Swollen Tongue After Dental Procedure – Diagnosis and Management Of Type III HAE

Z. Khan, MD, M. Roberge, MD
UPMC Mercy

Introduction

- Hereditary Angioedema (HAE) characterizes as recurrent, self-limiting episodes of angioedema of varying severity involving the gastrointestinal tract and upper airway.

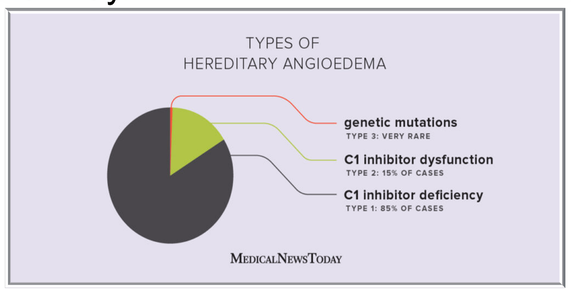

Fig 1. Incidence of HAE

Case Presentation

- A 50-year-old female with unknown family history (adopted) and past medical history of upper extremity deep vein thrombosis and recurrent tongue swelling presented for a dental procedure requiring anesthesia.
- The patient developed a diffusely swollen and protruding tongue post-procedure affecting her speech but with normal swallowing and no respiratory distress.
- She developed these episodes in the fourth decade of her life, which seemed to have worsened with oral contraceptive use, but never required intubation before this visit.

Diagnosis

- Vasculitis panel, complement levels, and C1 esterase inhibitor level were only notable for a weakly positive Antinuclear Antibody (1:40 titer); repeat laboratory tests were unremarkable except for mildly low immunoglobulin G level with no involvement of the vocal cords, epiglottis, or oropharynx on visualization with flexible laryngoscopy.

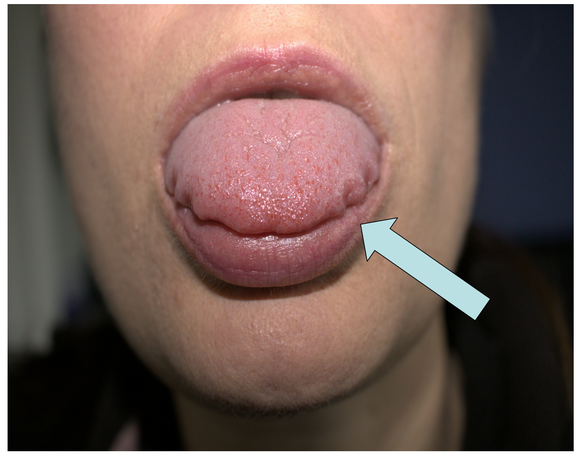

Fig 2: Swollen tongue due to Angioedema

Laboratory test	Type I HAE	Type 2 HAE	Type 3 and idiopathic HAE
C4 concentration	Low	Low	Normal
C1-INH concentration	Low	Normal/high	Normal
C1-INH function	Low	Low	Normal

Abbreviations: C1-INH, C1-inhibitor; HAE, hereditary angioedema.

Table 1: Different types of HAE

Management

- Formerly, varying combinations of steroids, antihistamines, amphetamines, and diazepam, were used with variable success for these self-limiting episodes.
- She was intubated for airway protection and received methylprednisolone, famotidine, and diphenhydramine. The patient was extubated after her initial treatment but continued having acute flare-ups that required varying combinations of PRN Diphenhydramine, Diazepam, Furosemide, and Corticosteroids.
- The patient had a brief respite in her symptoms with a trial of plasma infusions every 12 hours for three days, which led to four days without symptoms but failed treatment with Danazol and Icatibant (bradykinin B2 receptor antagonist)
- Given her history of deep vein thrombosis, tranexamic acid was contraindicated. For maintenance and prevention, the patient was trialed on a C1 Esterase Inhibitor (Haegarda), which reduced her acute episodes from three to four times a day to once daily.

Discussion

- Hereditary Angioedema can be classified into three subtypes based on the pathophysiological cause of the swelling.
- The quick onset and underlying risk of airway compromise warrants urgent intervention. The management is aimed at the prevention of flare-ups with Tranexamic acid, Danazol, and Haegarda, along with early abortive treatment of acute episodes with Icatibant and Plasma infusions.
- There are limited prophylactic and abortive treatment options for Hereditary Angioedema requiring a personalized, per-patient approach as a response to the treatment can be highly variable.

References

- https://www.researchgate.net/figure/Diagnosis-of-types-1-2-3-and-idiopathic-HAe_tbl1_275664236
- https://www.medicalnewstoday.com/articles/facts-stats-hereditary-angioedema#types
- https://stock.adobe.com