

CASE REPORT

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# Quincke's disease: a case report

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## Abstract

**Background:** Isolated angioneurotic edema of the uvula is termed Quincke's disease. It is a rare clinical disorder of acute onset with few known causes. It may be encountered in any emergency setup and must be dealt with rapidly and with utmost vigilance for prevention of progression and complications.

**Case presentation:** A young adult, 3 months post Frey's procedure for chronic pancreatitis, presented with acute onset throat discomfort and gagging progressive over 8 h. Examination showed isolated edematous hyperemic uvula with normal oropharyngeal structures and adequate airway. Prompt antihistaminic and corticosteroid therapy caused relief of symptoms over 2 h with no recurrences.

**Conclusion:** Any symptom suggesting orofacial edema must not be trivialized. Awareness about this rare but acute condition, even in the background of unknown etiology, in all medical personnel is essential. Early diagnosis with appropriate management can prevent life-threatening airway obstruction and hypoxemia.

**Keywords:** Case report, Quincke's disease, uvular edema, Angioedema

## Background

Quincke's disease or isolated angioneurotic edema of the uvula is a rare clinical entity with various etiologies reported but not many substantial underlying causes defined.

Being a rare condition, epidemiology of Quincke's disease has not been documented in literature; however, multiple etiologies have been explored and defined over the last 2 decades [1–6]. The etiology of isolated angioedema of the uvula is usually an immediate type I hypersensitivity reaction by trauma, thermal injury, infections or drugs, and even idiopathic in some cases [7]. As the name indicates the condition is not associated with other hypersensitivity reactions such as skin rash, hypotension, or tachycardia and occurs as a localized, non-pruritic subcutaneous swelling that comes on rapidly. The kinnin and complement pathway mediated type 1 hypersensitivity has been implicated by many authors as the pathophysiological basis of Quincke's disease, although exact mechanism in case of idiopathic Quincke's edema, such as in our case, is not known [6].

Owing to allergic nature of the disease, investigations such as complete blood picture with total and differential leucocyte counts along with Ig E levels have been given importance in the past. Mast cell tryptase at 1–2 h and 24 h after onset have been suggested by Patel et al [8] to substantiate the pathogenesis suspected. Markedly reduced C1 esterase levels in such patients indicate a hereditary etiology. Airway management is the primary strategy in all cases of Quincke's edema. Suggested treatment plan includes continuous monitoring, oxygen therapy and epinephrine if indicated with H1 and H2 blocking drugs and dexamethasone as steroid therapy [9–11].

Due to its acute spontaneous presentation and potential for airway compromise, there is a need for study of this condition in its rare idiopathic background and awareness about the appropriate management that must be undertaken. The following case report aims to study a rare case of idiopathic Quincke's edema in the light of unknown etiology.

## Case presentation

An eighteen-year-old boy with 3 months post Frey's procedure for chronic pancreatitis came for routine check-up to the surgical team with the complains of throat

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discomfort with foreign body sensation since 8 h. Upon check-up by the on-call ENT team, patient reported feeling “something touching the back of the tongue” each time he swallowed that made him gag a few times. He refused any associated throat pain, cough, fever, or difficulty breathing. Family members reported slight nasality in the patient’s voice since that morning. Patient was on tablet tramadol one dose taken the previous day, on tablet pancreatin 170 mg and tablet dimethicone 80 mg daily since 3 months and had no allergic responses to those drugs. There were no other history of known food, drug, or inhaled allergies or exposures; no history of consumption of sea food/nuts the previous day; and no family history of angioedema. His vitals were stable. Respiratory examination was normal. Local examination showed grossly edematous uvula of about  $4 \times 1$  cm size projecting beyond the base of tongue with no other oropharyngeal abnormalities as shown in Fig. 1. Image to left shows isolated edema of the uvula with normal oropharyngeal structures; (top right) hyperemia and edema of uvula magnified image at presentation can be seen (bottom right) after single dose of intravenous dexamethasone and pheniramine maleate when patient reported symptomatic relief. Airway looked adequate with normal epiglottis and normal vocal cord movements. No urticarial rash or hives on physical examination. Complete blood picture had revealed mild leukocytosis (total white blood cell count,  $11,500/\text{mm}^3$ ) Neutrophilia with normal eosinophil counts. Erythrocyte sedimentation rate (ESR) was 21 mm/h, CRP 6 mg/L, and total IgE levels of 290 IU/mL. C1 esterase inhibitor level, functional C1 inhibitor activity, complement C3 and C4, and radioallergosorbent test (RAST) could not be done by the patient due to financial constraints.

