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CASE REPORT

A Case of Generalized Lichen Sclerosus et Atrophicus

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A 62-year-old female, with previous history of asthma and hypertension, presented with generalized hyperpigmented skin lesion, found a year ago. Physical examination revealed brown colored lichenified and sclerotic patches on the lower abdomen and flexural areas of extremities. Punch biopsy was performed and histopathological examination revealed hyperkeratosis, follicular plugging and thinning in epidermis. In dermoepidermal junction, cleft like space separating atrophic epidermis and dermis was seen. Also, lichenoid lymphocytic infiltration was observed in mid-dermis. Based on clinical and histopathological findings, a diagnosis of generlaized lichen sclerosus et atrophicus (LSA) was made. Other laboratory examinations were unremarkable. As there is no standard treatment for LSA, the patient received various treatments including topical steroid, tacrolimus and narrow-band ultraviolet B therapy. The skin lesion has softened and its color improved after treatment. LSA is defined as infrequent chronic inflammatory dermatosis with anogenital and extragenital manifestations. Generalized type is rare and genital involvement is the most frequent and often the only site of involvement. We report this case as it is an uncommon type of LSA with generalized hyperpigmented and sclerotic skin lesion in a postmenopausal female patient. **(Ann Dermatol 32(4) 327～330, 2020)**

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INTRODUCTION

Lichen sclerosus et atrophicus (LSA) is a rare chronic inflammatory skin disease originally defined by Hallopeau[1] in 1887; it mainly invades the genital region but can also include the extragenital area. An exact diagnosis and treatment are critical because of the possibility of atrophy, scar formation, and malignant changes[2]. The overall prevalence is about 0.1% to 0.3%; it is more common in female than in male, with its range varying from 10:1 to 1:1[3-5]. In female, there are two peaks of prevalence throughout the lifespan: at puberty and postmenopause. The pathogenesis of LSA has not yet been clarified. In one report, LSA seemed to be related to autoimmune diseases, as approximately 25.5% of the patients had autoimmune diseases and approximately 42% had autoimmune antibodies[6]. In addition, trauma and infection may affect the development of LSA. The clinical manifestation includes hyperpigmented or hypopigmented papules and plaques with sclerosis and atrophy involving the vulva, perineum, and perianal region in female. It can be asymptomatic but can also cause itching, pain, and dysuria. In male, it peaks around the late 40s and mainly involves the penis, causing phimosis. The incidence of extragenital involvement is only approximately 6%, with a higher incidence in female (11%～13%)[3,6]. Commonly involved extragenital areas include the buttock, thigh, chest, back, and axilla, and systemic involvement is very rare. In addition, there are few symptoms, and many are accompanied with morphea in cases of systemic involvement[7]. Herein, we report a case of generalized LSA without morphea that occurred in a postmenopausal female.

CASE REPORT

A 62-year-old female presented with extensive skin lesions accompanied by mild itching. In the past year, she gradually developed diffuse scattered brown-colored lichenified patches on her abdomen, axilla, and flexural areas of her extremities. Skin lesions showed irregularly shaped hyperpigmented patches with sclerosis (Fig. 1). The genital area showed no symptoms. She had no specific family history or past medical history except asthma and hypertension. Autoantibodies or abnormalities of hormone level such as thyroid hormones were not found in laboratory tests. She underwent skin biopsy for a diagnosis to be made. On performing histopathologic examinations (Fig. 2), follicular plugging with compact hyperkeratosis was observed in the epidermis. Thinning of the epidermis and hydropic degeneration of the basal cells were found. A cleft-like space due to the separation of the atrophic epidermis and dermis was also observed. Edema and collagen homogenization existed in the upper dermis, and inflammatory cell infiltration was observed in the mid-dermis. Based on the clinical features and histopathological findings, the patient was finally diagnosed with generalized LSA. She began treatments that consisted of systemic and topical steroids. Because there is no definite treatment guideline for generalized LSA, various treatments had been attempted, including topical steroids, tacrolimus, and narrow band ultraviolet B (UVB). After approximately 18 months of treatment, pigmentation and sclerosis of the abdomen and extremities were reduced and her symptoms were alleviated (Fig. 3). The patient is now continuing outpatient treatment.
We received the patient's consent form about publishing all photographic materials.

