Reader's Case Forum: How would *you* manage this?

Sudden Macroglossia -- Diagnosis and Management.

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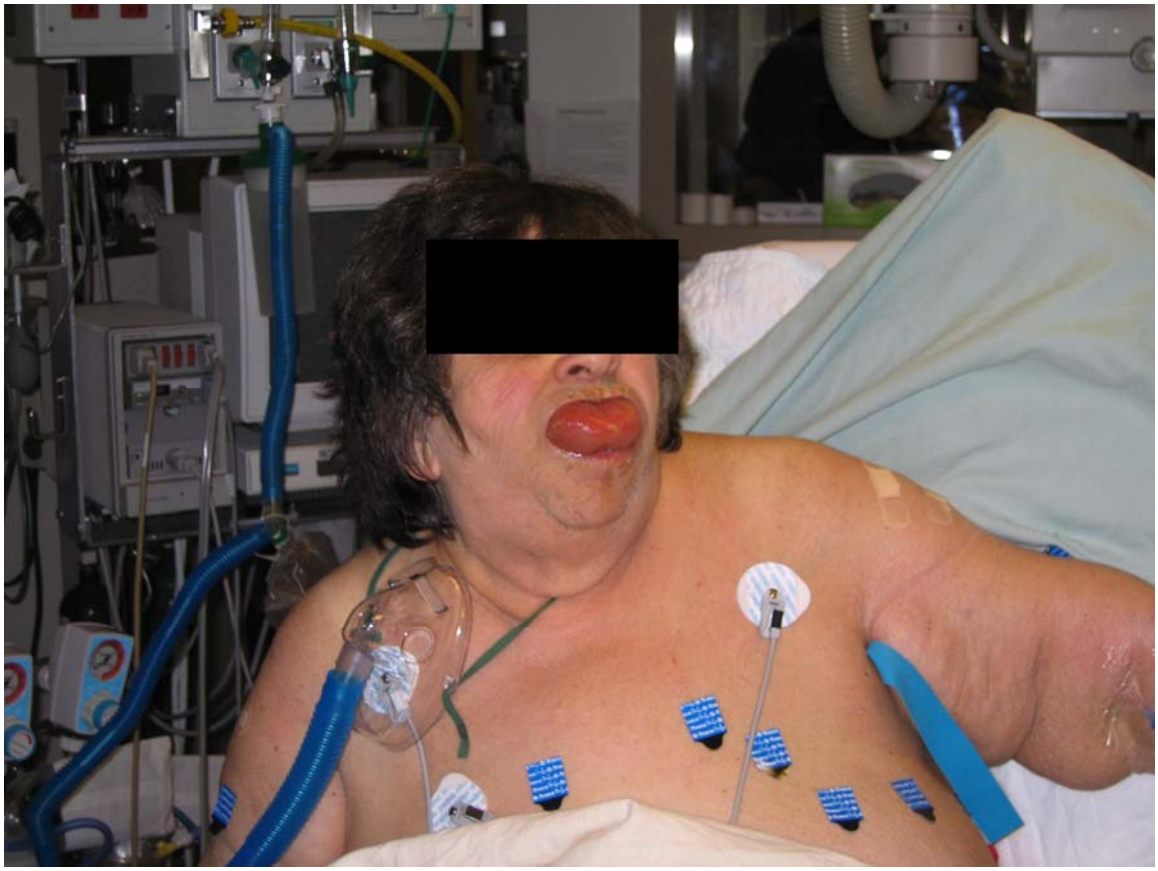
MsSH Words: Macroglossia, Angioedema, Airway, ACE Inhibitor

Case History:

A seventy-six year old woman presented to the Emergency Department. The nursing home where she lives was concerned that her tongue had swollen suddenly over the past two hours to the point where she could no longer speak, let alone take anything by mouth. She was not reported to have any difficulty breathing, rash, vomiting, fever, or mental status changes.

Her past medical history included hypothyroidism, diet controlled diabetes, and hypertension for which she was treated with levothyroxine and enalapril for the past several years. She was on no new medications. There was no history of angioedema in the past, no known trauma or dental problems or treatments recently.

On presentation the patient was accompanied by EMS who reported hemodynamic stability during transport and no evidence of respiratory distress or hypoxia. She was obese, upright and seated, cooperative, but unable to communicate due to massive macroglossia (see figure).



Her skin was pink and dry there was no rash or facial swelling. Blood pressure was 145/92, heart rate 89, temperature 36.5 (97.8) and pulse oxymetry showed a saturation on room air of 95%. There was good air entry to the lungs via nasal breathing. No stridor or wheeze was noted. Heart sounds were regular without murmur or rub, and the abdomen was soft and nontender. She had no focal neurological deficit, and no signs of injury.

