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CORRESPONDENCE

Erythema annulare centrifugum in a patient with stiff-person syndrome

Stiff-person syndrome (SPS), also called stiff-man syndrome, is a rare autoimmune neurological disease characterized by painful episodic spasms and progressive muscle rigidity. Anti-glutamic acid decarboxylase (GAD) antibodies are positive in 60% of SPS, which reduces the production of the inhibitory neurotransmitter gamma-aminobutyric acid (GABA) and causes muscle stiffness.[1] SPS is accompanied by other autoimmune disorders such as type 1 diabetes mellitus, chronic thyroiditis, vitiligo, and pernicious anemia, and it is treated with systemic corticosteroids, intravenous immunoglobulin, muscle relaxants, antiepileptic drugs, and GABAergic agents. Vitiligo is a characteristic skin manifestation, appearing in 16% of patients with SPS.[2] There have been no reports of patients with erythema annulare centrifugum (EAC) accompanying SPS. This case report describes a patient with SPS accompanied by both EAC and vitiligo.

A 65-year-old Japanese woman with a history of type 1 diabetes mellitus, chronic thyroiditis, hyperlipidemia, and lung cancer began to wobble while walking and developed muscle stiffness 19 months before visiting our clinic. A neurologic examination revealed transient muscle stiffness triggered by body movements or mental stress and markedly elevated anti-GAD antibody (86,000 U/mL, normal value <1.5 U/mL). She was found to have vitiligo on both axillae and annular erythema on both thighs (Figure 1A–C). The erythema of the thigh was centrifugally expanded, and the central part was clear. The advancing edge of the erythema was slightly raised and had a trailing scale. The results of serum antibodies-related collagen diseases were all negative (antinuclear antibody <20, anti-dsDNA antibody, anti-ssDNA antibody, anti-SS-A antibody, anti-SS-B antibody, anticardiolipin antibody, and antineutrophil cytoplasmic antibodies). Histopathologic examination of a skin biopsy taken from her right thigh showed mild superficial perivascular lymphocytic infiltration with liquefaction degeneration (Figure 1D). There was no dense perivascular infiltration, which is known as a "coat sleeve" appearance. Based on these findings, we diagnosed her as SPS accompanied by superficial type EAC.

Erythema annulare centrifugum was first reported in 1916 by Darier. Subsequently, EAC was pathologically classified into superficial and deep types by Ackerman, based on lymphocytic infiltration into the epidermis.[3] The pathogenesis of EAC is unclear, but it is considered to be a delayed-type hypersensitivity response to variety of antigens, including malignancy, infections, drugs and other autoimmune diseases.[4] When EAC occurs as a

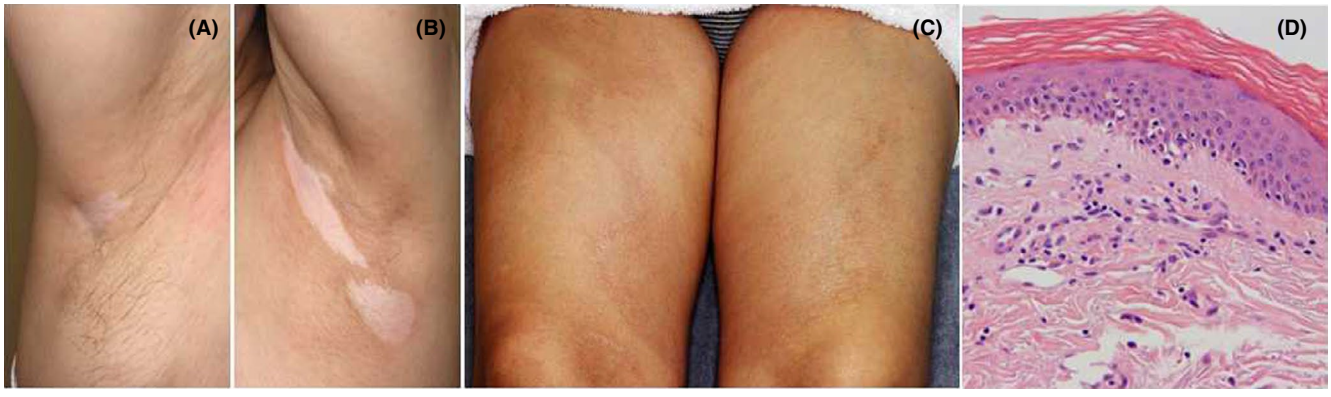

FIGURE 1 (A–C) Photographs of this patient, showing (A, B) vitiligo in both axillae and (C) annular erythema on her thighs. (D) Skin biopsy of the lesion on her right thigh, showing superficial perivascular lymphocytic infiltration with liquefaction degeneration. (HE × 40)

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paraneoplastic phenomenon, it has been designated paraneoplastic erythema annulare centrifugum eruption (PEACE). PEACE typically precedes the clinical diagnosis of malignancy and may recur with subsequent relapses.[5] We did not consider our patient as PEACE because she had suffered from lung cancer before the appearance of EAC and had been no recurrence of it. It seemed that EAC was unrelated to drugs because no new drug was taken or medication change before the onset of EAC. Treatment of diazepam for SPS gradually eliminated her muscle stiffness and EAC on her thighs in parallel. This clinical course may suggest that EAC in this patient was a responsive manifestation to SPS as a so-called dermadrome.

However, in order to discuss the relationship between EAC and SPS, it is necessary to accumulate cases similar to our case.

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DECLARATIONS

Approval of the research protocol : No human participant was involved in this study.
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CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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