Title: Face Value: A Case of Small Lymphocytic Lymphoma And Its Connection To Acquired Angioedema

Authors
Monica Moreno (LSU Internal Medicine), Anna Chataginer (LSU Internal Medicine), Spencer, Lemoine (LSU Internal Medicine), Merrick Firroz (LSU Internal Medicine), Seema Walvekar (LSU Internal Medicine)

Case Presentation
A 67-year-old female with a history of stage III Small Lymphocytic Lymphoma and hypertension presented to the emergency department with one week of progressive neck, facial, and lip swelling accompanied by erythema and yellow crusting of the lips. On presentation, she was hemodynamically stable; however, given concern for potential airway compromise, she was admitted to the intensive care unit for close monitoring and possible intubation. Computed tomography of the neck demonstrated interval enlargement of submandibular and cervical lymphadenopathy. She was empirically started on broad-spectrum antibiotics and dexamethasone for suspected infectious and inflammatory etiologies. Notably, she had experienced a similar episode three months prior, during which lymph node biopsy revealed complete effacement of nodal architecture by a diffuse infiltrate of atypical, minimally pleomorphic, small B-cells consistent with small lymphocytic lymphoma. During the current admission, she had not yet initiated chemotherapy and was evaluated by hematology/oncology and otolaryngology. Given concern for possible transformation to a more aggressive process, repeat imaging, bone marrow biopsy, and lymph node biopsy were performed. Her symptoms improved, and she was transitioned to oral antibiotics. The differential diagnosis for her facial swelling included progressive lymphadenopathy causing impaired lymphatic or venous drainage versus Angioedema; however, normal complement and C1 esterase inhibitor levels made angioedema less likely. She was discharged with hematology and oncology follow up for ongoing management of her lymphoma.

Discussion
Acquired angioedema is a rare, non-allergic severe cervicofacial swelling linked to an acquired C1-inhibitor deficiency and is associated with autoimmune disease or lymphoproliferative disorders, most specifically SLL and CLL. One determined mechanism is that the underlying B-cell disorder creates neutralizing antibodies against C1 esterase. Although rare, there are case reports of angioedema as the initial presentation for lymphoma. Though this patient's C1 esterase levels were normal, her lymphoma was diagnosed during her first hospitalization for facial and neck swelling, and these levels were not obtained until her second hospitalization. Though her SLL is not associated with acquired angioedema, patients that present with angioedema and concern for lymphoma on imaging, should be evaluated for this as it will not improve with steroids alone. It can be managed with plasma-derived C1-INH concentrates. A large number of our patient population take ACE-inhibitors and we learn this as a classical adverse effect, knowing to stop the medication. However, understanding that angioedema could be a presentation for lymphoma is important, and imaging beyond the head and neck can reveal a more insidious, life threatening process beyond what we see on the outside.