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CASE REPORT

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Hereditary Angioedema: A Case Report and Clinical Review

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ABSTRACT

Hereditary angioedema (HAE) is a rare genetic disorder characterized by recurrent angioedema without urticaria or pruritus, involving the skin, upper airway, and gastrointestinal mucosa; swelling resolves within two to five days, yet laryngeal involvement can cause life-threatening airway obstruction. HAE results from C1 esterase inhibitor (C1-INH) deficiency (Type I) or dysfunction (Type II), disrupting the complement and contact systems and leading to excessive bradykinin production, which increases vascular permeability and tissue swelling. Prevalence is ~1 in 50,000, with autosomal dominant inheritance and onset in childhood or adolescence, though late-onset cases occur. We report a 6-year-old Filipino boy with right facial swelling after a suspected insect bite on the upper eyelid; family history prompted referral to a Pediatric Allergologist. Initial diphenhydramine, hydrocortisone, and ranitidine maintained stability with improvement over two days; outpatient assays showed decreased C1-INH function and level, confirming HAE. Early recognition, timely diagnosis, appropriate management, and education, especially in resource-limited settings, are crucial for outcomes.

Keywords: hereditary angioedema, C1 esterase inhibitor, C1-INH deficiency, C1-INH inhibitor, C1 inhibitor deficiency hereditary angioedema

INTRODUCTION

Hereditary angioedema (HAE) is a rare genetic disorder caused by either a dysfunctional C1 esterase inhibitor gene (C1-INH) or a deficiency of C1-INH. This condition follows an autosomal dominant inheritance pattern and affects approximately 1 in 50,000 individuals. C1-INH is involved in various pathways, including fibrinolysis, coagulation, and the contact and complement systems. There are three types of HAE: Type I, which accounts for 85% of cases, is characterized by a deficiency of C1-INH; Type II, representing 15% of cases, involves a dysfunctional C1-INH; and Type III, also known as estrogen-dependent HAE, is associated with normal C1-INH activity but has several identified mutations. All three types of HAE present similarly, with recurrent episodes of non-pitting edema in subcutaneous and submucosal tissues, affecting the lips, face, neck, extremities, oral cavity and larynx. When the larynx is involved, it can be potentially life-threatening. Symptoms typically begin in childhood or young adulthood and often worsen around puberty, usually manifesting as recurrent swelling or abdominal pain. Some patients may also develop a serpentine, non-pruritic rash. Most acute episodes of HAE resolve within 1–2 days. The pathophysiology of HAE is mediated by bradykinin. In Types I and II HAE, when C1-INH is deficient or defective, the contact system is activated, leading to continuous production of kallikrein, which results in uncontrolled proteolysis of high-molecular-weight kininogen and the generation of bradykinin. This increase in bradykinin leads to heightened vascular permeability, causing edema.[1,2]

CASE

This case describes a 6-year-old Filipino male living in Cupang, Muntinlupa, who presented with right facial swelling. One day before admission, the patient woke up with a questionable insect bite on his right upper eyelid and allegedly slept late the previous night. Swelling of the right eye was noted, which then progressed to swelling of the right side of the face. He was given cetirizine (5 mg/5 ml, 5 ml twice), but it provided no relief. There were no other associated signs or symptoms present. On the day of the consult, there was continuous progression of facial swelling, which prompted a visit to the emergency room, where he was admitted.

The patient has a significant past medical history, including urticaria diagnosed at the age of 2, for which he was treated with prednisone (1 mg/kg/day). He has experienced multiple episodes of angioedema: the first attack occurred in 2022 secondary to an insect bite, resulting in swelling of the left ear; the second attack was noted after riding an airplane, with swelling of the genitals and gluteal area; and the third attack occurred in August 2023 following physical trauma and, allegedly, an insect bite, presenting with facial swelling. Additionally, the patient was diagnosed with asthma at 2 years old, for which he uses salbutamol nebulization as needed. His asthma attacks usually occur once a month, with the last attack occurring at 4 years of age. He was previously admitted for amoebiasis in November 2023. The patient has a positive history of allergies to cow's milk and dog dander, as confirmed by an allergy panel done in 2018.

