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Life-threatening complications following orthognathic surgery in a patient with undiagnosed hereditary angioedema.

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Abstract

As described in the literature, hereditary angioedema (HAE) is an autosomal dominant disease that presents with recurrent events of angioedema caused by a) deficiency or b) functional alteration of the plasma protein C1 inhibitor (C1-inh); this enzyme is involved in the regulation of the complement, kallikrein-kinin, fibrinolytic, and coagulation systems. HAE is characterized by episodes of edema in the larynx, facial structures and tissues, gastrointestinal tract, or extremities. Laryngeal edema has been reported to occur predominantly after oral surgery. We describe the case of an 18-year-old Asiatic male, reporting an unremarkable medical history, who experienced complications following orthognathic surgery. Thirty hours post-op, the patient developed severe facial, pharyngeal, and glottic edema that compromised the airway, and an emergency tracheal intubation was performed. He was tested for C1-inh plasma levels, showing a sub-normal concentration and indicating a diagnosis of HAE. The patient received fresh-frozen plasma and improved throughout the day as his condition stabilized. Several cases of HAE following oral surgery have been reported, but, to the authors' knowledge, this is the first case reported following orthognathic surgery. This patient's treatment will be described, and a literature review of the disease and management methods will be provided.