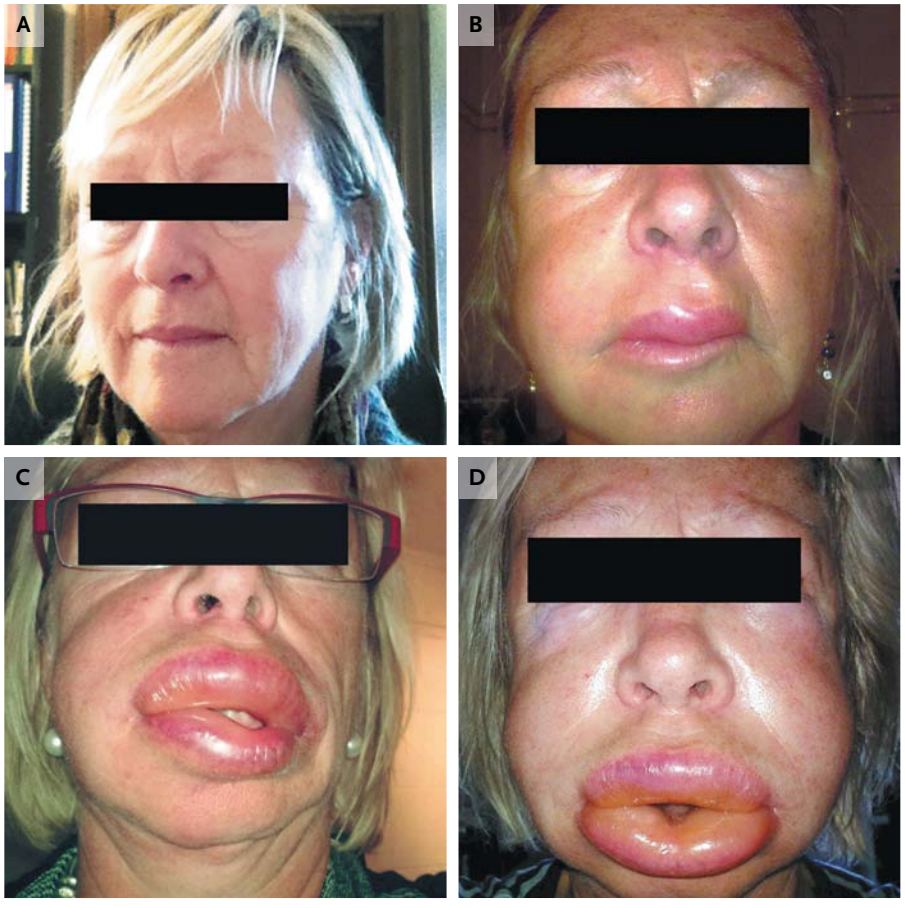
IMAGES IN CLINICAL MEDICINE

Disfiguring Angioedema



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A 54-year-old woman presented to our clinic with progressive swelling of her face. (Panel A shows the patient's face without swelling and Panels B through D show two different episodes of swelling, with Panels B and C showing her face 2 hours and 6 hours, respectively, after the beginning of one episode, and Panel D showing her face 4 to 6 hours after the beginning of another episode.) The patient did not have shortness of breath or difficulty breathing during either episode. The patient had a history of almost 10 years of periodic swelling of the tongue, larynx, trunk, and extremities, starting between the ages of 30 and 35 years. These episodes occurred without prodrome or any discernible cause and generally lasted from several hours to 3 days, irrespective of intake of antihistamines or glucocorticoids. She was not taking an angiotensin-converting–enzyme (ACE) inhibitor or an angiotensin II–receptor antagonist. Complement levels were normal, and functional C1 inhibitor activity was 122%. A tentative diagnosis of type III hereditary angioedema was made. Unlike types I and II hereditary angioedema, type III is not caused by a deficiency of C1 inhibitor but is thought to result from the periodic accumulation of bradykinin through alternative mechanisms. Maintenance therapy with tranexamic acid was initiated, and the patient was instructed to avoid intake of ACE inhibitors and angiotensin-receptor blockers. At 3 years of follow-up, her attacks appear to be both less frequent and less pronounced.

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