[NM-209] Severe Hereditary Angioedema and General Anesthesia for portacath placement.

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Introduction: Hereditary angioedema (HAE) is a rare disease caused by a deficiency of C1 esterase inhibitor. It is characterized by recurrent episodes of swelling, without accompanying hives or purities, that often affects the skin or mucosa of the upper respiratory and gastrointestinal tracts, face and extremities.(1,2,3,4) This swelling is often self-limiting, however the laryngeal edema associated with attacks may be life threatening.(2,3) Various triggers have been report including mild trauma, stress, medications or menstruation.(1,2,3)

Case description: A 17-year-old 65 kg female with a history of hereditary angioedema, presented for surgery for a port placement secondary to her frequent episodes of angioedema. She reported having repeated episodes of airway swelling and abdominal swelling. She required the medication Cinrynze, a plasma derived C1 esterase inhibitor product, every three days. This medication is administered intravenously and given the patient had difficult IV access the decision was made to place a port. Her other medications included Firazyr, Ecallantide and an EpiPen on an as needed basis. She had to administer Ecallantide, a medication used for the treatment of HAE attacks given subcutaneously, as frequently as once or twice a day. Prior to surgery she was premedicated with oral versed and lidocaine spray was used for IV start. She was then given Cinryze prophylactically one hour prior to surgery. With difficult airway equipment, an emergency tracheostomy kit and ENT surgeons on standby anesthesia was induced with midazolam, ketamine, fentanyl and vecuronium. Propofol was avoided due to pain on injection. Laryngoscopy revealed no edema and the airway was secured with a 6.0 endotracheal tube following the administration of 3cc’s of lidocaine via a laryngotracheal topical anesthesia kit (LTA). Anesthesia was maintained with sevoflurane. She was reversed with neostigmine and glycopyrrolate and ondansetron was given for PONV prophylaxis. She remained hemodynamically stable throughout the case and extubated deep. She did not develop any evidence of angioedema and was discharged home after an uneventful PACU course.

Discussion: Severe angioedema can occur in the perioperative period, causing fatal airway obstruction. It is important to understand and take the necessary precautions when managing patients with HAE. This case demonstrates that successful management of this rare condition can be obtained with administering prophylactic C1 esterase inhibitor and meticulous measures to avoid triggers.

References:
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