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CORRESPONDENCE

Intertwined vascular skin manifestations in a patient with Sjögren syndrome: A case report

Dear Editor,

Sjögren syndrome (SS) is a common autoimmune disease that exhibits broad organ-specific and systemic manifestations.[1] Here, we report a case of SS with intertwined vascular skin manifestations, in which skin biopsy played an important role in precise diagnosis.

A 78-year-old Japanese woman had suffered from dry mouth for eight years, and been diagnosed with SS based on a positive result for anti-SS-A antibody and sialadenitis one year before the first visit. She had repeatedly experienced multiple palpable purpuras on her lower extremities, too. Six months later, she developed dysesthesia of her extremities and hypanakinesia of her lower legs which resulted in her activities of daily living (ADL) disability. She was diagnosed with mononeuropathy multiplex based on the laterality of neurological symptoms and axonal degeneration of the sensory nerve on a nerve conduction study of her left arm, and treated with a methylprednisolone pulse followed by high-dose oral prednisolone (1 mg/kg/day),

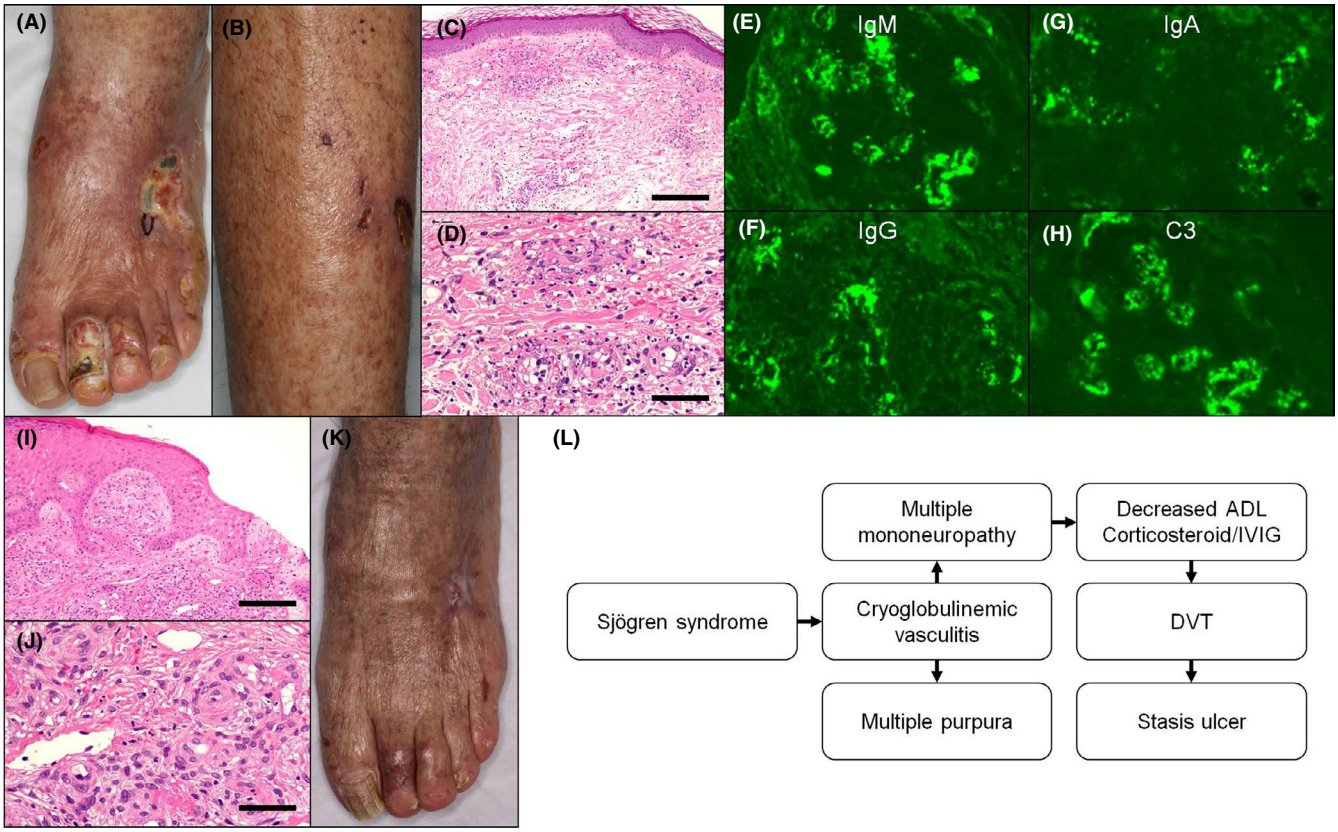

FIGURE 1 Clinical and pathological findings of the present case. (A, B) Multiple palpable purpuras and skin ulcers on the left foot (A), and multiple palpable purpuras and crusts on the right lower leg (B). (C–J) Hematoxylin and eosin staining and direct immunofluorescence of the skin biopsy samples obtained from the palpable purpura (C–H) and the edge of the ulcer (I, J). The bars represent 200 μm (C, I) and 50 μm (D, J), respectively. (K) A clinical image after a 2-month period anticoagulation and local treatments. (L) Predicted pathophysiology of the present case. ADL, activities of daily living; DVT, deep venous thrombosis; IVIG, intravenous immunoglobulin

and repeated intravenous immunoglobulin (IVIG). After tapering the dose of prednisolone, she also developed multiple skin ulcers on her lower extremities (Figure 1A,B). Laboratory tests revealed cryoglobulinemia, a high plasma level of D-dimer (11.3 μg/ml; normal, ≤1.0 μg/ml) and a low plasma level of activated partial thromboplastin time (20.5 s; normal, 24.3–36.0 s). Venous ultrasonography exhibited acute central deep venous thrombosis (DVT) mainly spreading from left external iliac to popliteal veins. Skin biopsy from the palpable purpura revealed