Three Recent Cases of ACE-I Related Angioedema Causing Severe Airway Obstruction

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Case Description:

Case 1: 56-year-old male with repaired TOF, CAD, and HTN presented with worsening swelling of the lips and tongue. He denied dyspnea or hoarseness. His primary care physician evaluated his lower extremity edema 1wk prior to admission and increased his lisinopril from 10mg BID to QID. In the ER, the patient was treated with PO Solu-Medrol 125mg, Benadryl 50mg, epinephrine SQ, and racemic epinephrine without improvement and was admitted to the MICU where he had dyspnea, hoarseness, and diaphoresis. He was transferred to the OR, nebulized and topicalized with lidocaine, but was still dyspneic and restless while O2sat dropped from 100% to 30%. Nasal cannula was converted to 100% O2 face mask (10L/min). Bag-mask ventilation was difficult, fiberoptic intubation was unsuccessful, LMA #3 improved ventilation (O2sat 80%), however BP dropped from SBP of 120s to the 60s. Tracheostomy was successful but pulseless VTach was noted. IV epinephrine 1mgx5, Ca2+ 1gx2, Mg2+infusion, vasopressin 40units x2, and defibrillation x2 brought the patient to NSR. In the MICU, a GCS score of 3T with no improvement of neurological status made his family decide to withdraw care on day 3. Time from the ER to the OR was 5 hours and 6 minutes. C1 esterase inhibitor levels were WNL; however, C4 level was not measured.

Case 2: 62-year-old male with H/O HTN and lisinopril-induced angioedema 2yrs prior to admission presented with swelling of the tongue and lips with no resolution after PO Benadryl 50mg. Symptoms progressed to dysphasia and dyspnea. One day prior to admission, he was seen at an urgent care clinic and was prescribed enalapril for HTN. In the ER he was treated with IV Benadryl 50mg, Pepcid 20mg, and Solu-Medrol 125mg, but dyspnea worsened. An anesthesiologist was called and noted the patient’s muffled voice and inability to speak, and transferred him to the OR. Awake fiberoptic intubation with topical lidocaine was successful on the 2nd attempt. The patient remained hemodynamically stable with an O2sat >95%, was transferred to the PACU and then to the ICU, and was successfully extubated on day 3 with no complications. Time from the ER to the OR was 2 hours and 45 minutes. C1 esterase inhibitor deficiency labs were not determined.

Case 3: 59-year-old female H/O HTN treated with lisinopril presented with worsening swelling of the tongue and lips 3 hours after her usual 10mg lisinopril. The patient was transferred to the OR for awake fiberoptic intubation which was successful on the first attempt. The patient remained hemodynamically stable with O2sat >95%, was transferred to the PACU and then to the ICU, and was successfully extubated on day 2 with no complications. C1 esterase inhibitor concentrate was administered after her airway was secured. Time from the ER to the OR was 2 hours and 30 minutes. Results of C1 esterase inhibitor deficiency labs are pending.
Discussion:

Angioedema secondary to ACE-I usually occurs after beginning therapy or increasing dose, but may occur after years of use. ACE degrades bradykinin therefore ACE-I causes accumulation of bradykinin which leads to increased vascular permeability causing angioedema. Symptoms resolve 24-48hrs after discontinuing ACE-I. Airway management is not always necessary and should be decided by a case-by-case basis. Significant improvement in outcome was found with expediting time from the ER to the OR when intubation is necessary. ACE-I use can unmask Hereditary Angioedema (HAE) in undiagnosed individuals. For this reason, patients with suspected ACE-I related angioedema should undergo testing for C1 esterase inhibitor deficiency. Both ACE-I related angioedema and HAE have elevated bradykinin levels leading to angioedema. Treatment with C1 esterase inhibitor concentrate (suppress the local over-production of bradykinin) or FFP (degradation of excess bradykinin by Kininase II) have both been shown to be effective in ACE-I related angioedema and HAE.

Summary:

We reported 3 cases of angioedema secondary to ACE-I use which caused severe airway obstruction requiring endotracheal intubation. Significant improvement in outcome was found with expediting time from the ER to the OR for airway management when intubation was necessary. Treatment with C1 esterase inhibitor concentrate (suppress the local over-production of bradykinin) or FFP (degradation of excess bradykinin by Kininase II) have both been shown to be effective in ACE-I related angioedema and hereditary angioedema.