Case Report

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Recurrent ICU Admissions for Airway Protection in a Young Female with Hereditary Angioedema: A Case Report

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ABSTRACT

Hereditary angioedema (HAE) is a rare condition that may involve life-threatening upper airway swelling. A 19-year-old female with known C1 esterase inhibitor deficiency presented multiple times with lip and tongue swelling without stridor. She was electively intubated for airway protection during each episode and treated with Berinert®, resulting in symptom resolution. Hormonal and prophylactic therapy was initiated after repeated ICU admissions. This case highlights the need for high suspicion and early airway intervention in HAE, even in the absence of overt respiratory distress. Prompt airway management and tailored long-term treatment are essential in managing recurrent angioedema.

Introduction

Hereditary angioedema (HAE) is a rare autosomal dominant disorder caused by a deficiency or dysfunction of C1 esterase inhibitor [1, 2]. It presents with episodic, non-pruritic swelling that can involve subcutaneous or submucosal tissues, including the upper airway. Airway involvement, while less common, poses the greatest risk of mortality [3]. This report presents a case of recurrent ICU admissions for airway protection in a young female with HAE.

Case Presentation

A 19-year-old female with a known history of C1 esterase inhibitor deficiency presented to the Emergency Department with acute lip and tongue swelling. Despite the absence of dyspnea or stridor, elective intubation was performed due to the high risk of airway compromise. She was admitted to the ICU and received intravenous Berinert® (C1-INH concentrate). Her symptoms gradually improved, and she was extubated successfully. Over the next 6–8 months, she experienced two more similar episodes requiring ICU care and early airway intervention. She

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was under immunology follow-up and was initiated on prophylactic therapy and hormonal modulation due to recurrent presentations. She was a poor historian, and no specific triggers could be identified.

Discussion

Hereditary angioedema (HAE) is a potentially life-threatening condition characterized by episodes of non-pruritic, non-urticarial swelling, most often affecting the skin, gastrointestinal tract, and upper airways. It is caused by either a quantitative or functional deficiency of C1 esterase inhibitor (C1-INH), leading to unchecked bradykinin production and increased vascular permeability [2,4]. Unlike histamine-mediated angioedema, HAE does not respond to corticosteroids, antihistamines, or epinephrine, necessitating condition-specific treatment [2].

Laryngeal edema is the most feared complication of HAE due to its rapid progression and high risk of asphyxiation. Bork et al. demonstrated that up to 30% of untreated laryngeal attacks may be fatal, emphasizing the need for early airway protection in suspected cases, even in the absence of overt respiratory symptoms [1]. In our patient, despite the absence of stridor or dyspnea, the decision to perform early elective intubation was guided by the recognition that orofacial swelling may precede airway obstruction. This highlights the importance of clinical vigilance and a low threshold for airway intervention, consistent with current guidelines [4, 5].

Management during acute attacks focuses on targeted therapies such as plasma-derived C1-INH concentrate (Berinert®), recombinant C1-INH, bradykinin receptor antagonists (e.g., icatibant), or kallikrein inhibitors [6, 3]. In this case, administration of intravenous Berinert® led to rapid symptom improvement, aligning with established efficacy data for on-demand treatment [6].

Recurrent ICU admissions in this patient prompted evaluation of long-term prophylactic strategies. The 2020 U.S. HAE Medical Advisory Board Guidelines recommend long-term prophylaxis in patients with frequent or severe attacks, especially those requiring repeated ICU or emergency interventions [3]. Attenuated androgens, antifibrinolytics, and monoclonal antibodies targeting kallikrein (e.g., lanadelumab) are among the prophylactic options [3, 4].

Additionally, hormonal modulation played a role in this patient's management plan. Estrogens are known to exacerbate HAE symptoms, particularly in women, through their effect on bradykinin production pathways. Hormonal regulation, including switching contraceptives and cycle suppression, is often necessary in female patients [4, 5].

Another important aspect is the recognition of prodromal symptoms, which may precede an HAE attack by several hours. Prematta et al. reported that up to 85% of patients experience early signs such as tingling, fatigue, or a non-painful rash, which can aid early diagnosis and treatment [7]. However, our patient was a poor historian, making it challenging to identify triggers or prodromes a common issue in real-world practice.

This case underscores the necessity of a multidisciplinary approach involving emergency, ICU, and immunology teams. Effective acute care, combined with personalized long-term management, is essential in preventing morbidity and reducing ICU burden in patients with hereditary angioedema.

Conclusions

This case underscores the need for early airway intervention in hereditary angioedema involving the orofacial region, even when symptoms appear clinically stable. A multidisciplinary approach involving ICU and immunology teams is vital for preventing recurrent ICU admissions.

Patient Consent

Verbal consent was obtained from the patient for publication of this case report.

Conflicts of Interest

The author declares no conflicts of interest.

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