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LETTER TO THE EDITOR

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A case of microscopic polyangiitis initially presented with erythema multiforme-like skin eruptions

Dear Editor,

Microscopic polyangiitis (MPA) is a systemic, necrotizing small vessel vasculitis with positive antineutrophil cytoplasmic antibody (ANCA).[1] MPA can involve extracutaneous organs such as lungs and kidneys.[2] MPA shows various cutaneous manifestations; however, erythema multiforme-like eruptions are rarely reported as an initial presentation.

An 83-year-old man presented with a pruritic skin rash and a fever of 38°C following a sore throat of four-day duration. He

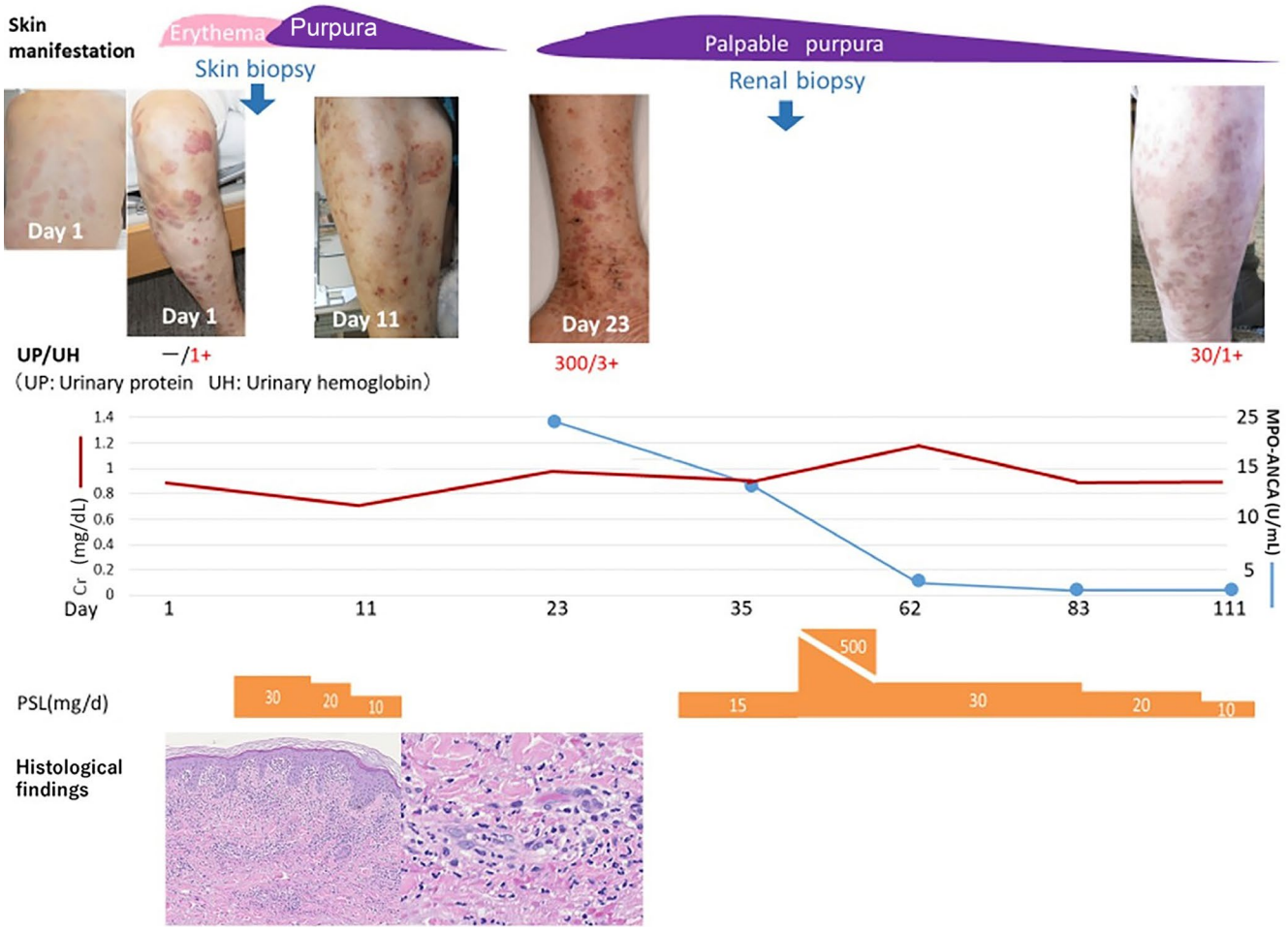

FIGURE 1 Clinical course of the patient. The extent of skin manifestation and represented photographic images are shown at the top. On day 1, well-demarcated erythematous patches of various sizes on the torso and extremities can be noted. Some of the patches showed a bull's eye appearance. The line graph in the middle shows the change in the serum creatinine level, change in the anti-MPO-ANCA antibody titre, and the dosage of prednisolone during the course of therapy. Histological findings at the bottom showed perivascular and interstitial neutrophil infiltration, and cellular debris and erythrocyte extravasation were present in the oedematous papillary dermis and upper reticular dermis at higher magnification

had a medical history of diabetes mellitus, angina pectoris, and prostate cancer. His torso and lower extremities showed well-demarcated, erythematous patches of various sizes, some of which showed a bull's eye appearance (Figure 1). A blood test upon admission revealed a white blood cell count of 9.1 × 10^9/L, neutrophils 6.4 × 10^9/L, eosinophils 0.2 × 10^9/L, blood urea nitrogen 11.5 mg/dL, serum creatinine 0.90 mg/dL, HbA1c 7.8%, and CRP 6.83 mg/dL. Urinalysis showed mild haematuria. The blood test results were suggestive of past infection of Epstein-Barr virus and cytomegalovirus. Anti-mycoplasma antibodies were negative. Erythema multiforme was clinically diagnosed. Oral medications including aspirin, rosuvastatin, sodium rabeprazole, bisoprolol fumarate hydrochloric acid, flivas, sitagliptin phosphate hydrate, and flutamide were all stopped, but the fever remained above 38°C, and the erythematous macules turned purpuric. On day 4, oral prednisolone 30 mg/ day was started, and the fever lowered swiftly, the purpura started to vanish. The prednisolone was tapered off on day 11 with complete resolution of the purpura.

