A Laryngeal Attack of Hereditary Angioedema: A Case Report

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INTRODUCTION: Hereditary angioedema (HAE) is an immunodeficiency disease involving potentially life-threatening bradykinin-mediated attacks of cutaneous, gastrointestinal, or laryngeal swelling. Attacks may be spontaneous or triggered by injury, infection, stress or other factors. Because of its unique pathophysiology, attacks are unresponsive to emergency treatment with epinephrine, antihistamines, or corticosteroids. Due to rarity of the disease, clinicians may be unaware of emergency treatment of HAE.

PURPOSE: Describe an acute laryngeal attack of HAE and discuss etiology, pathophysiology, clinical presentation, diagnosis, and prophylactic and emergency treatment of HAE types I and II.

CASE PRESENTATION: A 70-year-old male presented to the hospital with acute laryngeal angioedema. During diagnostic examination, it was discovered that he had undiagnosed pneumonia and urinary tract infections which likely triggered the attack and also required treatment.

DISCUSSION: The protease, C1-inhibitor, normally inhibits the immune system from spontaneously activating. HAE is an autosomal dominant disease involving deficient (type I) or defective (type II) C1-inhibitor which leads to increased bradykinin and angioedema. Gastrointestinal and laryngeal attacks are the most lethal. HAE presents as recurrent episodes of abdominal pain or angioedema without itching/rash. Diagnosis is made through family history and measurements of C4, C1-inhibitor, and C1-inhibitor functionality. Emergency treatment requires intravenous C1-inhibitor concentrate such as icatibant (Firazyr®). Prophylactic measures include trigger identification/avoidance and possible life-long administration of C1-inhibitor concentrate.

CONCLUSION: This patient presented with laryngeal swelling successfully treated with icatibant, as well as identification and treatment of two infections which triggered his attack. Healthcare provider knowledge of HAE is essential for correct diagnosis and treatment.