+</sup> cells (Figs. 1g, h, and i). In addition, the aggregations also contained S-100 protein<sup>+</sup> and CD1a<sup>+</sup> cells. (Figs. 1j and k). There were similar pattern of lymphocytes infiltration around vessels (not shown). We made a final diagnosis of morphea in association with lichen nitidus. Physical and serological findings were negative for systemic sclerosis. Although the papules disappeared after topical corticosteroid treatment for 3 months, morphea lesions were not improved by treatment for 6 months.

### DISCUSSION

Morphea, also known as localized scleroderma, is classified into subtypes based on clinical and histopathological findings. In the recent Padua Consensus classification, morphea subtypes include circumscribed morphea, linear scleroderma, generalized morphea, pansclerotic morphea and mixed morphea.<sup>1</sup> Clinically, in linear morphea, sclerosis is distributed on unilateral limbs or the face and head along Blaschko's lines. On the other hand, there are a few reports of linear nitidus along Blaschko's lines.<sup>2</sup> A postzygotic mutation of epidermal progenitor cells has been suggested as a mechanism of a linear distribution.<sup>3</sup> Consequently, the mutation may be induced by trigger factors such as infections, vaccinations or trauma and give rise to a consecutive immune reaction.<sup>2, 3</sup> However, the genetic involvement is not clear in our case because mutation analysis has not performed.

There are some cases of linear lichen nitidus associated various cutaneous diseases (including oral lichen planus, lichen striatus and psoriasis).<sup>4–6</sup> Furthermore, there is an unusual histopathologic pattern of lichen

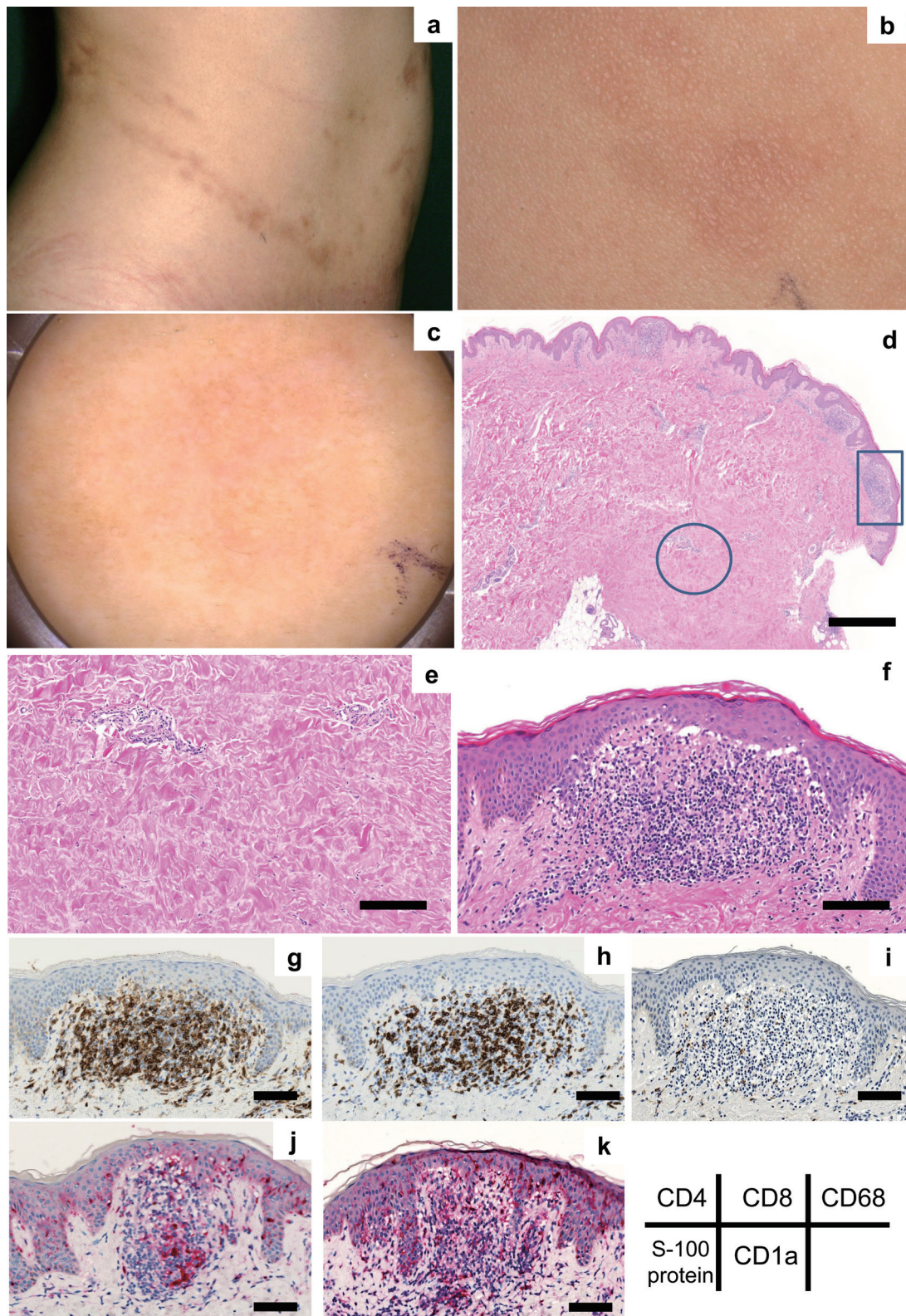
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**Fig. 1.** (a) Multiple lightly red-brown plaques along Blaschko's lines on the trunk. (b) There were multiple milky-white papules within the plaques. (c) Dermoscopy showed some ill-defined milky circular structures. (d) Histopathological examination showed sclerosis in the deep dermis (hematoxylin and eosin). Bar = 1 mm. (circle, e, square, f). (e) Inflammatory cells near eccrine glands were also seen (hematoxylin and eosin). Bar = 25  $\mu$ m. (f) There was well-circumscribed, papillary dermal lymphohistiocytic infiltration (claw clutching a ball) (hematoxylin and eosin). Bar = 100  $\mu$ m. (g, h, i, j and k) Lymphohistiocytic aggregates in a claw-clutching-a-ball fashion were mainly composed of many CD4<sup>+</sup> cells and CD8<sup>+</sup> cells, some S-100 protein<sup>+</sup> and CD1a<sup>+</sup> cells and a few CD68<sup>+</sup> cells. Bar = 100  $\mu$ m.

nitidus that could be confused with lichen scrofulosorum.<sup>7</sup> However, to our knowledge, there has been no report of simultaneous occurrence of lichen nitidus and morphea. We report here the first case of lesions distributed along Blaschko's lines.

A previous study showed infiltration of many CD4<sup>+</sup> and CD8<sup>+</sup> lymphocytes and some CD1a<sup>+</sup> cells in morphea lesions.<sup>8</sup> Intriguingly, marked infiltration of CD4<sup>+</sup> cells rather than CD8<sup>+</sup> cells and an increased number of Langerhans cells have also been suggested to play a crucial role in the immune response in lichen nitidus.<sup>9</sup> Although it is possible that these distinct entities developed coincidentally, similar proinflammatory immune responses might be induced by the infiltration of CD4<sup>+</sup> lymphocytes and Langerhans cells, leading to simultaneous occurrence of lichen nitidus and morphea in our case.

In conclusion, we consider that immunological and genetic factors may play important roles in the pathogenesis of our unusual clinical presentation.

*The authors declare no conflict of interest.*

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