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CORRESPONDENCE

A case of IgG4-related skin disease with plaques on the lower legs

A 69-year-old man was referred to our hospital with a 10-year history of nonpruritic brown plaques on the lower legs. Skin examination revealed a soft brown plaque (30 mm in diameter) on the right lower leg and brown-colored indurated plaque measuring 100 mm on the major axis of the left one (Figure 1A,B). Pathological analysis revealed dense infiltrating lymphocytes and plasma cells around blood vessels and fat septum and marked fibrosis extending from the superficial dermis to subcutaneous fatty tissue (Figure 1D,E). More than 70% of Immunoglobulin G (IgG) + plasma cells were found to be IgG4-producing (Figure 1F,G). Furthermore, obliterative phlebitis was observed in the dermis (Figure 1H,I). No lymph follicle formation or eosinophilia was observed. Laboratory test results showed elevated levels of IgG4 (186 mL/dL; normal, 4.5–117 mg/dL). Computed tomography (CT) revealed low-density areas surrounding the thoracic spine and abdominal aorta (Figure 1J,K), without head and neck lesions. The patient was diagnosed with IgG4-related disease (IgG4-RD) according to the comprehensive diagnostic criteria proposed by Umehara et al.[1] Oral administration of corticosteroids (30 mg/day) was initiated and IgG4 level was normalized 1 month after treatment (86.5 mg/dL). Along with decreasing IgG4 levels, the plaques gradually faded and flattened (Figure 1C), and the low-density areas surrounding the thoracic spine and abdominal aorta gradually decreased. Prednisolone was tapered and maintained at a dose of 9 mg/day, without recurrence.

IgG4-RD is a systemic inflammatory disease characterized by elevated levels of serum IgG4 and infiltration of IgG4-positive plasma cells and tissue fibrosis.[2] Some patients with IgG4-RD exhibit skin lesions, which Tokura et al. defined as IgG4-related skin disease (IgG4-RSD).[3] It commonly manifests as pruritic nodules, papules, pustules, and plaques, which are predominantly located on the head and neck.[4] In patients with IgG4-RSD, large plaques on the extremities have not been reported. Herein, we describe a rare case of IgG4-RSD with a large plaque of 100 mm on the major axis of the lower leg. Since topical steroids have less effect on IgG4-RSD, oral administration of corticosteroids is the current first-line therapy, for other organs as well.

The histological features of IgG4-RSD differ slightly from those of IgG4-RD. Regarding IgG4-RD, the number of IgG4+ plasma cells per high-power field (HPF) is often ≥50. However, in patients with IgG4-RSD, these plasma cells rarely exceed 50. Fibrosis is often milder in patients with IgG4-RSD than in those with IgG4-RD. Additionally, obliterative phlebitis has rarely been reported in patients with IgG4-RSD. In our case, we found more than 60 IgG4+ plasma cells per HPF and observed marked fibrosis and obliterative phlebitis, which is a rare case with atypical features. Although the high level of fibrosis in this patient may have been caused by chronic inflammation for 10 years, the precise mechanisms remain unclear. We planned to carefully monitor this patient for relapse while tapering prednisolone.

CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

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

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

Registry and the Registration No. of the study/trial: N/A.

Animal Studies: N/A.