Family history showed a maternal history of angioedema, a paternal history of asthma, and a maternal history of diabetes mellitus (Figure 1). The patient is the second child in a family of eight members, residing in a two-story, well-ventilated concrete house with no pets. He was born full term via repeat Cesarean section to a 43-year-old G2P2 mother, with no noted fetomaternal complications. His mother had no infections during pregnancy and was not exposed to radiation, smoking, or alcohol. Newborn screening revealed a positive result for G6PD deficiency. Immunizations were completed at a private clinic. The patient was formula-fed since birth with partially hydrolyzed milk. Other feeding and developmental history were unremarkable. Review of systems was unremarkable. Pertinent negatives include the absence of fever, weight loss or gain, pruritus, redness, jaundice, chest pain, orthopnea, easy fatigability, weakness, abdominal pain, hematuria, dysuria, urinary frequency and urgency, and heat or cold intolerance.

On physical examination, the patient had swelling of the right eyelid that was non-erythematous, non-pruritic, non-pitting, and non-tender, without a central punctum. There were also swollen lips on the right side of the face, which were non-erythematous, non-pitting, and non-tender, without ulcerations. There was right anterior cervical lymphadenopathy (1 cm) without masses. The rest of the physical examination was unremarkable.

Course in the ward

Upon admission, the patient was hooked to intravenous fluids and placed on a hypoallergenic diet. A complete blood count with platelet count was performed, revealing normal hemoglobin, hematocrit, and white blood cell count with neutrophilic predominance and a normal platelet count (Table 1).

Medications started were diphenhydramine 30 mg IV every 8 hours (1 mg/kg dose), hydrocortisone 60 mg IV every 12 hours (2 mg/kg dose), and ranitidine 80 mg IV every 12 hours (15 mg/kg dose). The patient was referred to a pediatric allergist with an initial impression of angioedema, likely secondary to insect-bite hypersensitivity, and to consider hereditary angioedema. An additional medication, montelukast 4 mg chewable tablet, was also started once a day.

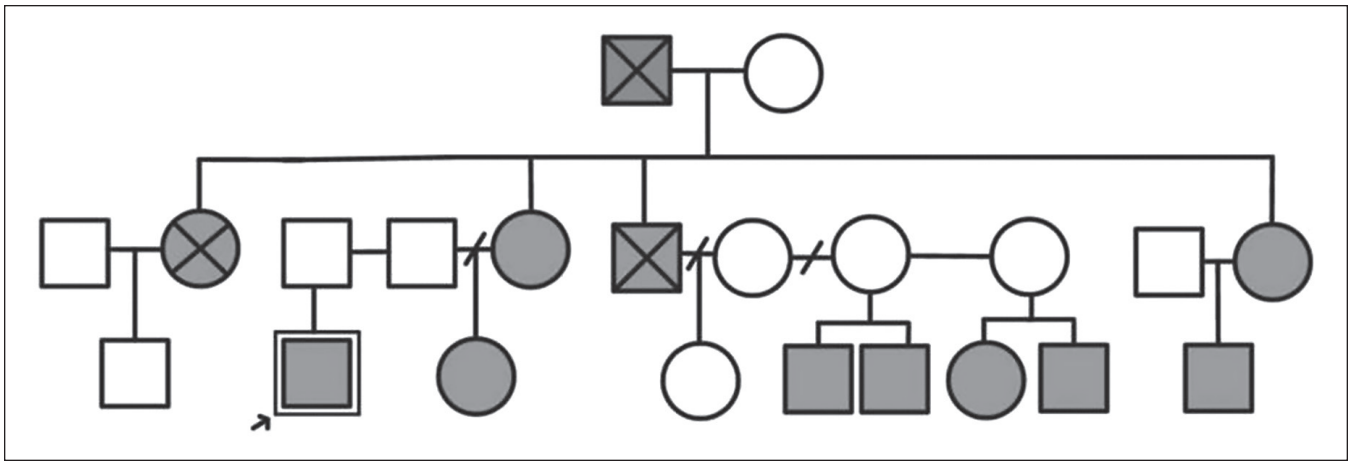

Figure 1. Family genogram.