A skin specimen collected on hospital day 1 from an erythematous macule on the lower leg showed perivascular and interstitial neutrophil infiltration, nuclear debris with swollen endothelial cells, and erythrocyte extravasation in the dermis (Figure 1). Neither immunoglobulin nor complement deposition was observed on a direct immunofluorescence test.

On day 23, he had a flare of multiple, palpable purpuras on the lower legs and was placed on oral prednisolone 15 mg/day. A blood test taken on the day was positive for MPO-ANCA at 22.0 U/mL (normal range <3.5 IU/mL), proteinuria (>300 mg), and haematuria. Although we recommended an anew skin biopsy, the patient was not cooperative, and therefore, we diagnosed the patient with MPA based on the previous histopathological finding and clinical presentation. A chest computed tomography did not show any pulmonary involvement. A renal biopsy revealed crescentic glomerulonephritis.

Steroid semipulse therapy (PSL 500 mg/day for three days) followed by oral prednisolone (30 mg/day) ameliorated the skin manifestations, serum creatinine level, and MPO-ANCA titre (Figure 1).

The prevalence of cutaneous symptoms in MPA ranges from 19% to 62.4% in the previous reports.[2–4] The most frequently reported skin manifestation is palpable purpura, followed by erythema, papules, livedo, wheals, and other miscellaneous eruptions. There is one report of erythema multiforme-like presentations during the course of MPA treatment.[5] Our case is unique in that MPA initially presented with erythema multiforme-like eruptions with possible renal involvement suggested by positive urinary haemoglobin early in the course of the disease. To the best our knowledge, this is the first report showing histologically that MPA can exhibit erythema multiforme-like eruptions as the initial cutaneous presentation.

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CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

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