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Hereditary Angioedema Laboratory values in relation to C1- esterase inhibitor treatment

	Admission Day 1	Admission Day 2	Admission Day 2	Admission Day 3	Admission Day 4	Outpatient Visit
Date & Time	11/13/2020 3:34 PM	11/14/2020 12:15 PM	11/14/2020 5:14 PM	11/15/2020 5:01 AM	11/16/2020 6:58 AM	7/16/21 2:35 PM
C4 Complement (mg/dL)	2.0 (L)	18.4	3.6 (L)	7.1 (L)	14.9	6 (L)
C1 Esterase Inhibitor (mg/dL)	28	C4 level obtained prior to C1 esterase inhibitor product	5:36 PM C1 esterase inhibitor	11:41 AM C1 esterase inhibitor		44 (H)
C1 Esterase Inhibitor, Functional	32 (L)		1200 units, plasma derived	1200 units, plasma derived		8 (L)
C1Q Binding	5.8 (H)					
C1Q Complement (mg/dL)	13					17.1

M123

TRAUMATIC "ANGIOEDEMA": EXTENSIVE SUBCUTANEOUS EMPHYSEMA MASQUERADING AS ANGIOEDEMA

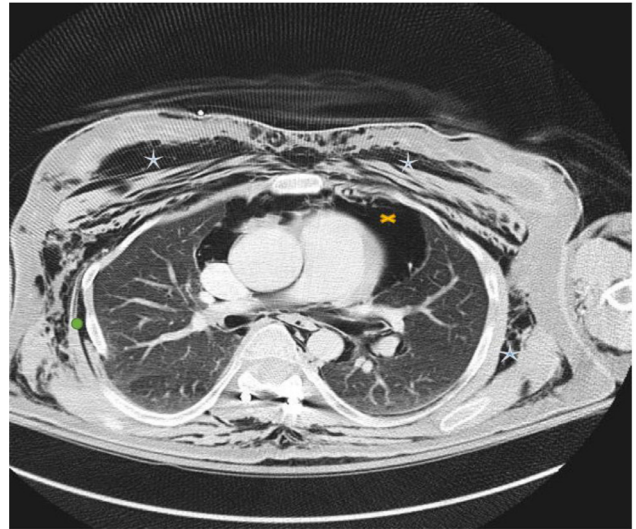
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Introduction: COVID 19 pandemic related precautions have resulted in suboptimal physical examination of the patients which may have affected the patient care and training of medical professionals. We present a case of a patient whose subcutaneous emphysema was misdiagnosed as angioedema.

Case Description: 25 years old female with cerebral palsy and developmental delay presented to hospital in June, 2020 with asymmetric left facial and periorbital swelling which developed an hour after her regular Depo-Provera injection. She had elevated blood pressure readings with tachycardia without hives, respiratory or gastrointestinal symptoms. Facial swelling did not improve after IM epinephrine, IV steroids and antihistamines. Flexible laryngoscopy noted mild edema of lingual surface of the epiglottis. Allergy service was consulted to evaluate for drug-induced anaphylaxis. Our physical examination revealed significant asymmetric periorbital edema, neck and chest swelling with distant breath sounds and distinct palpable crepitus. Serum Trypsin and C1-Esterase inhibitor levels were normal at 3.2ug/L and 28 mg/dl, respectively. Emergent imaging was recommended, which demonstrated multiple rib fractures, pneumomediastinum with extensive chest wall subcutaneous emphysema tracking to extra-cranial soft tissue. Patient was intubated and chest tube was placed due to concerns for airway compromise. She stayed in ICU for one week before being discharged to home.

Discussion: Facial edema, concerning for angioedema, is potentially life threatening condition and an allergic reaction should be higher on the differential diagnoses. Our case represents a learning opportunity regarding other potentially life threatening conditions that can mimic angioedema, requiring higher index of clinical suspicion and thorough physical examination.

CT Chest revealing subcutaneous emphysema.



CT scan of the chest reveals pneumomediastinum with extensive subcutaneous emphysema, in the setting of right Rib fractures (●)

➡ : Pneumomediastinum

★ : Areas of extensive subcutaneous emphysema

M124

COMPLETE RESOLUTION OF ACQUIRED ANGIOEDEMA FROM SPLENIC MARGINAL ZONE LYMPHOMA AFTER TREATMENT WITH RITUXIMAB

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Introduction: Acquired angioedema (AAE) due to C1 esterase inhibitor (C1-INH) deficiency is clinically similar to hereditary angioedema (HAE), but typically develops in older patients. There is invariably an underlying disease such as a lymphoproliferative disorder, autoimmune condition, solid tumor, etc. that either drives autoantibody production against normal C1-INH and/or causes consumptive depletion of normally functioning C1-INH.

Case Description: A 68-year-old Caucasian male with past medical history of hypertension, presented 1 year previously with an episode of lip angioedema followed by recurrent episodes of abdominal pain lasting > 12 hours, associated early satiety, and gradual weight loss. He also developed peripheral neuropathy, splenomegaly of unclear etiology, and hypogammaglobulinemia. Workup of his angioedema led to the discovery of low C4 and C1-INH levels, and prophylactic treatment with Icatibant was initiated to treat further attacks. Given his age, splenomegaly, weight loss, and other clinically relevant information, he was evaluated for malignancy, especially lymphoproliferative disorders. He was subsequently diagnosed with Splenic Marginal Zone Lymphoma (SMZL), a clonal B cell neoplasia, and received one cycle of Rituximab with profound improvements of his angioedema-related symptoms, splenomegaly, and labs, including normalization of his quantitative C1-INH and C4 levels and hypogammaglobulinemia.