http://dx.doi.org/10.5021/ad.2014.26.6.764

Segmental Vitiligo and Extragenital Lichen Sclerosus et Atrophicus Simultaneously Occurring on the Opposite Sides of the Abdomen

Hemin Lee, Yoon Jee Kim, Sang Ho Oh

Department of Dermatology, Cutaneous Biology Research Institute, Yonsei University College of Medicine, Seoul, Korea

Dear Editor:

A 10-year-old female patient visited the dermatology clinic because of hypopigmented skin lesions on both sides of her abdomen that occurred 3 years ago (Fig. 1A, B). There were no associated symptoms or history of infectious diseases, stressful events, or trauma. Her parents did not recall any history of familial autoimmune diseases. Laboratory tests on autoimmune panels (antinuclear antibody, anti-DNA screening, anti-Scl 70) were all negative. On Wood's light examination, hypopigmented lesions on the right abdomen showed marked accentuation that was clinically consistent with segmental vitiligo (SV). On the other hand, the lesions on the left side of the abdomen revealed only mild accentuation. Whereas biopsy was not performed on the lesion suspicious for SV, histopathological examination of a hypopigmented plaque on the patient's left abdomen showed follicular plugging and fibrosis with edema in the upper dermis, suggesting lichen sclerosus et atrophicus (LSA) (Fig. 1C, D). Although rare, there are incidences of morphea presenting with vitiligo, requiring attentive differential diagnosis with LSA. Yet, features that suggest morphea, such as thickened collagen bundles or atrophic eccrine glands, were not seen.

There have been previous reports of SV and LSA occurring simultaneously. However, most of the reported cases were SV that co-occurs with genital LSA[1,2] or extragenital LSA in a non-SV (NSV) patient[3]. Thus, to our knowledge, we here report the first case of extragenital LSA occurring with SV.

The significance of LSA and SV of synchronous onset can be interpreted as a deviation from a previously known mechanism for SV. Also, we believe that it suggests a possible relation between the mechanisms of two very different diseases. Although an exact pathomechanism for LSA is not yet known, there is an unproven consensus that LSA is an autoimmune disease, on the basis of evidences such as the presence of circulating antibodies targeting extracellular matrix protein 1 and the frequent association of LSA with other autoimmune diseases[4]. Concerning SV, although it is mostly thought to involve a neural mechanism, there is a view that an autoimmune basis adds to its onset as well. A case report on SV occurring concurrently with alopecia areata, psoriasis, and halo nevus supports this hypothesis, as the three diseases except SV are known to have an autoimmune basis[5]. Hence, from our interesting case of SV and LSA occurring on the opposite sides of the abdomen along Blashko's line, we suggest that there is a possibility of a common initiating factor or a shared pathomechanism existing between SV and LSA. Furthermore, if autoimmunity or auto-inflammatory reaction takes place in the onset of SV, our case could provide supporting evidence for SV sharing a common link with NSV.

Received October 4, 2013, Revised October 31, 2013, Accepted for publication November 7, 2013

Corresponding author: Sang Ho Oh, Department of Dermatology, Cutaneous Biology Research Institute, Yonsei University College of Medicine, 50 Yonsei-ro, Seodaemun-gu, Seoul 120-752, Korea. Tel: 82-2-2228-2080, Fax: 82-2-393-9157, E-mail: oddung93@yuhs.ac