Legend:

Male
Female
Deceased male
Deceased female
Male with HAE
Female with HAE
Index patient
Deceased male with HAE
Deceased female with HAE

Table 1. Complete blood count

	11/22/23
Hgb	12.1 g/dL
Hct	38.4%
WBC	8.2 cumm
RBC	5.44 M/uL
Platelet count	370 cumm
Neutrophils	84%
Lymphocytes	14%
Monocytes	2%
MCV	70.6 fL
MCH	22.2 pg
MHC	31.4%
MPV	8.8 fL

On the first hospital day, the patient had stable vital signs and showed minimal regression of facial swelling (Figure 2). During pediatric allergist rounds, the patient's history was reviewed, and tests for C1 inhibitor (level and function) and C4 levels were ordered. Hydrocortisone was switched to prednisone 10 mg/5 ml at 7.5 ml twice daily, and diphenhydramine was changed to Levocetirizine 0.5 mg/5 ml at 5 ml twice daily, while montelukast and ranitidine were discontinued. Tranexamic acid was started intravenously at 20 mg/kg/day.

On the second hospital day, the patient continued to have stable vital signs and exhibited marked improvement in facial swelling, now presenting with only minimal swelling of the right eyelid (Figure 2). The patient tolerated oral medications and was deemed fit for discharge.

After discharge, tests for C1 inhibitor level (126 mg/L, normal values 210-380 mg/L) and function (C1-inhibitor activity 26%, normal values 70-130%) were requested as an outpatient, and the results were below normal.

DISCUSSION

Etiology and pathogenesis

HAE is a rare autosomal dominant disorder due to either deficiency or dysfunction of the serine protease inhibitor (serpin) C1 inhibitor (C1-INH). It is described as recurrent episodes of angioedema without urticaria or pruritus.[2] The prevalence of HAE is estimated at approximately 1 individual per 60,000.[1,2] Males and females are affected equally, and there are no known differences in prevalence among ethnic groups.[3] There are two types of HAE. The first is HAE Type I, characterized by C1 inhibitor deficiency, which accounts for 85% of cases. The second is HAE Type II, characterized by C1 inhibitor dysfunction, accounting for only 15% of cases. In the patient's case, HAE Type I is strongly suspected, as all affected relatives have a C1 inhibitor value of zero in diagnostic tests.[1-4]

There are several triggers and exacerbating factors associated with HAE. The most common triggers are stress, either mental or physical, and dental procedures. Physical triggers include mild trauma, such as dental work, intubation, and tongue or ear piercing, which may induce local

