Session 3557
Interesting Cases II – Sunday, March 15, 2020

Case Report #3

Author John Trickett, MD
Training Program Mayo Clinic - Arizona

Case Title
A Sensation of Tongue Swelling When the Tongue Is Not Swollen: Factitious Angioedema

Summary
A 48-year-old female presented to her local ED for a sensation of swollen lips and tongue multiple times one year prior to presentation at our Allergy-Immunology clinic. She was treated during these episodes with epinephrine, corticosteroids, and antihistamines. After having no episodes for nine months, she began sensing swollen lips and tongue, including episodes where she appeared to have dysarthria and respiratory distress. For most episodes, epinephrine was given. During one episode, she was intubated and mechanically ventilated. Laboratory testing and imaging were unremarkable. Ultimately, the patient was referred to our ED, and by working with ED and ENT teams, was documented on three separate occasions as having dysarthria that was variable and associated with tongue thrusting (no angioedema). A diagnosis of factitious angioedema was made and the patient was evaluated by psychiatry. Following this, her chart was flagged and the episodes stopped occurring with no further ED visits reported.

Patient Presentation
A 48-year-old female with a past medical history of multinodular thyroid disease status-post isthmusectomy and allergic rhinitis presented to the Emergency Department (ED) following an episode of reported lip and tongue swelling over one year prior to presentation to the Allergy-Immunology outpatient clinic at our institution. Following a nine-month period with no episodes, she made multiple visits to the ED with concerns related to the inability to talk due to tongue and lip swelling. She denied improvement with icatibant or high-dose steroids and by report, her speech became more muffled so she was then intubated and treated with epinephrine and higher doses of steroids. She was eventually extubated 2 days later, and in discussions with the critical care specialists involved in her care, there was not obvious evidence of angioedema and questions existed about the indications for the initial intubation. C1 esterase, C4, C3, ESR, CRP, and tryptase level during this episode of reported angioedema were all within normal limits.
Over the next two months, she continued to experience a sensation of tongue and lip swelling leading to repeated hospitalizations and intubations. Multiple subspecialists in her city were consulted including Allergy-Immunology, ENT, and dermatology. Further workup including thyroid, leukotriene, prostaglandin studies, autoimmune panels, salivary gland biopsy, and additional imaging studies were all negative. She reported improvement of her symptoms only after receiving fresh frozen plasma (FFP) in the ED.

Diagnosis

Factitious angioedema was diagnosed after multiple ENT exams in the ED revealed no objective findings of angioedema.

Testing

She was then seen in our Allergy-Immunology outpatient clinic where all initial laboratory and imaging studies were conducted, reviewed, and found to all be unremarkable. At that time, the patient was reporting that the episodes were occurring every 48 hours. Given the importance of documenting the degree of angioedema, the safety of the airway, and the possibility of tongue thrusting, the Allergy-Immunology team suggested she be seen at our institution's ED during an episode. The Allergy-Immunology team communicated with the ED and ENT teams, including the concern that the angioedema could represent tongue thrusting. At the first ED visit, despite instructions from the Allergy-Immunology team to consider ENT evaluation prior to giving treatment, she received FFP and was examined shortly afterward by the ENT team who found no evidence of angioedema (Photo 1). At the second ED visit (48 hours later), no treatment was given and no angioedema was found on exam. At the third ED visit (another 48 hours later), evidence of tongue thrusting and its association with dysarthria was very clear, and the ENT, ED, and Allergy-Immunology team made a diagnosis of factitious angioedema. This diagnosis was further supported by the fact that all laboratory testing remained negative during these evaluations.

Treatment

A follow-up visit to the Allergy-Immunology outpatient clinic was made to review the work-up once again with the patient, explain the diagnosis, and present an opportunity to ask questions. The patient was not able to offer a specific external award for tongue thrusting, and the patient and Allergy-Immunology team agreed to proceed with a psychiatry consultation. She was seen by psychiatry who discovered that her son died in an accident when he was six years old and that she may have PTSD from this experience. She also had a previous near-suicidal attempt by overdose in her teenage years from depression. Psychiatry recommended personality testing for alexithymia, CBT, and an SSRI for comorbid trauma and anxiety. The final component of the treatment plan involved a conference between the Allergy-Immunology team and her local providers including her primary care provider and local ED leadership. Following a review of the evidence, we agreed to flag the patient's electronic record so that providers could consider this diagnosis if she presented for care in the future.

Patient Outcomes

The episodes stopped occurring and no further ED visits were reported.

Lessons Learned

Factitious disorder is defined in the DSM-V as "the falsification of medical or psychological signs and symptoms in oneself… in the absence of obvious external rewards." It is noted that up to 1% of patients who present to the hospital have factitious complaints. Angioedema is a rare presentation of factitious disorder on self with literature consisting of mainly case reports. Feldman et al. reported a series consisting of four cases of "angioedema," which were later deemed to be manifestations of factitious disorder. In our patient, no external reward could be identified. Not all patients who present with a clinical concern for angioedema have angioedema that can be objectively confirmed. In particular, tongue, throat, and larynx angioedema can be difficult to diagnose, especially without expert ENT

examination during an episode. Factitious angioedema should be considered if angioedema cannot be confirmed despite multiple examinations and repeatedly negative laboratory data. A detailed laboratory evaluation for angioedema may not be indicated when the diagnosis of angioedema has not been objectively confirmed.