Case Report
Eurasian Journal of Critical Care

Hereditary Angioedema and Allergic Reaction Due to Fresh Frozen Plasma

Serdar Derya, Şükrü Gürbüz, Muhammed Ekmekyapar, Hakan Oğuztürk, Muhammet Gökhan Turtay, Neslihan Yücel, Abdullah Keyfo Kama
Department of Emergency Medicine, Inonu University, Malatya, Turkey

Abstract

Hereditary angioedema (HA) is a rare disorder characterized by recurrent angioedema attacks and autosomal dominant transmission. Plasma C1 occurs due to esterase inhibitor deficiency. Angioedema is most commonly seen in the extremities, oropharynx, and visceral organs. Intestinal mucosal edema can cause temporary obstruction and severe abdominal pain that can interfere with acute abdomen. It should be kept in mind that antihistamines, stereoids, androgens, FFP (Fresh frozen plasma) and C1 esterase inhibitors used in the treatment of these patients may also develop allergies and these drugs should be used with caution. We aimed to present a 51 years old female patient (Figure-1) with a diagnosis of hereditary angioedema in our case which developed FFP (Fresh frozen plasma) allergy.

Key Words: Hereditary angioedema, FFP (Fresh frozen plasma), allergy

Introduction

Hereditary angioedema (HA) is a rare autosomal dominant disorder characterized by recurrent angioedema attacks. It is caused by the lack of plasma C1 esterase inhibitor. The disease that may begin in puberty, sometimes in childhood, is a condition characterized by subcutaneous and mucosal edema[1]. In patients with HA attacks, the concentrations of C4 and C2 decrease while the concentrations of C3 and C1 remain normal. Thus, there are also those who recommend C4 measurement as a screening test for hereditary angioedema[2]. C1, C2, C4 decrease chronically in case of the absence of this enzyme which functions in the complement system. Complement deficits become more apparent during attacks. We aimed to present a 51-year-old female patient who has a diagnosis of hereditary angioedema and developed FFP (fresh frozen plasma) allergy in our case.

Case report

A 51-year-old female patient with complaints of swelling and erythema on the face and neck applied to the emergency department. The patient has a history of HA. There was edema and erythema in the neck and face, no uvula edema. The patient ‘s vital parameters including blood pressure, pulse, respiratory rate and fever were respectively measured as 130/90 mmHg, 74 /min, 16/min and 36° C. In the laboratory tests of the patients, white blood cell (WBC), hemoglobin (HB), hematocrit (HCT) and platelets (PLT) from hemogram parameters were respectively 11,4 10^3/ml, 16,6 g/dl, 50,5% and 280 10^3/ml, glucose and C-reactive protein (CRP) from biochemical parameters were respectively 143 mg/dl and 0,48 mg/dl (normal range is 0 to 0,35) and the other parameters were normal. She had developed allergies to antihistamines and steroids during previous attacks. Patient with hereditary angioedema was consulted dermatology department. Dermatology suggested giving 2 units of FFP to the patient. We started giving FFP to the patient. The patient who received FFP treatment stated that she experienced a feeling like a lump in his throat, burning sensation on her face and difficulty in swallowing. FFP treatment was stopped immediately. It was seen that swelling and erythema in the face increased and uvula edema developed during the patient’s control examination (Figure 1). Then, 1000 IU (2 vials) of C1 esterase inhibitor was given to the patient. It was seen that the patient’s complaints and swelling of the uvula decreased. While FFP therapy is given to patient with hereditary angioedema, it should be absolutely considered that the patient may develop allergies to the FFP and the patient should be closely monitored with attention.

Corresponding Author: Şükrü Gürbüz **e-mail:** sukrugurbuz@gmail.com
Received: 17.01.2019 • **Accepted:** 21.02.2019

