
Isolated Uvular Angioedema in a Teenage Boy

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Citation

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Abstract

Isolated angioedema of the uvula, also known as Quincke's disease, is a rare presentation of angioedema, requires prompt therapy and careful monitoring of symptom improvement because of the potential for airway compromise. Other possible etiologies for uvular swelling should also be considered: hereditary angioneurotic edema, localized trauma, inhalation injuries or exposures, adverse reactions to medications, and infectious causes. We present a case of isolated uvular swelling in a teenage boy, without a clear inciting event or cause, which resolved with aggressive therapy for angioedema.

CASE REPORT

A 17-year-old teenage boy was transferred to our institution's emergency department (ED) from an outlying hospital with a presumptive diagnosis of "swollen uvula." He had a history of acute throat pain, neck swelling,odynophagia, dysphagia and difficulty breathing that started that morning when he awoke, approximately 6 hours before his arrival to the ED. The patient noticed that his uvula was red and swollen when he looked at his throat in the mirror. His symptoms improved somewhat by leaning forward.

He had a history of asthma as a younger child, but had not used his albuterol inhaler in over five years. He was taking no medications and had no known drug or food allergies. He also denied any history of trauma, tobacco or illicit drug use, or any inhalant use or exposure. The patient also complained of some mild nasal congestion, rhinorrhea, slight nausea, and subjective fever. He denied cough, vomiting, diarrhea, toothache, headache, chest pain, rashes, or pruritus.

The patient was initially noted by the ED triage nursing personnel at the referring hospital to be "unable to speak." However, on exam, he was noted to be afebrile, with a normal respiratory rate, heart rate, blood pressure, and room air oxygen saturation. He had no respiratory distress, drooling, wheezing, stridor, trismus, or gagging. His cardiopulmonary exam was unremarkable, but his oropharyngeal exam demonstrated a swollen, erythematous, and elongated uvula without exudate or discharge. He had normal dentition, gingiva, tongue, palate, and tonsils. He had no urticaria, and no face or neck swelling.

The outside emergency department administered a subcutaneous dose of terbutaline, and intravenous doses of dexamethasone, famotidine, diphenhydramine, and clindamycin to treat for a possible allergic or infectious etiology of his symptoms. His throat pain was treated with meperidine. He had a lateral neck plain radiograph that revealed no epiglottis swelling, no foreign body, or increased swelling of the prevertebral soft tissue. Computerized tomography of his neck and a complete blood count were also interpreted as normal.

Upon arrival to our ED, he had normal vital signs, his pain was much improved, but he still felt that his uvula was swollen. He was able to recline on the hospital gurney without difficulty. His uvula was still noted to be swollen, erythematous, and elongated enough to be draped over the posterior aspect of his tongue. (Figure 1) He received an additional dose of dexamethasone, scheduled diphenhydramine every 6 hours and was admitted to the inpatient ward for overnight observation. The presumptive admitting diagnosis was isolated uvular angioedema from an allergic reaction, but a C4 complement level was also drawn to help exclude the possibility of an initial presentation of C1 esterase deficiency. His hospital stay was uneventful and his uvular changes completely resolved within 18 hours after admission. His C4 level was 24 mg/dl (normal range 11-61 mg/dl) and he was discharged with an epinephrine auto-injector and instructions to continue taking his scheduled oral antihistamine for the next 48 hours.

Figure 1



DISCUSSION

Isolated angioedema of the uvula or Quincke's disease of the uvula, is a relatively rare presentation of angioedema of the upper airway. It is characterized by acute swelling of the uvula, which may come to rest on the tongue. The associated signs/symptoms include: throat pain, odynophagia and/or dysphagia, difficulty with handling oral secretions, gagging, hoarseness, and often a sense of throat "tightness or fullness."

The etiology of isolated angioedema of the uvula is usually due to a Type 1 hypersensitivity reaction. It is usually characterized by recurrent, localized, non-demarcated, non-pruritic subcutaneous swelling that appears rapidly and resolves within hours to days.

Fever and associated surrounding cellulitis of the uvula warrant an investigation for infectious causes. Case reports have described isolated uvulitis with Group B streptococci, *Haemophilus influenzae* type B, *Candida*, and anaerobic bacteria as probable etiologic agents. Associated tonsillopharyngitis and epiglottitis have also been acknowledged as potential complications.^(6,7,14,15)

Causes for isolated uvulitis, other than allergic and infections, include:

1. post anesthesia and deep sedation, both with and without endotracheal intubation,
2. secondary to airway instrumentation
3. medications and drugs: including angiotensin-converting enzyme inhibitors, after
4. marijuana and cocaine use (3,4,7,8,9), and after the use of juice from *Ecballium elaterium*, a plant used

as a homeopathic remedy for sinusitis. (10,18)

5. Snoring or obstructive sleep apnea (16,17)

Hereditary angioedema and C1 esterase inhibitor deficiency are more commonly causes for repetitive/chronic episodes of angioedema.⁽¹⁾

Direct visualization is always crucial in the initial evaluation of the patient presenting with isolated uvular angioedema if the patient's airway protective reflexes are intact.

A lateral neck plain radiograph should be considered to help rule out epiglottitis, since several previous reports of uvulitis occurring concurrently with *Haemophilus influenzae* type b epiglottitis have been described.^(6,7)

Screening for C1 esterase inhibitor deficiency can be accomplished by checking C3 and C4 levels. A normal C3 level and low levels of C4 will suggest C1 esterase deficiency, with C1q levels distinguishing between acquired and inherited forms. Unfortunately, acquired C1 esterase deficiency has a high concordance rate with other autoimmune and lymphoproliferative disorders, necessitating further studies.⁽²⁾

Management priorities focus on maintaining a patent airway with proper positioning, utilizing more invasive and/or surgical techniques if necessary, along with parenteral H1 and H2 histamine blockers and corticosteroids. Parenteral epinephrine can also be used with patients in extremis.^(1,7) Dexamethasone has been considered the medication of choice considering its potent anti-inflammatory properties and long half-life.⁽³⁾ Documented cases of post-anesthetic uvular edema treated with diphenhydramine have assumed an allergic etiology.⁽⁴⁾ There are also rare case reports of topically applying epinephrine directly on the uvula itself or performing a partial uvulectomy for relief of airway obstruction.^(1,5,9)

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