Isolated angioedema of uvula (Quincke's disease): A rare presentation

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Abstract
Isolated uvular angioedema (Quincke's disease) is a rare clinical presentation. This condition may be a part of life threatening urticaria and anaphylaxis. Here we report a case of 35 year old male with acute onset isolated uvular angioedema. This case is reported for rare occurrence and management implications.

Keywords: Isolated uvular angiodema, Quincke's disease, Angioedema, Idiopathic.

Introduction
Isolated uvular angioedema, or Quincke's disease, is a relatively rare presentation of angioedema of the upper airway. Majority of cases are of idiopathic origin, while other causes are allergy to food or airborne allergen, drugs (ACE Inhibitors, NSAID), hereditary angioneuroticoedema, oropharyngeal trauma, pharyngotonsillar infection.[1] Swelling of uvula can be a part of generalized oedema of upper airway, as a manifestation of urticaria and anaphylaxis. This condition can lead to upper airway obstruction and respiratory distress, so needs careful evaluation and management.

Case Report
A 35 year old male presented to otolaryngology emergency of our institute with complains of feeling of lump in throat and mild difficulty in swallowing since he got up from bed at early morning. There was no complains of respiratory difficulty, itching or swelling over other part of body. He had history of air travel and alcohol intake along with non-vegetarian dinner in previous night. He was a smoker, taking 1 packet of cigarette per day since 5 years. He had no known history of allergy to any drug or food. There was no history of co-morbid medical condition. And family history was negative for allergy, asthma, urticaria, hereditary angioneuroticoedema. His vitals were stable at presentation, blood pressure 128/86 mm of Hg, pulse 78/min, respiration rate 16/min. Systemic examinations were unremarkable. Oral cavity and oropharyngeal examination revealed large, oedematous uvula with normal appearing tonsil and posterior pharyngeal wall (Fig. 1). Indirect laryngoscopy examination showed normal epiglottis and vocal cords. Routine blood investigation showed total leukocyte count of 7000/cmm, with normal differential count. Liver function test and renal function test were normal. Patient was givendexamethasone injection, pheniramine maleate injection in emergency ward and observed for 1 day and discharged on next day on oral anti-histaminics for 48 hours as his general condition was stable. On 4th day follow up patient was asymptomatic and uvula was appearing normal.

Discussion
Swelling of uvula can be a presentation of urticaria and anaphylaxis. When associated with generalized swelling of pharynx and larynx, the condition could be life threatening, by compromising airway. Though swelling of uvula in isolation is rare, it can be due to varied etiology, among which idiopathic is most common.[1] Other causes being, allergy to food or airborne allergen, hereditary angioneuroticodema, trauma etc. The patient in the present report had isolated angioedema of uvula, without any urticaria, and responded well to steroid and antihistaminic. As the patient had heavy alcohol intake in the previous night, he may have taken some food allergen, though patient denying any past history of allergy to any food material.

Patients with hereditary angioneuroticoedema may have associated uvular oedema. But the condition typically affect young adolescent age group,[2] swelling episodes worsens over one to two days then resolves over next two days,[3] more than 90% patient experience abdominal pain,[4] positive family history may be present.[5] Hereditary angioedema responds to C1 esterase inhibitors and kallikrein inhibitors and not to glucocorticoids.[6] As the present case was not fitting to clinical criteria for hereditary angioedema and he responded well to glucocorticoid and anti-histaminics, so we did not go for complement level assessment.

Conclusion
In patients with uvular oedema, presence of concomitant epiglottitis must be ruled out by laryngoscopic examination and/or X-ray soft tissue neck lateral view, so as to avoid grave complications arising out of latter. Initial management must address to maintain a safe airway.

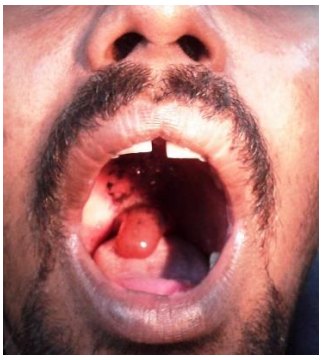

Fig. 1: Oropharyngeal examination shows grossly oedematous uvula. (Patient consent was taken for photograph)

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