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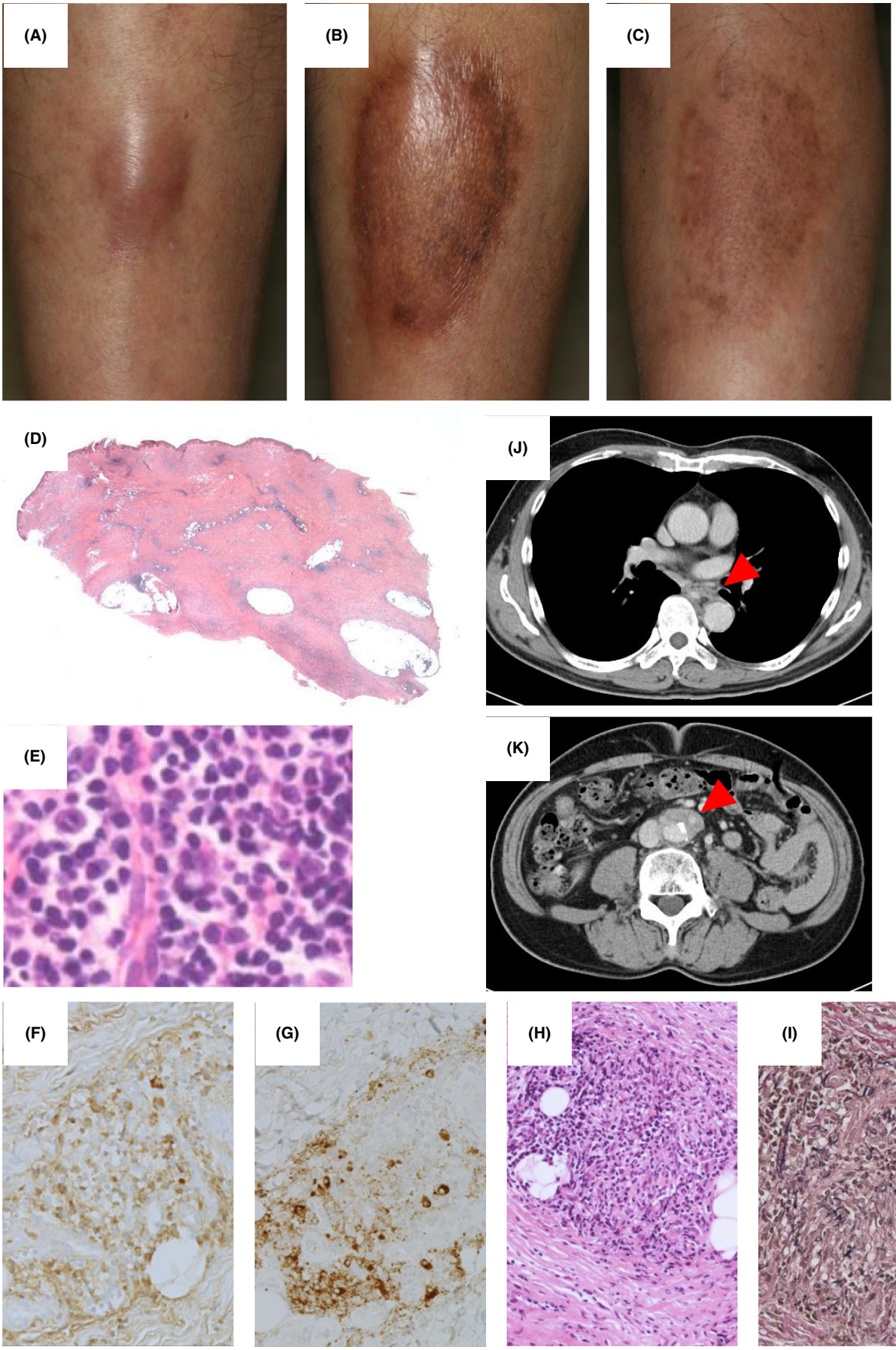
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FIGURE 1 (A, B) Clinical appearance at the first visit. There is a soft brown plaque of 30 mm in diameter on the right lower leg and brown-colored indurated plaque of 100 mm on the major axis of the left one. (C) Clinical appearance on the left lower leg 7 months after starting treatment. Plaques are faded and flattened. (D, E) Histopathological findings of the left lower leg (hematoxylin eosin staining, ×10 (D) and ×400 (E)). Focal and massive infiltrations of lymphocytes and plasma cells are observed. (F, G) Immunostaining for IgG (F) and IgG4 (G). (H, I) Obliterative phlebitis is seen (hematoxylin eosin staining (H) and elastca-van gieson staining (I)). (J, K) CT shows low density areas surrounding the thoracic spine (J) and abdominal aorta (K) (arrowhead).

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