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Visual Case Discussion

Angiotensin converting enzyme inhibitor induced Quincke's disease: A case report

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ABSTRACT

Purpose: To describe and alert practitioners to potentially rare but life-threatening side effects due to angiotensin converting enzyme inhibitors Summary: Angioedema is caused by capillary leakage and subsequent swelling of surrounding tissues. This can be life threating emergency if the tongue, larynx or oropharynx is affected as it can lead to airway obstruction. Isolated uvular angioedema, also known as Quincke...s Disease, was first described in 1882 and has since been a relatively rare form of angioedema of the upper airway. The first known case of Quincke...s disease due to ACE-I was first reported in 1988 and consequent reports have been rare. Conclusion: We report a case of Quincke...s Disease in a patient taking lisinopril that was successfully treated.

1. Introduction

Angioedema is caused by capillary leakage and subsequent swelling of surrounding tissues. This can be life threating emergency if the tongue, larynx or oropharynx is affected as it can lead to airway obstruction. Isolated uvular angioedema, also known as Quincke's Disease, was first described in 1882 and has since been a relatively rare form of angioedema of the upper airway.[1] While angioedema can be due to hereditary causes, it can also be caused by thermal injury, trauma, infections. The first known case of Quincke's disease due to an angiotensin converting enzyme inhibitor (ACE-I) converting enzyme inhibitor was first reported in 1988 and subsequent reports have been rare.[2] We report a case of Quincke's Disease in a patient taking lisinopril.

2. Visual case discussion

A 59-year-old man with a significant medical history of hypertension, chronic lower back pain, and intermittent polysubstance abuse presents to the ED with a 2-day history of "sore throat." The patient reported that he had associated pain with swallowing and mild dry cough when he feels a "tickle" in the back of his throat. The patient reports being able to swallow both solids and liquids. He denied any change in his voice, difficulty maintaining oral secretions, shortness of breath, drooling, fevers, chest pain, new onset rash, any recent trauma or thermal injuries from smoking or food. He reported that he engaged in oral sexual intercourse with a receptive female partner. The patient reported compliance with daily lisinopril for his hypertension and does not take medication for his low back pain.

Vital signs included a temperature of 97.7 F (35.6C), pulse of 91 beats per minute, 18 respirations per minute, and a blood pressure of 155/108, and an oxygen saturation of 99% on ambient air. Complete blood count, basic metabolic panel and all other blood work was within normal limits, including complement (C4 and C1) levels. Acute infectious workup was all negative as well, including Step A, Gonorrhea, Chlamydia, Influenza, and COVID-19. Physical exam was unremarkable except for the flexible endoscopy. Findings showed no nasopharyngeal, oropharyngeal, supraglottic, glottic or hypopharyngeal lesions. Epiglottis was crip, with clear posterior cricoid and bilateral pyriformis. True vocal folds were mobile bilaterally. The only pertinent finding was an isolated erythematous uvula (Fig 1). The patient was given 10 mg of dexamethasone intravenously and discharged with oral prednisone to complete a 5-day course and started on a new antihypertensive

3. Discussion

Quincke's disease, or isolated uvular angioedema remains a rarity

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Fig. 1. A patient with isolated uvular edema after taking lisinopril.

and can be caused by infections, burns, trauma or medications. This condition can lead to rapid airway compromise and can potentially be life-threatening. While it remains rare, failure to recognize that this disease can be caused by ACE-I may lead to continuation of the medication and significant morbidity for patients. The mainstay of treatment for Quincke's disease, like angioedema or anaphylaxis, is histamine receptor blockers, corticosteroids, and/or intramuscular epinephrine in the setting of airway compromise.[3]

4. Conclusion

Anytime a patient presents with isolated uvular angioedema, besides the common causes, ACE-I should be kept as one of the differential causes. Mainstay of treatment is aimed at maintaining a viable airway, in conjunction with corticosteroids, antihistamines and epinephrine if the airway is compromised.

5. Questions

Question Text: Angiotensin converting enzyme inhibitor (ACE-I) induced Quincke's Disease is caused by anaphylaxis and the subsequent release of histamine causing isolated edema of the uvula?

Answer Options

a) True
b) False

Correct Answer = b

False. Quincke's Disease due to ACE-I is due to the build-up of bradykinin. The angiotensin converting enzyme is responsible for breaking down bradykinin. When the bradykinin molecule builds up and then attaches to the bradykinin receptor, it leads to local vasodilation and capillary leakage into surround tissue. The isolated local swelling of the uvula is termed Quincke's Disease, named for, Heinrich Quincke, the physician who first described it in medical literature.

Question Type: multiple choice

Question Text Which patient race has been associated with a higher prevalence of ACE-I induced angioedema?

Answer Options

a) African-American
b) Asian/Pacific Islander
c) White, non-Hispanic
d) Hispanic
e) Middle-East/North African

Correct Answer = a

In a 2008 retrospective cohort study Mahoney et al. found that out of 182 patients, 81% who developed ACE-I angioedema were black, even though black patients made up 72% of the entire cohort; the black patients were three times more likely to develop ACE-I angioedema. In a systematic review of pharmacogenomics, the gene region XPNPEP2 was associated with a higher incidence of angioedema. Woodward-Grice, et al. performed a multivariate analysis to look into gender and race-dependent prevalence of a gene polymorphism with ACE-I induced angioedema and found that the XPNPEP2 C-2399A gene polymorphism was more prevalent in black men and not white men or women.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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