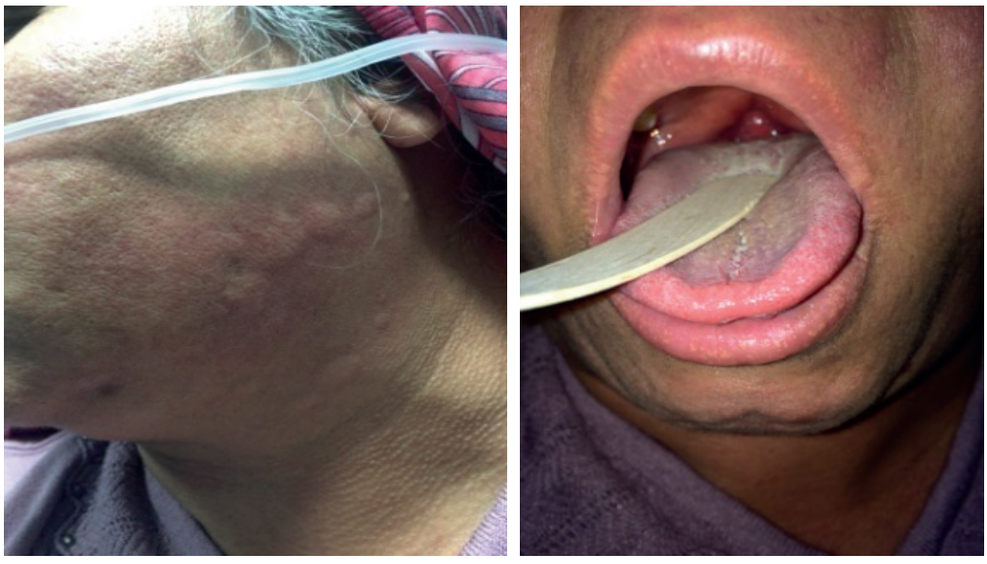

Figure 1. Increased redness and uvula edema after FFP treatment

Discussion

HA is a rare autosomal dominant disorder characterized by recurrent angioedema attacks. It is caused by the lack of plasma C1 esterase inhibitor. The disease that may begin in puberty, sometimes in childhood, is a condition characterized by subcutaneous and mucosal edema[1]. HA was first described by William Osler in 1888[3]. C1 INH is a functional or quantitative complement that regulates the initial proteins of the classical complement system. It Is a rare autosomal dominant (AD) disease characterized by the lack of the C1 complement inhibitor (C1 INH)[4-5]. 75% of the cases have family history and no gender difference similarly to the other AD diseases. HA is a disease characterized by attacks of edema which is well-circumscribed and non-pitting in the extremities, larynx, face and the body[5-6]. It is possible to confirm the diagnosis by detecting C1 INH levels. HA is usually self-limited and localized edema is prevalent due to the reversible increase in vascular permeability in its clinic. Patients have complaints such as subcutaneous edema, abdominal pain, nausea, vomiting, diarrhea, dysphagia and dysphonia. The most feared complication of hereditary angioedema is laryngeal edema. Laryngeal edema usually presents at older ages and occurs in less than 1% of attacks. Serum C4 levels are low during attacks and non-attack periods in nearly all cases with HA. C1 and C3 levels are normal. FFP may be useful for preoperative short-term prophylaxis and an acute attack. Researches on recombinant C1 INH, recombinant kallikrein inhibitor and bradykinin receptor antagonists continue. After the C1 esterase inhibitor administered to our patient, it was seen that the complaints of the patient and edema of the uvula were regressed.

Conclusion

As a result, patients with HA may visit the emergency services because of the complaints such as erythema, edema and shortness of breath. It should be kept in mind that allergies to antihistamines, steroids, androgens, FFP and C1 esterase inhibitors used in the treatment of these patients may develop and these drugs should be used with caution.

References

1. Huang YT, Lin YZ, Wu HL et al: Hereditary angioedema: a family study. Asian Pac J Allergy Immunol 2005;23:227-33.
2. Isselbacher KJ. Harrison's principles of internal medicine, 15th ed. McGraw-Hill, 2001
3. Osler W. Hereditary angio-neurotic oedema. Am J Med Sci 1888;95:362-7.
4. Frank MM. Hereditary angioedema: a half century of prog¬ress. J Allergy Clin Immunol 2004;114:626-8.
5. Witschi A, Krahenbühl L, Frei E, Saltzman J, Spath PJ, Müller UR. Colorectal intussusception: An unusual gastroin¬testinal complication of hereditary angioedema. Int Arch Allergy Immunol 1996;111:96-8.
6. Frank MM. Herditary angioedema: a half century of progress. J Allergy Clin Immunol 2004;114:626-8.