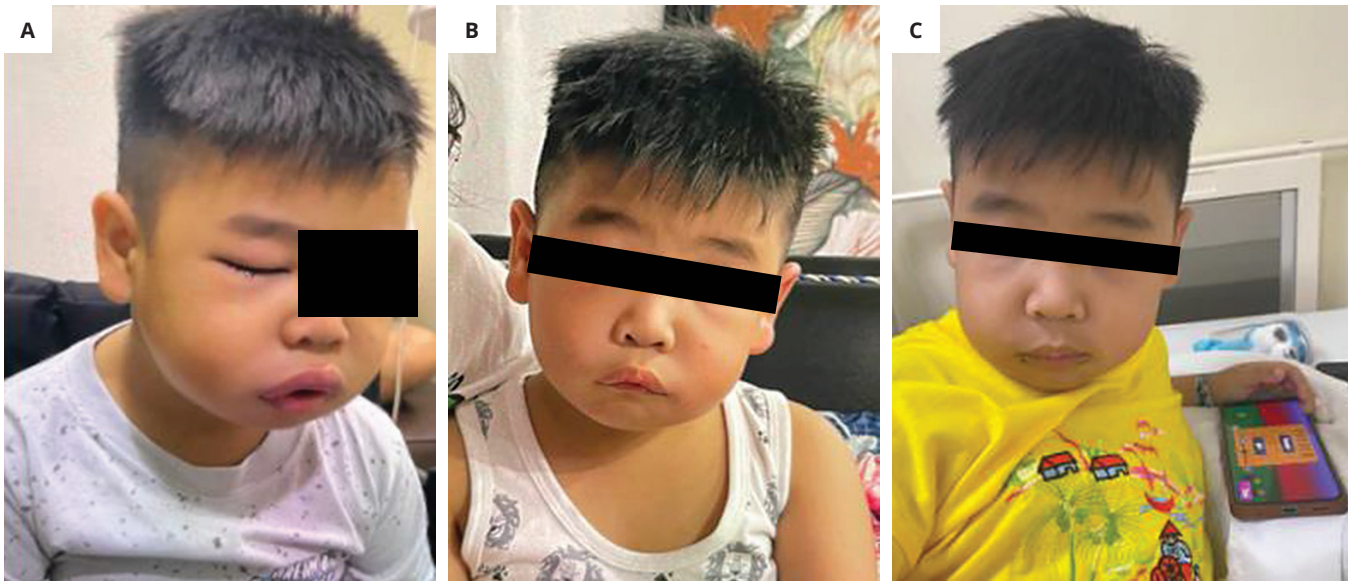

Figure 2. Clinical photographs of the patient during acute attacks at different time points during hospitalization. **(A)** Upon admission, **(B)** first day of treatment, **(C)** prior to discharge.

trauma and have been implicated in isolated case reports. In women, genital swelling can be precipitated by sexual intercourse, as well as activities like bicycle or horse riding. Infections and other stressors, including excitement, sleep deprivation, viral upper respiratory tract infections, bacteriuria, cold exposure, and prolonged sitting or standing, are also considered contributing factors.[3] In the case of our patient, the identified triggers are a lack of sleep and, to a lesser extent, an insect bite. The latter is thought to contribute not due to an allergen from the sting, but rather to the local skin trauma it induces. Although this patient is also being treated for angioedema secondary to insect bite hypersensitivity, classified as allergic/histaminergic angioedema, a study by Zotter and colleagues investigated trigger factors in HAE due to C1 inhibitor deficiency in 140 subjects. The study found that insect bites triggered HAE attacks in two patients, suggesting they could also be a trigger in our patient's case, as previously noted.[5]

Pathophysiology

Hereditary angioedema is an autosomal dominant disorder that leads to decreased levels of C1 esterase inhibitor. Normally, C1 esterase inhibitor acts as an acute-phase reactant that inhibits plasma kallikrein, which produces bradykinin. Under healthy conditions, C1 esterase inhibitor effectively inhibits kallikrein and prevents the production of bradykinin, a pro-inflammatory mediator. Therefore, C1 inhibitor helps decrease bradykinin levels under normal circumstances.[1,5,6]

In HAE, there is a deficiency of C1 esterase inhibitor, resulting in uninhibited kallikrein and increased bradykinin production. The excess bradykinin, which is proinflammatory, leads to vasodilation and increased vascular permeability. Additionally, C1 inhibitor deficiency or dysfunction results in low levels of complement component 4 (C4), because the C1 complex normally cleaves C4 in the classical complement pathway. This effect is further exaggerated in cases of C1 inhibitor deficiency. Molecular events leading to angioedema include the local activation of the contact system proteases, factor XII and plasma prekallikrein, on endothelial cell surfaces. An increased bradykinin level will cause bronchoconstriction, leading to a dry cough; vasodilation, leading to hypotension; increased vascular permeability, leading to angioedema; and cutaneous and gastrointestinal attacks, or worse airway compromise.[1]