DISCUSSION

In both male and female, LSA mainly occurs in the genital area and sometimes accompanies symptoms such as itching and dysuria. When extragenital areas are involved, most cases show localized invasion, and generalized involvement affecting more than 2 anatomic regions[8] is very rare. Our patient manifested with hyperpigmented skin lesions diffusely scattered around the abdomen, axilla, and flexural areas of the extremities with mild itching. In some previously reported cases, systemic blisters or ulcerations were accompanied by generalized LSA. Our case is thought to be a rare case of generalized LSA, as blisters or ulcerations were not observed. In addition, preceding studies

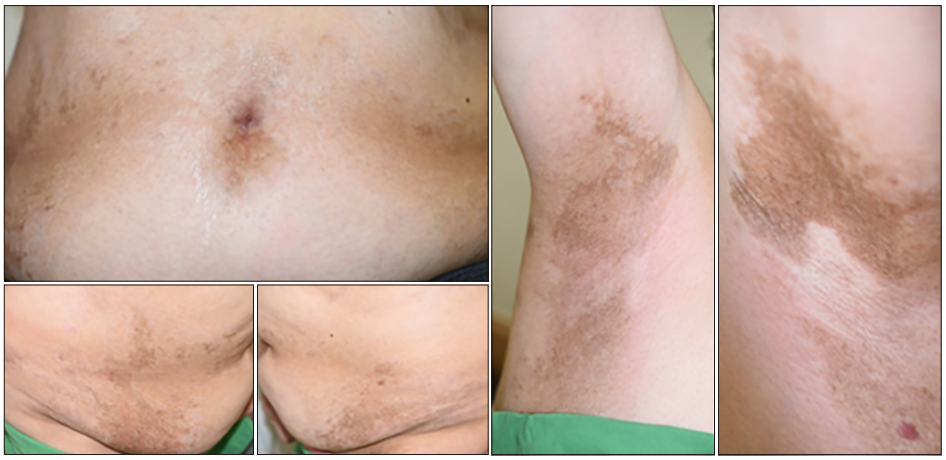

Fig. 1. Irregularly shaped hyperpigmented patches with sclerosis were seen at first visit.

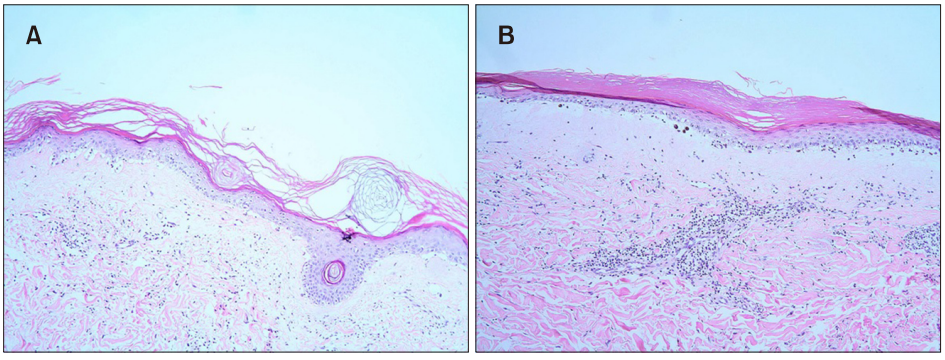

Fig. 2. Histopathological examination revealed hyperkeratosis and follicular plugging in epidermis (A), dermoepidermal separation, and inflammatory cell infiltration in mid-dermis (B); H&E stain, ×100.