Patient was treated with single doses of injection intravenous dexamethasone (8 mg) and pheniramine hydrogen maleate immediately and observed for any signs of distress or excessive drooling. Patient had relief of symptoms in 2 h and uvular edema completely resolved after 24 h with no adverse events or deteriorations. Antihistamines were prescribed for 3 days thereafter. Subsequent follow-ups showed completely normal uvula with no further recurrences.

## Discussion

First described by Quincke in 1882 [12] and thence named after him, isolated non-hereditary uvular edema or “Quincke’s disease” is a rare condition reported in both adult and pediatric ages [9]. The clinical manifestations of Quincke’s disease described by Milton in 1876 and its mechanism by Heinrich Quincke in the late 1800s, the manner of development of this condition, i.e., acute localized swelling of the skin and mucous membranes, it was thereafter considered as an angioneurosis. It is a unique form of angioneurotic edema of the upper airways where a patient presents with symptoms like foreign body sensation of throat, cough, gagging, voice change, or breathing difficulty in an acute manner in the absence of any other constitutional symptoms like fever or rash. As such, it may be easily missed or refuted by medical professionals in its early stages and may worsen rapidly.

Only a handful cases with Quincke’s disease have been reported and hence an exact epidemiology of the condition is not yet known. Underlying etiologies explored and reported in literature until now include food allergies such as nut or prawn [1] infections, mechanical pressure/trauma regional [2] or general anesthesia [9], herbal agents



**Fig. 1** (Left) Isolated edema of the uvula with normal oropharyngeal structures, (upper right) magnified image of the oropharynx at presentation, (lower right) and magnified image of oropharynx when patient reported symptomatic relief

[3], drugs such as ACE inhibitors [4], or ARB [5]. However, idiopathic etiology is rarely reported [6].

It is usually caused by type1 hypersensitivity reaction [2]. The pathogenesis behind the inflammatory edema is thought to be mediated through the complement system which can be through the classical pathway in which C1 is activated by an antigen-antibody complex to form C1 esterase, which binds C4 and C2. This activates C3 followed by the rest of the sequence leading to cell lysis. Hereditary form of Quincke's disease probably follows this pathway due to deficient C1 esterase inhibitor which is the main regulator for both kinnin and complement pathways. In the alternate pathway, however, there is direct activation of C3 endotoxins, polysaccharides that ultimately leads to production of anaphylotoxins which cause increased vascular permeability by histamine release through mast cell degranulation. This maybe the mechanism in conditions such as drugs, food allergens, or anesthetic agents that cause Quincke's disease. However, in our case the etiopathogenesis seems idiopathic as the exact cause of this reaction in our patient could not be specifically attributed to any identifiable inciting agent. The patient was on long-term treatment with the same set of medications and 3 months post a definitive surgery. Hence, hypersensitivity to drugs or anesthetic agent did not seem to be a causative factor here. A delayed drug response to his treatment regimen was still kept in mind, but after the initial management he did not show any recurrence in our long-term follow-up while on the same medications. As C1 esterase levels were not measured here, the possibility of hereditary C1 esterase deficiency was still considered. Lack of any recurrence in our long-term follow-up of this patient with no direct correlating or precipitating agents identified denoted a more likely case of idiopathic etiology.

Investigations for probing into the allergic nature of the disease including CBC, ESR, skin prick test, and RAST must be done. In recurrent cases, C1 esterase levels and mast cell tryptase levels need to be measured. Although some of the tests were done in our case, C1 esterase inhibitor level, functional C1 inhibitor activity, complement C3 and C4, and RAST could not be done due to unavailability of sufficient finances and mast cell tryptase levels of 1–2 h and 24 h levels that need to be measured immediately at onset for baseline had not been done in our case and it did not seem instructive to check the levels several hours after the reaction when the patient presented. Direct visualization or a lateral neck plain radiograph should be considered to help rule out epiglottitis and throat swab can be undertaken if in doubt of any infective etiology.

Quincke's must not be confused with a similar presenting entity: Isolated uvulitis where the primary management includes antibiotic therapy owing to its infective etiology.

Complications and consequences of this condition could be morbid, even lethal in some instances. Due to the bulk of edema and sudden acute onset, the uvular edema can cause obstruction to airway passages. The most dreaded and potentially fatal manifestation of Quincke's disease is the oropharyngeal edema which can potentiate into a lethal airway obstruction occurring as a result of the uvular edema to develop into glottis obstruction and laryngeal edema. This may cause airway compromise and life-threatening hypoxemia [4].

