

Angioedema triggered by pulp extirpation—a case report

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Abstract

Background C1-esterase inhibitor deficiency results in episodes of non-allergic edema of parts of the body. Edema of the face may be triggered by dental therapy.

Case report We report a case of C1-esterase inhibitor deficiency which was detected in a 42-year-old woman. The patient was completely unaware that she had this disorder or of any related family history, and the patient developed an intense facial angioedema after pulp extirpation of lower premolar tooth.

Discussion In this case, the diagnosis of angioedema due to C1-esterase inhibitor deficiency was established at a later stage. The differing causes of C1-esterase inhibitor deficiency are briefly discussed and the treatment modalities outlined.

Keywords C1-esterase inhibitor deficiency · C1-inhibitor deficiency · Angioedema

Introduction

Angioedema due to C1-esterase inhibitor (C1-INH) deficiency is characterized by episodes of edema of a localized area of the body. Patients may also have episodic swelling of the bowel wall leading to abdominal symptoms [1]. Of interest to dentists and facial surgeons is the fact that perioral edema can be triggered by facial trauma or dental therapy, which on involvement of the upper airway can lead to asphyxiation. The reported triggers for angioedema

include extraction of teeth [2], pulp extirpation [3], or even dental impressions [4]. The disease manifests due to a quantitative or functional deficiency of C1-INH.

We present a case in which the sequela was encountered after pulp extirpation.

Case report

Forty-two-year-old female was admitted to the hospital emergency with a swelling of the entire left side of the face and lower lip (Fig. 1). The swelling began when the patient had returned home after pulp extirpation of the lower left premolar under local anesthesia. At first, she noticed a small swelling over the lower lip that gradually increased throughout the day. With vital signs being normal, patient developed a firm, non-tender diffuse swelling extending from infraorbital rims to the lower border of mandible, bilateral submandibular, and submental regions with eversion of the upper and lower lips (Fig. 2), and obliterating upper and lower labial/buccal vestibule. Laboratory parameters revealed Hb 10.9 mg/dl, total WBC 6,730 cells/cumm, WBC differential count N62 L34E01M03B0, and ESR 81 mm at 1 h.

With possibility of allergy, resuscitation began with administration of hydrocortisone sodium succinate, methyl prednisolone acetate, ranitidine, and pheniramine maleate. The oxygen saturation was constantly monitored and endotracheal intubation planned; should that fail, surgical airway access. However, the patient maintained her oxygen saturation without respiratory distress.

A detailed medical history revealed patient had suffered recurrent bouts of abdominal pain, without an obvious cause, and ascites. Five years ago, an “insect bite” near the lips had resulted in facial edema.

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Fig. 1 Angioedema after pulp extirpation

Over the next 48 h, there was gradual regression of edema (Fig. 3) and a patch test was negative; further parameters revealed serum IgE of 209.4 IU/ml (reference up to 200), N96, L04, B0. Because of the edema that developed following pulp extirpation, slow response to therapy with corticosteroids and antihistamines, and history of abdominal disorders, the possibility of angioedema due to C1-INH deficiency was considered. The C1-INH level was tested and found to be 0.05 g/L (reference range and units, 0.21–0.39 g/L).

Discussion

The C1-INH, normally, prevents complement cascade auto-activation and has an inhibitory effect on activated kallikrein (generated through the contact or kallikrein–kinin system) [5]. When C1-INH activity is deficient, there is vasodilation and increased vascular permeability. The clinical result is a non-pitting edema. The attack may last from 2 to 5 days. C1-INH deficiency in a majority of cases is an inherited autosomal

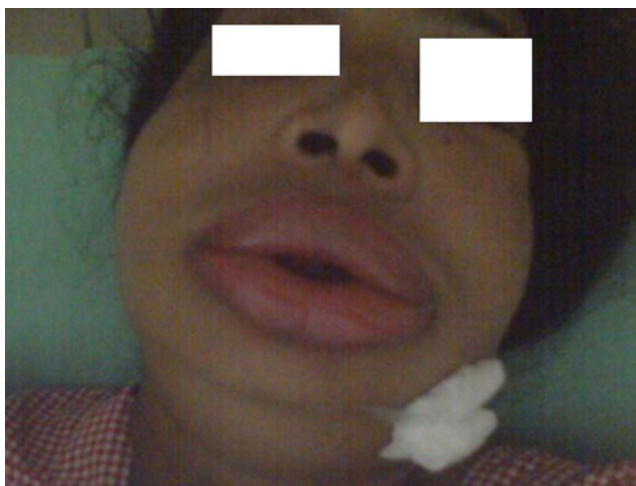


Fig. 2 Patient at the time of maximum swelling



Fig. 3 After 48 h, edema resolved

dominant condition, referred to as “hereditary angioedema” (HAE). In patients without a family history, it is thought to be a spontaneous mutation [1]. HAE may be type I or type II depending on whether there is a quantitative or a functional deficiency of C1-INH, respectively [6]. Further, another type of inherited angioedema is described in female patients who showed normal C1-INH level and function [7, 8]. Angioedema can also occur because of acquired disorder, acquired angioedema (AAE) [9]. The genetic mutations of HAE are not present. Type I AAE occurs in patients with B cell lymphoproliferative, autoimmune, infectious, or neoplastic diseases where C1-INH is rapidly consumed and synthesis cannot keep up. In type II AAE, auto-antibodies directed against the C1-INH molecule lead to its inactivation [5]. Another clinically relevant form of angioedema occurs in some patients on angiotensin converting enzyme (ACE) inhibitor therapy [10].

The diagnosis and treatment of dental patients who develop these sequelae can be very difficult until the diagnosis is established. In this case, the patient was completely unaware of the condition and any related family history. The increasing edema after dental treatment leads to initial consideration of allergy. Allergic angioedema, however, is often associated with urticaria. It is mediated by the release of histamine; hence, it responds to treatment with corticosteroids and antihistamines [8]. A lack of response to these drugs coupled with the medical history suggested the possibility of angioedema due to C1-INH deficiency.

Treatment with epinephrine, corticosteroids, and antihistamines may not resolve an acute attack of angioedema due to C1-INH deficiency. The airway has to be secured, either through endotracheal intubation or surgically, in patients with airway difficulty. A rapid resolution of the edema is brought about by replacement with C1-INH concentrates,

when available. C1-INH can also be replenished by transfusing fresh frozen plasma; however, an exacerbation of symptoms can occur. Attenuated androgens and anti-fibrinolytics have been used for therapy and prophylaxis [1, 5, 8]. ACE inhibitor therapy-related angioedema is managed by replacing ACE inhibitors by angiotensin II receptor blockers or by the medication icatibant, a bradykinin B2 receptor antagonist [11].

The patient was referred for counseling, therapeutic, and prophylactic measures to manage the disorder before she sought further dental treatment.

Conflict of interest declaration NIL

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