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Make Your Diagnosis

Funky Quincke's for Make Your Diagnosis Category

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A 20-year-old woman, 20 weeks gravid with twins, with no history of asthma or allergies presented to the emergency department with symptoms of hyperemesis gravidarum. She was admitted to the obstetrics floor for treatment.

The day after admission, she complained of a sore throat; at that time a nurse's examination of the oropharynx was negative. Later in the day, acutely, she began to complain of dyspnea, dysphonia, and globus sensation. She received intravenous fluid hydration and antihistamine to lower stomach acidity (Famotidine). She denied recent antibiotics or change in medication, inhalation or ingestion of drugs, trauma, or smoking.

On examination she was in acute distress and in tripod position. Heart rate was 110 beats/minute and respiratory rate 28 breaths/minute; oxygen saturation (pulse oximeter) was 98 percent. Lungs were clear without stridor or wheeze. However, examination of her mouth showed isolated severe swelling of the uvula, 2 x 1.5 cm by visual estimation, without erythema, post nasal drip, hemorrhage or exudate. The pharynx and the soft tissue surrounding it were without swelling or edema, Fig. 1.

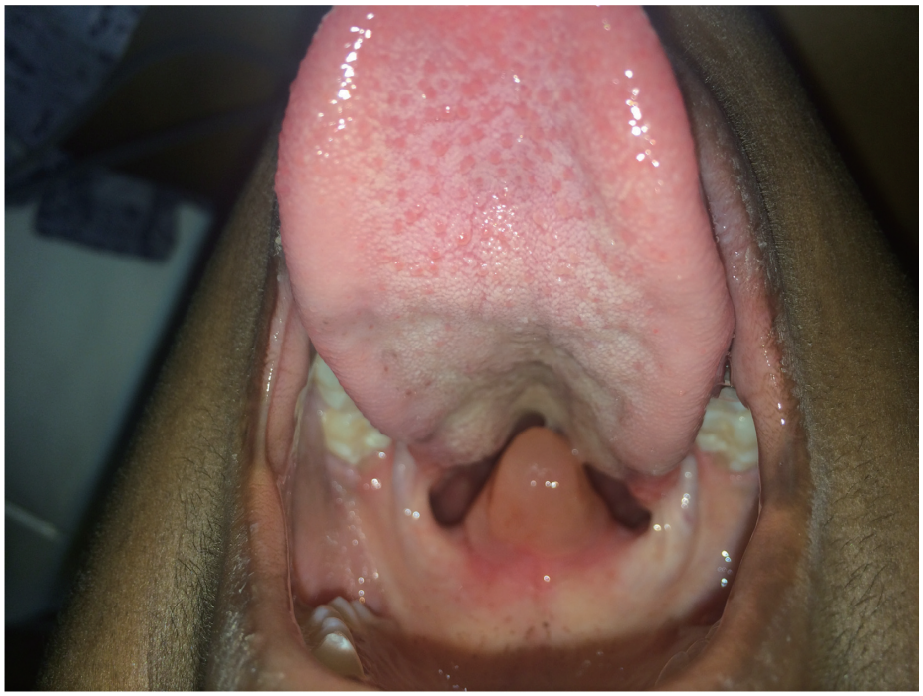

Fig. 1. Uvula size and normal pharynx appearance at the time of initial patient's symptoms.

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She was given epinephrine, antihistamine (H1 and H2 blockers) and corticosteroids but there was no improvement in symptoms. She was then transferred to the ICU where her breathing, drooling, and inability to speak worsened. She was intubated in a controlled environment by the critical care physician with direct video laryngoscopy using C-MAC 4. Her epiglottis and vocal cords appeared normal.

Her respiratory viral panel tests, influenza swab, MRSA nasal swab and rapid Strep tests were all negative. The patient was tested for underlying hereditary causes like hereditary angioedema type I (C 1 esterase inhibitor deficiency) and type II (atypical C1-inhibitor proteins). Her serum complement factor 4 (C4), C1 inhibitor (C1-INH) antigenic protein and C1 inhibitor (C1-INH) functional level came back negative. Ceftriaxone and Azithromycin were started covering the typical and atypical most common respiratory infectious organisms like Legionella pneumophila, Mycoplasma pneumoniae, Chlamydophila pneumonia, Haemophilus and Streptococcus species were given. Her antibiotics were switched to a broader coverage with Piperacillin-Tazobactam to cover intra-abdominal source since her initial complaint was nausea vomiting and abdominal pain. After 3 days the swelling resolved and the patient was extubated, Fig. 2.

Her presentation was consistent with isolated uvular angioedema (Quincke`s disease), with unknown etiology. Quincke's disease is a rare condition that can be caused by mechanical trauma, chemical or thermal injury, medication reactions, infection, or hereditary angioedema. It is an acute medical emergency because it can cause airway compromise, especially in high risk patient as our pregnant lady with her twins. Although no established guidelines for treatment here in the USA, usual management consists of initial empiric antibiotics for possible common infection, corticosteroids, antihistamines, epinephrine and, if necessary, intubation. Refractory swelling not responding to the medical treatment can be managed with surgical intervention and uvulectomy. With rapid and appropriate treatment patients can be expected to have full recovery.

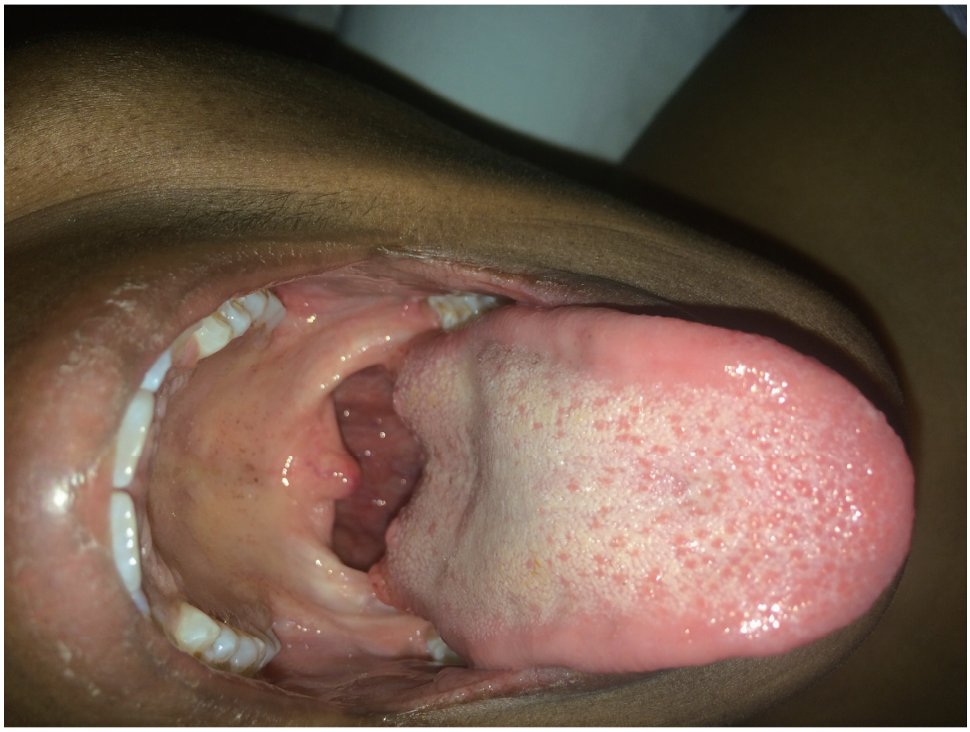

Fig. 2. Uvula size 4 days after initial patient's symptoms and one day after extubation.