Her tongue completely filled the oral cavity and protruded. There were no signs of hematoma and no tenderness. Direct examination of the oropharynx was precluded by the massive tongue.

What is the diagnosis?

Angiotensin Converting Enzyme Inhibitor (ACE-I) induced angioedema.

But she has been on enalapril for years?

This entity may appear any time after initiation of treatment with ACE-I. It typically appears only after months of treatment. It is uncommon (perhaps 0.1% of patients treated with ACE-I get it) (1) but not rare given the prevalent use of ACE-I. The cause is not known, but may related to bradykinin and a concomitant inflammation. (2)

Is this an allergy?

The exact mechanism is not known but it is not an IgE mediated phenomenon as in anaphylaxis. In one study it was the most common cause of upper airway angioedema presenting to the ED (3).

Is it serious?

In the latter study 18 of 45 (40%) of patients required ICU placement, of these 5 required intubation (two nasally, one orally and one required cricothyrotomy). There was no

mortality. (3) There may be some predisposition in patients with previous.

What is the duration of the problem?

The key to stopping the process is to stop the medication causing the problem. However once the angioedema is present what is the natural course, when is the peak effect, and how long does it last? There is no clear answer in the literature, except that it lasts for hours to days, can relapse and can progress to the need for intubation to insure the airway, although most cases will not.

What to do in the ED?

The first question of course is of immediate respiratory compromise. If this is present then immediate airway intervention is necessary and this will depend on the clinical scenario: Is there a reasonable likelihood of success of oral or nasal intubation? Is there time available for preparation due to respiratory compromise or is cricothyrotomy the fastest method available. Is there time to initiate fiberoptic evaluation and intubation? Is ENT consultation available and in what time frame.

The usual treatment for anaphylactic angio edema (epinephrine, antihistamine medication, corticosteroids) are not thought to help in cases of non-allergic angioedema (4) but are recommended as second line therapy (3-4).

In the case presented there is no present airway obstruction.

How would you proceed?

The options:

1. Continued observation with medical treatment of stopping ACE-I, administer IV steroids and IM/SC adrenaline, and antihistamine medication.
2. Nasal Intubation in the ED
3. Bronchoscopic nasal intubation in the ED
4. Bronchoscopic nasal intubation in the OR with ENT standing by for possible cricothyrotomy

<u>Exploring the Options</u>

1. Continued observation with medical treatment of stopping ACE-I, administer IV steroids and IM/SC adrenaline, and antihistamine medication. Advantages: The patient is currently stable, this is the least invasive course without complications associated with any more aggressive course of action. Disadvantage: If the angioedema progresses securing the airway may be more difficult and need to be done more urgently with potentially more complications than if done now. Requires close follow up (ICU) and ability for immediate response to any deterioration.

2. Nasal Intubation in the ED. On the face of it oral intubation was not a viable option due to tongue size. Advantages: Can be done with patient in sitting position with minimal sedation. If successful secures the airway in the ED without having to move the patient. Disadvantages: Blind procedure, and although there is no clinical evidence of angioedema of the posterior pharynx it is still possible. Also, epistaxis due to the trauma of the procedure could cause aspiration or even asphyxiation.

3. Bronchoscopic nasal intubation in the ED. Advantages: More controlled nasal intubation under vision. Can be done with patient in sitting position with minimal sedation. If successful secures the airway in the ED without having to move the patient. Disadvantages: Epistaxis due to the trauma of the procedure could cause aspiration or even asphyxiation. ED cricothyrotomy back up. All in all a relatively

major procedure procedure for busy ED resources.

4. Bronchoscopic nasal intubation in the OR with ENT standing by for possible cricothyrotomy. Advantages: More controlled nasal intubation under vision. Controlled environment of OR with maximal preparation for surgical airway before attempting intubation. Disadvantages: Patient needs to be moved to OR. Possibility of airway trauma and secretions.

In this case given the rapid onset (over 2 hours), the obesity of the patient and the size of the swelling, the treating physicians decided that control of the airway was indicated despite the lack of airway compromise at present. The latter however allowed for maximal preparation, and consultation with ENT and anesthesia,. The patient was taken to the operating room and underwent bronchoscopy which revealed no laryngeal edema. Nasotracheal intubation over the bronchoscope without complication.

After follow up examination by ENT, and with clinical improvement of the tongue swelling, the patient was extubated after twenty four hours and discharged with instructions not to use ACE-I or angiotensin receptor antagonists ever.

ISRJEM Invites your comments on management of this case, or questions for the author.

Email: Forum@isrjem.org

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Competing interests: None Declared
Funding: None

This manuscript has been peer reviewed.

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