Clinical manifestations

The age at which attacks of HAE begin varies, with rare reports of initial episodes occurring during the perinatal period. Approximately 40 percent of patients experience their first HAE attack before the age of 5, and 75 percent have their first attack by age 15, although repeated attacks in preadolescent children are uncommon. The onset of initial symptoms does not differ between boys and girls. HAE attack frequency ranges from weekly occurrences to one or two episodes per year.[1,2] In the case of the patient, the first attack occurred during his fourth year of life, with attacks occurring twice yearly. Many hereditary angioedema attacks involve only one site at a time, although multi-locational attacks can also occur. HAE attacks are self-limited, typically lasting two to five days, and can range in severity from inconvenient cutaneous swelling to life-threatening upper airway edema. Approximately 50 percent of patients experience

all three manifestations during their lifetime.[3,4] In the case of the patient, the right side of the face, including the lips, was affected, but the swelling eventually subsided and returned to normal on the third day. While this resolution may be associated with the steroids administered, it is important to recognize that HAE is self-limiting in most cases, typically lasting 2 to 5 days, as seen in our patient.

Prodromal symptoms of hereditary angioedema may include fatigue, nausea or vomiting, myalgias, and flu-like symptoms. Some patients develop skin changes characterized by a serpentine, mottled, and/or "chicken-wire" pattern of erythematous discoloration, commonly referred to as the HAE prodromal rash, as shown in the accompanying picture. Erythema marginatum occurs in approximately 42-58% of cases. Prodromal symptoms typically arise within 24 hours before the onset of angio-edema; however, not all prodromal symptoms lead to an angioedema episode.[2] In the case of our patient, no prodromal symptoms were experienced.

The characteristic features of angioedema attacks typically involve three anatomical locations: the skin (cutaneous attacks), the gastrointestinal tract (gastrointestinal attacks), and the upper airway. The angioedema builds over the first 24 hours, then gradually subsides over 48 to 72 hours. Swelling may last up to five days in some patients. Attacks can also last longer if the swelling spreads from one site to another. Cutaneous attacks of angioedema are common and can be temporarily disfiguring, although they are generally not dangerous. In a retrospective study of 221 patients, skin swelling was observed in 97 percent of 131,110 angioedema episodes. The extremities, face and genitals are the most commonly affected areas, although swelling can occur at any site.

The swelling typically occurs in nondependent areas and is non-pitting and non-pruritic. In addition to swelling, cutaneous attacks are often associated with pain and dysfunction. An episode usually begins in the skin with a peculiar tingling sensation or a feeling of fullness and irritation, followed by swelling and a sense of tightness within the next two to three hours. Facial and lip angioedema accounts for approximately 3 percent of episodes of subcutaneous edema. Swelling in these areas should be closely monitored by the patient or their caregivers, as up to 30 percent of episodes of upper airway edema are preceded by facial or labial edema. Gastrointestinal attacks of angioedema present with symptoms such as gastrointestinal colic, nausea, vomiting, and diarrhea, which result from bowel wall edema. Laryngeal swelling can occur in isolation or alongside swelling of the lips, tongue, uvula and soft palate. Initial signs and symptoms of upper airway attacks may include a sore, scratchy, or itchy throat; a sensation of tightness or a lump in the throat; dysphagia; voice changes; hoarseness; roughness of voice; or a resonant, "barky" cough. Laryngeal edema occurs in approximately half of all patients over their lifetimes; however, only a few percent experience recurrent episodes, and in a large retrospective study, laryngeal attacks accounted for less than 1 percent of all angioedema episodes. Laryngeal attacks are less common in patients over the age of 45, with tooth extraction and oral surgery being common triggers. In comparison to adults, children may experience asphyxia more rapidly due to their smaller airway diameter.[1-3] In our patient, there were no gastrointestinal or upper airway manifestations.

