**BACKGROUND**

C1 inhibitory deficiency is associated with the syndrome of hereditary angioedema (HAE). HAE presents with submucosal swelling of facial, abdominal, laryngeal, or oropharyngeal structures that can lead to significant airway collapse and respiratory distress. Edema formation is related to reduction or dysfunction of C1 inhibitor (C1-INH), which results in the release of bradykinin and C2-kinin mediators, which continue to enhance vascular permeability and lead to fluid extravasation (1). Treatment with corticosteroids and antihistamines are ineffective. Diagnosis is made by a constellation of clinical symptoms, decreased C4 levels, and decreased C1 esterase (2).

**PERIOPERATIVE MANAGEMENT**

Preoperative: Mallampati 1 airway, IV placed preop, 10ml/kg NS bolus in ED

Intraoperative: Minimal facial manipulation during preoxygenation via face mask. Modified rapid sequence intubation to secure airway. Patient was intubated easily without incident. C1 inhibitor concentrate was available in the OR and kept with patient postoperatively to treat an acute attack. An adequate cuff leak was confirmed and the patient was extubated awake.

Postoperative: Post op ICU observation was recommended to surgery staff to watch for signs/symptoms of respiratory distress. The patient was discharged on post op day 2 without an acute attack. Follow up of lab work shows low C1-INH levels consistent with her family history of hereditary angioedema.

**DISCUSSION**

Due to good tolerance of airway manipulation, adequate tidal volumes and respiratory effort, and minimal secretions we opted for conservative management and did not administer C1-INH concentrate (C1-INH). Unless head, neck intervention C1-INH is usually not routinely necessary for other surgical procedures (2). Emergent treatment of a HAE attack is a dose of C1-INH concentrate 10-20 units/kg. Onset is ten minutes after infusion starts. Intravenous volume replacement is needed to correct fluid extravasation. C1-INH concentrate is expensive and unavailable in many facilities and countries (4). If C1-INH is unavailable 1unit-2units of FFP can be transfused to the patient to increase C1-INH levels (2). In patients with history of severe reactions, C1-INH and androgens can be given prophylactically before scheduled surgery (3).

**REFERENCES**