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CASE STUDY

Normocomplementemic urticarial vasculitis with laryngeal and intestinal tract edema

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Abstract

An 81-year-old Japanese man presented with a history of recurrent eyelid swelling and purpura on the face, neck, and limbs. Because the initial clinical presentation was angioedema alone, the patient was treated with an H1-receptor antagonist and tranexamic acid as for an idiopathic angioedema. The patient also experienced dyspnea simultaneously with edema on the face and limbs and was thus taken to the emergency room, where laryngeal edema was confirmed on laryngeal fiber. A good response to hydrocortisone injection was observed in the patient. ACE inhibitors were never prescribed for the patient, and there was no family history of angioedema. Laboratory data indicated normocomplementemia, and skin biopsies revealed leukocytoclastic vasculitis. Therefore, the patient was diagnosed with NUV. Following hospitalization, the patient experienced appetite loss and the CRP level increased, presenting with thickening and stranding around colon tissues on abdominal CT. These symptoms responded well to prednisone treatment. Given that the initial clinical manifestation of the current case was mainly angioedema, physicians should consider that angioedema may in rare cases be diagnostic for UV.

KEYWORDS
angioedema, intestinal tract edema, laryngeal edema, normocomplementemic urticarial vasculitis, steroid

Extracutaneous disease is often associated with decreased serum hemolytic complement activity in urticaria vasculitis. Hypocomplementemic urticarial vasculitis (HUV), and HUV syndrome (HUVS) are considered forms of severe vasculitis associated with several systemic findings, including angioedema, pulmonary manifestations, arthritis, arthralgia, glomerulonephritis, and uveitis.[1] Especially, angioedema is the initial clinical presentation in 50% of HUVS patients.[2] By contrast, normocomplementemic urticarial vasculitis (NUV) patients normally have minimal or no systemic involvement and frequently have a better prognosis.[3] We herein present a severe case of NUV with angioedema which exhibited laryngeal and intestinal tract edema.

An 81-year-old man was referred to our dermatology department with a chief complaint of recurrent eyelid swelling and purpura on the face, neck, and limbs (Figure 1A,B). Five years before the first visit, the patient developed swelling on the eyelids and limbs that lasted