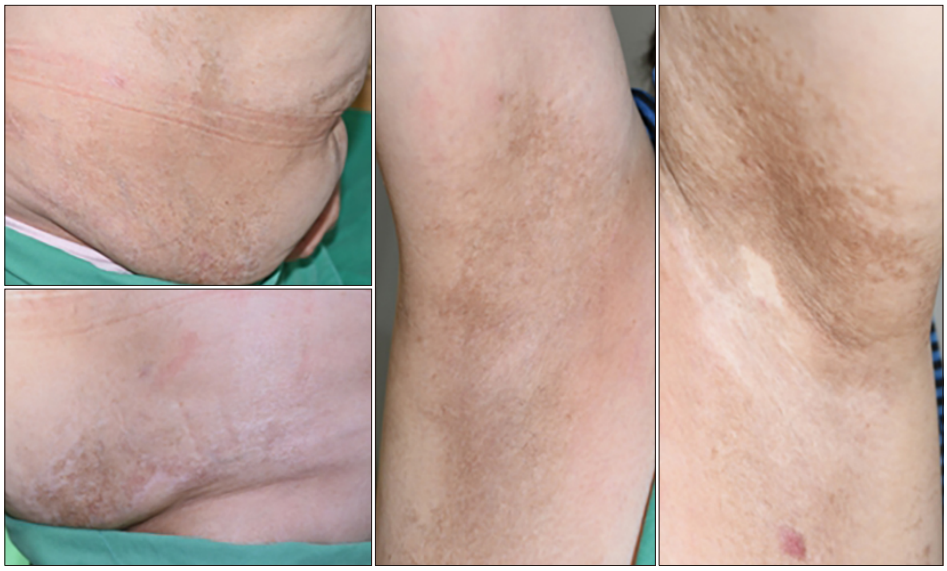

Fig. 3. Pigmentation and sclerosis of the abdomen and extremities were reduced after treatment.

about LSA hypothesized that this may be related to autoimmune factors because 21.5% of patients had autoimmune diseases and 42% had autoantibodies[6]. However, there was no specific laboratory finding in our case.

Histopathologically, atrophic epidermis accompanied by compact hyperkeratosis and follicular plugging was noted. A cleft-like space can be observed in the dermoepidermal junction due to epidermal thinning and dermal edema. Inflammatory cell infiltration can be observed in the middermis. Morphea should be differentiated from LSA. Morphea shows relatively normal epidermis without follicular plugging and does not include subepidermal separation; fibrosis is present with collagen and elastic fibers at the dermis. Inflammation of the subcutis can also be observed. According to previous reports on generalized LSA, many cases were accompanied by morphea[7]. In the present case, the histopathological findings were consistent with LSA and did not show any features of morphea.

In addition, this case is meaningful because the patient improved through treatments including topical steroids, tacrolimus, and narrow band UVB. Although there have been reports about successful treatments of genital LSA with topical steroids and tacrolimus[9,10], there is no established treatment guideline for generalized LSA. As our case showed alleviation of symptoms and improved skin lesions, various treatments including topical steroids, tacrolimus and narrow band UVB can be considered for use in patients with generalized LSA; however, future studies are needed to confirm this result. Herein, we reported a case of generalized LSA successfully treated with a combination of topical steroids, tacrolimus, and narrow band UVB.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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REFERENCES

1. Hallopeau H. [Du lichen plan et particulierement desa forme atrophique]. Ann derm Syph 1887;8:790. French.
2. Fistarol SK, Itin PH. Diagnosis and treatment of lichen sclerosus: an update. Am J Clin Dermatol 2013;14:27-47.
3. Wallace HJ. Lichen sclerosus et atrophicus. Trans St Johns Hosp Dermatol Soc 1971;57:9-30.
4. Hyman AB, Brownstein MH. Lichen sclerosus et atrophicus. In: Demis DJ, Dobson RL, McGuire J, editors. Clinical dermatology. Hagerstown, MA: Harper & Row, 1977:1-7.
5. García-Bravo B, Sánchez-Pedreño P, Rodríguez-Pichardo A, Camacho F. Lichen sclerosus et atrophicus. A study of 76 cases and their relation to diabetes. J Am Acad Dermatol 1988;19:482-485.
6. Meyrick Thomas RH, Ridley CM, McGibbon DH, Black MM. Lichen sclerosus et atrophicus and autoimmunity: a study of 350 women. Br J Dermatol 1988;118:41-46.
7. Uitto J, Santa Cruz DJ, Bauer EA, Eisen AZ. Morphea and lichen sclerosus et atrophicus. Clinical and histopathologic studies in patients with combined features. J Am Acad Dermatol 1980;3:271-279.

8. Kreuter A, Tigges C, Gaifullina R, Kirschke J, Altmeyer P, Gambichler T. Pulsed high-dose corticosteroids combined with low-dose methotrexate treatment in patients with refractory generalized extragenital lichen sclerosus. Arch Dermatol 2009;145:1303-1308.
9. Dahlman-Ghozlan K, Hedblad MA, von Krogh G. Penile lichen sclerosus et atrophicus treated with clobetasol dipropionate 0.05% cream: a retrospective clinical and histopathological study. J Am Acad Dermatol 1999;40:451-457.
10. Hengge UR, Krause W, Hofmann H, Stadler R, Gross G, Meurer M, et al. Multicentre, phase II trial on the safety and efficacy of topical tacrolimus ointment for the treatment of lichen sclerosus. Br J Dermatol 2006;155:1021-1028.