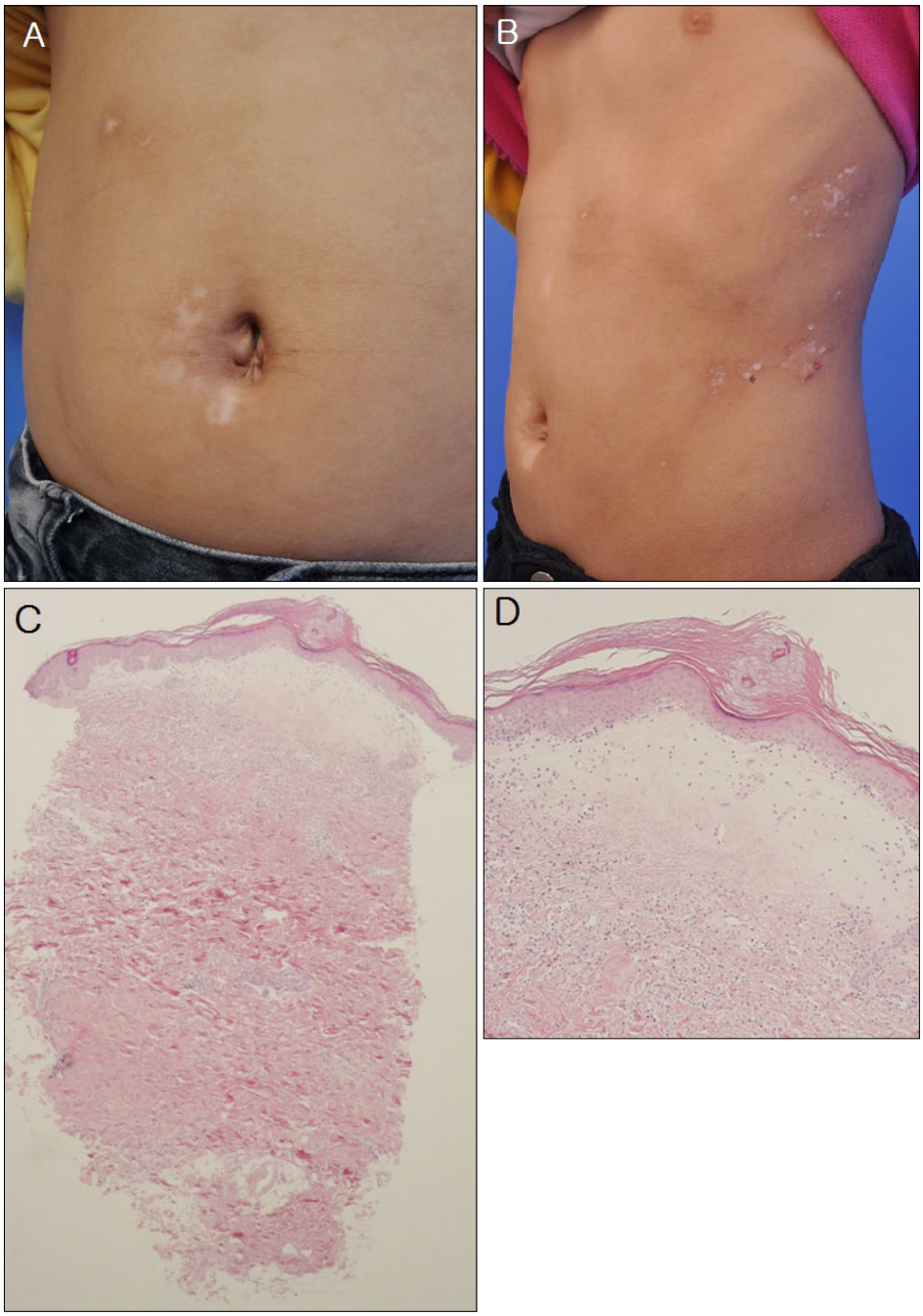

Fig. 1. (A) Well-demarcated hypopigmented patches without surface changes on the right abdomen. (B) Multiple hypopigmented papules and plaques occurring with atrophic surface change and hardening on the left abdomen. The lesions shown in both (A) and (B) had occurred spontaneously 3 years ago. (C) Histology demonstrates marked fibrosis with edema in the upper dermis, suggesting lichen sclerosus et atrophicus (H&E, ×40). (D) Higher magnification shows epidermal hyperkeratosis, superficial perivascular lymphohistiocytic infiltrate, and homogenization of the dermal collagen in the superficial dermis (H&E, ×100).

REFERENCES

1. Osborne GE, Francis ND, Bunker CB. Synchronous onset of penile lichen sclerosus and vitiligo. Br J Dermatol 2000;143:218-219.
2. Weisberg EL, Le LQ, Cohen JB. A case of simultaneously occurring lichen sclerosus and segmental vitiligo: connecting the underlying autoimmune pathogenesis. Int J Dermatol 2008;47:1053-1055.
3. Guerriero C, Manco S, Paradisi A, Capizzi R, Fossati B, Fabrizi G. Extragenital lichen sclerosus and atrophicus treated with topical steroids and retinoids in a child with vitiligo. Int J Immunopathol Pharmacol 2008;21:757-759.
4. Oyama N, Chan I, Neill SM, Hamada T, South AP, Wessagowit V, et al. Autoantibodies to extracellular matrix protein 1 in lichen sclerosus. Lancet 2003;362:118-123.
5. Picardo M. Vitiligo: new insights. Br J Dermatol 2012;166:472-473.