Diagnostics

In families with known C1 inhibitor deficiency hereditary angioedema (C1-INH-HAE), first-degree relatives, regardless of whether they are symptomatic or asympto-matic, should be screened for C1 inhibitor and C4 levels at the earliest convenience. Specimens can be obtained through umbilical cord blood in neonates and peripheral blood in infants and older children. Regardless of the initial test results, screening should be repeated after the child reaches one year of age. It is advised that, until a diagnosis of C1-INH-HAE is ruled out through two separate tests, with the second test performed after age 1, the pediatric patient should be considered to have inherited C1-INH deficiency. This repeated testing beyond the first year of life is necessary because complement levels in patients under one year old are typically lower than normal, which may lead to false positive results.[1,6]

A negative family history does not exclude the possibility of C1 inhibitor deficiency hereditary angioedema (C1-INH-HAE). Clinical suspicion of C1-INH-HAE-like symptoms at any age warrants screening, regardless of the presence or absence of family history. C1-INH-HAE screening includes evaluation of functional and antigenic C1-INH levels, as well as C4 levels. If the initial screening suggests C1-INH-HAE, a second test should be conducted to confirm the diagnosis. If testing indicates C1-INH-HAE, all first-degree relatives in the ascending line, including symptom-free individuals, should also be screened.[1]

In this patient, C1 inhibitor levels and function were found to be below normal, while C4 levels have not yet been tested. Given the patient's history of angioedema attacks and a strong family history of HAE, it is reasonable to consider that the patient has inherited C1 inhibitor deficiency or HAE until the C4 level results are obtained.

HAE can be identified through laboratory testing that reveals a deficiency of C1 esterase inhibitor, which leads to abnormally high levels of bradykinin. Patients with HAE typically exhibit low levels of C4, C1-INH protein, and C1-INH function. Low C1-INH levels result from a

deficiency associated with the absence of the SERPING1 gene. Furthermore, C4 levels should be monitored during suspected HAE episodes, but testing should only occur during acute attacks. Randomly checking the C4 level in HAE patients has a sensitivity of only 80%, although it can still support the diagnosis. Monitoring D-dimer levels may also aid in diagnosing an acute HAE attack, as these values may increase during such events. If any laboratory values are found to be less than or equal to 50% of normal, retesting should occur within 1 to 3 months to confirm accuracy and rule out any acute illness. Genetic testing can also diagnose HAE; however, it has limitations in predicting disease progression. A positive result from genetic screening for HAE does not necessarily indicate a patient's future symptoms or severity, as the same genotypic variation can manifest differently among individuals. To establish a diagnosis of HAE, patients should present with both clinical symptoms and positive laboratory findings.[1,2]

Treatment and prognosis

In some instances, attacks can be prevented through counseling, lifestyle modifications, and avoidance of triggering factors, particularly contact sports and activities that may involve physical trauma to tissues. While breastfeeding is recognized for its protective benefits against various diseases, it does not reduce or prevent C1-INH-HAE, or its associated symptoms.[1]

Prophylaxis begins with the identification and elimination or avoidance of precipitating factors. Therapeutic prophylaxis typically includes either short-term prophylaxis (STP) before events that increase the risk of precipitating an attack, or long-term prophylaxis (LTP) aimed at preventing attacks over an extended period. Ideally, for interventions that involve minor manipulation or may lead to tissue swelling, prophylaxis with 15 to 30 units per kilogram of plasma-derived C1-INH concentrate is recommended. Short-term prophylaxis with plasma-derived C1-INH should ideally be administered during the procedure or at least 1 hour before it, aiming to provide the treatment as close to the procedure as possible. However, since plasma-derived C1-INH is not available in the Philippines, we use fresh-frozen plasma (FFP) as an alternative, as it contains various proteins, including C1 inhibitor, which is deficient or dysfunctional in HAE. If licensed on-demand acute treatment medications are unavailable during planned procedures, the following options are recommended for short-term prophylaxis: oral attenuated androgens, primarily danazol at a dose of 2.5 to 10 mg/kg/day, with a suggested mean dose of 5 mg/kg/day (maximum 600 mg daily), or stanozolol and oxandrolone, which are used less frequently due to potential adverse drug reactions such as hypogonadism, increased cardiovascular event risk, fluid retention, and electrolyte imbalances. For major procedures or interventions that require airway manipulation, plasma-derived C1 inhibitor should be administered 1 to 6 hours before the procedure, with a second dose readily available. Again, due to the unavailability of plasma-derived C1 inhibitor, fresh frozen plasma serves as an alternative.