Recurrent or refractory Quincke's edema can also cause snoring or sleep apnea owing to the subpar airway conditions in the long term, giving rise to a chronic disorder with potentially fatal outcomes. It may also be an early presenting feature of hereditary C1 esterase deficiency which when missed can cause further angioneurotic stridor and that is refractory to standard treatment.

The immediate treatment of uvular edema depends on the degree of airway compromise and patent airway maintenance remains the most predominant primary strategy. In emergencies, intravenous H1 and H2 blockers, corticosteroids, and even epinephrine may be necessary. The resuscitation kit with an anesthetist team must always be ready on cue and facilities for endotracheal intubation and cricothyroidotomy should be available. After ensuring an airway devoid of obstruction, the underlying cause should be identified.

For patient with posttraumatic or inhalant uvulitis, steroids must be administered. Medications used to reduce swelling include epinephrine [9], diphenhydramine, cimetidine, and steroids. Dexamethasone has been considered the medication of choice considering its potent anti-inflammatory properties and long half-life [13].

Patients with suspected noninfectious cause, who do not respond to the above medications, may have a complement deficiency and should also receive plasminogen inhibitor e-aminocaproic acid [14, 15]. This hereditary etiology must be borne in mind especially in recurrent and refractory cases and measures such as fresh frozen plasma and beta epsilon amino caproic acid have all been tried in such conditions.

Symptoms usually resolve completely within 24–48 h; however, close observation of the patient for signs of respiratory distress or recurrence are necessary.

Recurrent uvular edema attacks have been attributed to underlying hereditary angioedema, acquired C1 esterase inhibitor deficiency, or can rarely point towards malignancy of parapharyngeal space as reported by Wareing et al. [7]. As this patient did not have any recurrent episodes, there was no necessity for evaluation with serum C1 esterase levels at that stage and hence not undertaken. However, owing to young age, the patient has been explained his condition and kept on long-term follow-up for the same.

Very rarely the invasive techniques of securing airways in the form of needle decompression may be required. Surgical management suggested includes uvular debulking with uvulotomy or uvulectomy [16] by lacerating or trimming the distal end of uvula and have been attempted by Evans et al. in 1987 [10]. But since there are no further reports regarding the same, surgical management owing to its high morbidity has a minimal to nil role in management protocol.

In our case, as the etiology was idiopathic, the mainstay of treatment was dexamethasone and H1, H2 histamine blockers which resolved the symptoms significantly. Overall prognosis of Quincke's disease is good and recurrence rates are low.

## Conclusions

Quincke's disease is a rare form of isolated angioedema of the uvula and etiologies are largely unknown. Investigations should include workup for allergy parameters and complement markers. Etiopathogenesis of the condition in the idiopathic background is less understood; however, complement mediated hypersensitivity is largely accepted as the mechanism in the allergic etiology of Quincke's edema. Symptoms such as acute onset foreign body sensation throat should not be dismissed and evaluated appropriately by otorhinolaryngologists. Awareness of this condition is necessary for early management. It can cause life-threatening hypoxemia through obstruction by glottic or laryngeal edema. Airway maintenance is the first priority with appropriate preparedness for invasive procedures. Dexamethasone is the treatment of choice. Epinephrine, H1, and H2 blockers are mainstay of treatment. FFP or beta epsilon amino caproic acid can be considered for C1 esterase deficiency cases refractory to all other treatment. Surgical debulking may be considered in recurrent or refractory cases. Further understanding of this rare entity, especially in the background of nonspecific etiology, is still in evolving stages and hence a thorough workup in all cases is quintessential.

## Patient perspective

Morning of my visit, I felt a lump-like sensation at the back of my throat more pronounced when I'd swallow. I mentioned it to my mother, but she scoffed it to be nothing. Over the next couple hours, it got worse making me gag a lot. Soon after the initial valuation, I was told my uvula was swollen and injections were given to me by the ENT doctor on call. The symptoms started to reduce, and the feeling completely went away in about 2 h.

## Abbreviations

ESR: Erythrocyte sedimentation rate; CRP: C reactive protein; RAST: Radioallergosorbent test

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## Authors' contributions

RRK done the literature search and is the guarantor. JSR and RRK done the concepts, design, definition of intellectual content, data acquisition, data analysis and manuscript preparation, editing, and review. All authors made corrections and approved the final manuscript.

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## Availability of data and materials

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## Ethics approval and consent to participate

There were no ethical issues in the case report. Informed consent was taken from the participant.

## Consent for publication

Consent for publication was obtained from the participant.

## Competing interests

The authors declare that they have no competing interests.

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