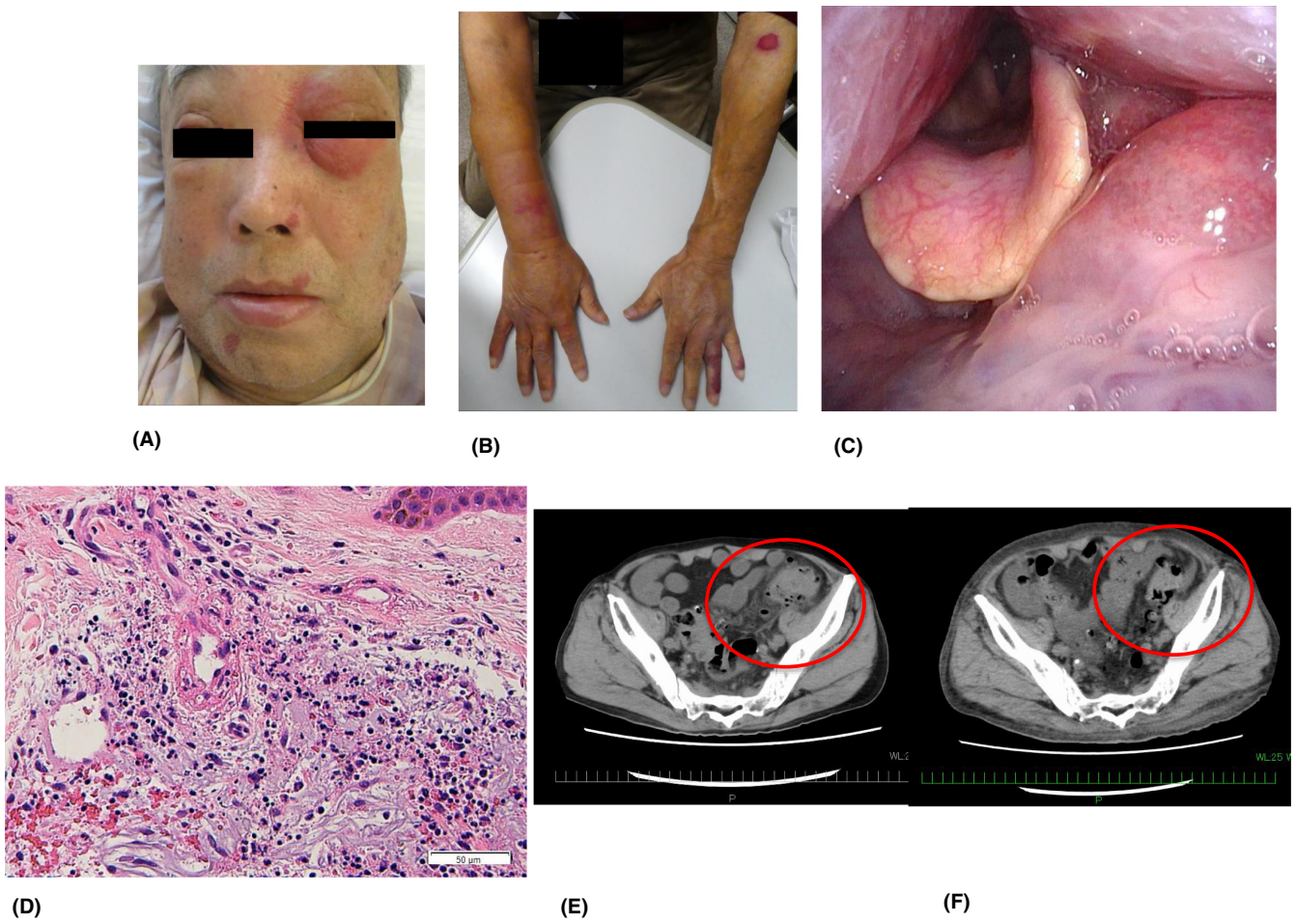

FIGURE 1 Cutaneous manifestations (A, B), laryngeal fiber finding (C), histopathological features (D), and CT findings (E, F). (A) Swelling of the face and hemorrhagic angioedema of the eyelids and lip. (B) Purpura and swelling on the forearm. (C) Epiglottis and laryngeal mucosa display edematous findings. (D) Neutrophils with nuclear dust infiltrated around the small vessel with fibrinoid degeneration in the dermis. (E) Before treatment; the panniculitis around sigmoid colon tissues (red circle). (F) At 28 days after treatment.

for several days. The patient was treated with an H1-receptor antagonist and with tranexamic acid as for idiopathic angioedema. The frequency of symptoms gradually increased, and the purpura persisted long after the swelling had resolved. The patient also experienced dyspnea simultaneously as edema on the face and limbs, and was taken to the emergency room, where laryngeal edema was confirmed on laryngeal fiber (Figure 1C). A good response to hydrocortisone injection was observed in the patient and presented at our department for evaluation of these symptoms. The patient had suffered from chronic renal dysfunction for 15 years because of hypertension. The laboratory tests revealed anemia, with a hemoglobin level of 9.4 g/dL, an increased CRP level of 3.2 mg/dL, and renal dysfunction with a creatinine level of 6.2 mg/dL. The levels of complement (C)3, C4, and C1q and the functional activity of the C1-inhibitor were normal. The antinuclear antibodies titer was slightly increased (1:80), whereas the ds-DNA antibody, Sm antibody, SS-A antibody, SS-B antibody, ANCA, rheumatoid factor, and cryoglobulins were all negative. Testing for hepatitis B and C were also negative. A biopsy specimen from the right forearm revealed leukocytoclastic vasculitis, which was shown by fibrinoid necrosis of the vessel walls, erythrocyte extravasation, and neutrophilic infiltrate with the formation of nuclear dust (Figure 1D). The patient was eventually diagnosed with NUV. Following hospitalization, the patient experienced appetite loss and the CRP level increased to 13 mg/dL. CT examination revealed thickening and stranding around colon tissues (Figure 1E). Initially, the patient was treated with 30 mg prednisone (0.5 mg/kg/day) and the appetite loss and CRP level improved, and the purpura on the face, neck, and limbs was absent after 2 days. One month after treatment, abdominal CT revealed that the thickening and stranding around colon tissues had improved (Figure 1F), and 2 months after treatment the creatinine had improved to 4.4 mg/dL.

We presented here a patient with NUV with laryngeal and intestinal tract edema that responded well to systemic steroid therapy. Angioedema occurs when the vasculitis affects capillaries and postcapillary venules in the deeper layers of dermis or submucosal tissue.[3] Therefore, angioedema comorbidity may contribute to the severity of urticarial vasculitis. Given that the initial clinical manifestation of the current case was mainly angioedema, physicians should consider that angioedema may in sometime cases be diagnostic for UV.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflicts of interest.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT

Approval of the research protocol: B210272.
Informed Consent: Obtained.
Registry and the Registration No. of the study/trial: N/A.
Animal Studies: N/A.

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