For long-term prophylaxis, most consider tranexamic acid the agent of choice in pediatrics; however, it is contraindicated in patients with a history of thromboembolism or known thrombophilia. The appropriate dose of tranexamic acid ranges from 20 to 50 mg/kg/day, split into two or three doses, with a maximum of 3 to 6 g/day. It is recommended to start at the lower dose and increase as needed to suppress events. Attenuated androgens, which stimulate the synthesis of C1-INH, are another option for long-term prophylaxis. C1-INH is a protein that regulates the complement system and bradykinin production. By increasing C1-INH levels, these medications help restore the balance of the complement system, thereby reducing the frequency and severity of angioedema attacks in patients with HAE. However, they are generally not considered long-term prophylaxis in pediatric patients before Tanner Stage V. After Tanner Stage V, attenuated androgens may be used while aiming to achieve the minimum effective dose. In the case of our patient, home medications included steroids and tranexamic acid. The steroids were not immediately discontinued due to a probable hypersensitivity to an insect bite and the need for tapering before discontinuation. Tranexamic acid was prescribed as a long-term prophylactic agent since it is the preferred choice for such cases, given that HAE is highly suspected. Since HAE is classified as a non-histaminergic angioedema, it will not respond to higher doses of steroids and antihistamines; therefore, plasma-derived C1-INH concentrates are approved for the acute treatment of C1-INH-HAE in pediatric patients, with a recommended dose of 20 units per kilogram. Additionally, ecallantide is FDA-licensed for the acute treatment of HAE attacks in patients with C1-INH-HAE aged 12 years and older, administered subcutaneously at a dose of 30 mg, and should only be given by a healthcare professional experienced in managing anaphylaxis due to the associated risk. Icatibant, while not licensed for pediatric use, is approved for acute treatment in patients aged 18 years and older, and a clinical trial for pediatric patients is currently ongoing. If licensed on-demand acute treatment medications are unavailable or inaccessible, 10 ml/kg of solvent-detergent plasma may be used on demand, as it is preferred over fresh frozen plasma for its reduced risk of transfusion-transmitted diseases. In this case, the patient's mother reported that they do not administer any medications during acute attacks and wait for the angioedema to subside; however, during this admission, she expressed concern about the potential for attacks, particularly due to family members who have died from HAE attacks, which included upper airway manifestations such as laryngeal spasm.[1,4-6]

The prognosis for patients with C1-INH-HAE varies; once attacks begin, they typically persist throughout the patient's life, although therapy can dramatically reduce the frequency of these attacks. Before effective therapies were introduced for HAE, up to one-third of patients died from asphyxiation. Despite the availability of effective treatments, deaths related to laryngeal attacks still occur regularly, although data on this are limited. A study of patients from Austria, Switzerland, and Germany published in 2004 reported a mortality rate as high as 13 percent.[2]

CONCLUSION

Hereditary angioedema is a rare genetic disorder, affecting about 1 in 50,000 people, caused by a deficiency or dysfunction of the C1 esterase inhibitor. It is inherited in an autosomal dominant pattern and presents in three types, all leading to recurrent, potentially life-threatening episodes of non-pitting edema. Symptoms usually start in childhood or early adulthood, often worsening around puberty, and are driven by increased bradykinin production, which causes vascular permeability and edema. Clinical suspicion of C1 inhibitor deficiency, hereditary angioedema-like symptoms at any age, warrants screening, regardless of the presence or absence of family history. If the initial screening suggests C1-INH-HAE, a second test should be conducted to confirm the diagnosis. Patients with HAE usually have low levels of C4, C1-INH protein, and C1-INH function, often due to a deficiency linked to the absence of the SERPING1 gene. The prognosis for patients with C1 inhibitor deficiency hereditary angioedema varies, with attacks typically persisting for life, though therapy can significantly reduce their frequency.

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Ethical Consideration
Patient consent was obtained before the submission of the manuscript.

Statement of Authorship
The authors certified fulfillment of the ICMJE authorship criteria.

Author Disclosure
The authors declared no conflict of interest.

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