Hemorrhagic Bulla in Churg-Strauss Syndrome

Takashi Irioka¹ and Hidehiro Mizusawa²

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Picture 1.

Picture 2.

Picture 3.

A 58-year-old man acutely developed sensory loss and weakness in the distal limbs. Physical examination revealed skin eruption; palpable purpura was noted in his calves and

feet. In the buttock, medial thighs, and calves, skin lesions progressed to confluent purpura and hemorrhagic bullae, and some became necrotic (Picture 1). Limb pain was so severe that he could not fall asleep. Eosinophilia was noted ($12,141$ cells/ mm^3) with elevated concentrations of C-reactive protein (13.1 mg/dL), immunoglobulin E ($2,247$ IU/mL, normally <20), and eosinophilic cationic protein (114 $\mu\text{g/L}$, normally <14.9). The titer of myeloperoxidase-antineutrophil cytoplasmic antibody was also elevated ($1,350$ EU, normally <20). Nerve conduction study disclosed multiple mononeuropathy. Skin biopsy revealed dermal hemorrhage (arrows in Picture 2) and perivascular eosinophils infiltration (Picture 3). The diagnosis of Churg-Strauss syndrome (CSS) was made, and we treated the patient with corticosteroids [methylprednisolone 1 g/day for 3 days followed by oral prednisolone (60 mg/day for 4 weeks with gradual taper)]. The patient has been in remission for many years with oral prednisolone (3 mg/day) and azathioprine (100 mg/day) treatment. CSS often causes various skin eruptions (1), and can cause hemorrhagic bullae leading to intolerable pain as seen in the present patient (2).

¹Department of Neurology, Yokosuka Kyosai Hospital, Japan and ²Department of Neurology and Neurological Science, Graduate School, Tokyo Medical and Dental University, Japan

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Correspondence to Dr. Takashi Irioka, t-irioka.neuro@ykh.gr.jp

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