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.

Fig 2. Swollen tongue due to Angioedema

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.

Table 1: Different types of HAE

<table>
<thead>
<tr>
<th>Laboratory test</th>
<th>Type 1 HAE</th>
<th>Type 2 HAE</th>
<th>Type 3 HAE</th>
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</thead>
<tbody>
<tr>
<td>C4 concentration</td>
<td>Low</td>
<td>Low</td>
<td>Normal</td>
</tr>
<tr>
<td>C1-INH concentration</td>
<td>Low</td>
<td>Normal/High</td>
<td>Normal</td>
</tr>
<tr>
<td>C1-INH function</td>
<td>Low</td>
<td>Low</td>
<td>Normal</td>
</tr>
</tbody>
</table>

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

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_fig1_275664236
- https://www.medicalnewstoday.com/articles/facts-stats-hereditary-angioedema#types