Unmasking a Hemifacial Swelling : Hereditary Angioedema

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Angioedemas are common causes for emergency department admissions, requiring knowledge of their etiology and management by healthcare providers in these settings. Hereditary angioedema (HAE) is a rare yet potentially life-threatening condition, often underdiagnosed, marked by recurrent bouts of swelling affecting different areas of the body, including the skin, gastrointestinal tract, and airways. These episodes can cause intense discomfort, functional limitations, and in severe cases, respiratory compromise.

The case on discussion was a pregnant 32-year-old female with no known medical illness and no known drug or food allergy presents with right-sided unilateral facial swelling since morning. She only complained of a tightening sensation over the right side of her face. A trial of T.Cetrizine, IV Hydrocortisone and IV Adrenaline did not help the patient. The patient did not develop any airway, breathing, or circulatory compromise throughout her 3 days of admission to the hospital. The patient was on regular IV hydrocortisone and Tab.Piriton during the admission. The facial swelling eventually subsided and the patient was discharged well.

HAE results from deficiency or dysfunction of C1-inhibitor(C1-INH) leads to unchecked activation of the complement system, which leads to overproduction of bradykinin. HAE attacks are transient, typically lasting two to five days, with manifestations ranging from cutaneous swelling to life-threatening upper airway edema. Common triggers include stress, hormonal changes, infections, and certain medications e.g ACE inhibitors. Response to treatment with antihistamines, corticosteroids, and epinephrine may distinguish histamine- and bradykinin-mediated angioedema.

The management of HAE patient should prioritize the standard airway, breathing, and circulation interventions as C1-INH concentrates or recombinants are generally not available in Malaysian emergency departments. Alternatively, fresh frozen plasma has been used to treat HAE in cases where first line therapies are not available. Although its use is controversial, Tranexamic Acid is also used acutely and prophylactically to treat HAE.

For optimal treatment and care, it is crucial to ascertain whether angioedema is histamine- or bradykinin-mediated. Given the absence of a dependable point-of-care test for distinguishing between these two pathophysiologies, emergency department physicians should acquaint themselves with existing indicators to guide treatment strategies effectively.

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