leukocytoclastic vasculitis in the dermal small vessels (Figure 1C,D) with depositions of multiclonal immunoglobulin (IgM, IgG, and IgA) and complement C3 on the vessel walls detected by direct immunofluorescence (Figure 1E–H). In contrast, skin biopsy from the edge of the ulcer on the dorsum of the left foot revealed vascular hyperplasia and dilation with bleeding (Figure 1I,J) without deposition of immunoglobulins or complement C3. She was finally diagnosed with cryoglobulinemic vasculitis (CV) resulting in palpable purpura and mononeuropathy multiplex, combined with DVT-induced stasis ulcers. The skin ulcers were almost epithelized with anticoagulation, intravenous injection of heparin followed by oral edoxaban (30 mg/day), and local treatments for 2 months (Figure 1K). Dysesthesia was partially ameliorated by methylprednisolone pulse but hypanakinesia and palpable purpura persistently continued.

In a previous observation of 558 patients with SS, cutaneous vasculitis, one of extraglandular manifestations of SS,[2] was noted in 52 patients (9%).[2] SS patients with cutaneous vasculitis included 14 patients (27%) with CV frequently presenting peripheral neuropathy.[2] Although sensory neuropathy is the most common form of SS-associated neuropathies, mononeuropathy multiplex can occur as a result of vasculitic neuropathy[3] as is the case with our case (Figure 1L). About 15% of and 65% of SS and CV patients developed vasculitic neuropathy, respectively.[4]

Sjögren syndrome patients have a fourfold risk of DVT compared with matched controls.[5] In our patient, ADL disability due to mononeuropathy multiplex, corticosteroid treatment, and IVIG[6] collectively contributed to the development of DVT (Figure 1L).

Sjögren syndrome accompanies diverse vascular skin manifestations as SS-associated extraglandular manifestations. The pathology of each manifestation should be dissected to establish the appropriate treatment strategy.

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None.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

DECLARATION SECTION

Approval of the research protocol: All procedures were approved by the Tsukuba University Hospital Ethics Committee.

Informed Consent: The patient diagnosed at Tsukuba University Hospital was included in this study with written informed consent.

Registry and the Registration No: N/A.

Animal Studies: N/A.

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