

Successful enucleation of radicular cyst after prophylaxis in a hereditary angioedema patient

Hereditör anjioödemli bir hastada profilaksi sonrasında radiküler kistin başarıyla çıkarılması

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Abstract

Hereditary angioedema (HAE) due to C1-INH deficiency (C1-INH-HAE) is an autosomal dominant inherited disease characterized by recurrent skin and mucosa swellings that commonly involve the extremities, intestines, face, genital area and upper airways. Dental procedures are important attack triggers in C1-INH-HAE patients. Attacks following dental procedures are commonly localized to the face and larynx. Laryngeal edema can cause death by asphyxiation. A 48-year-old female patient was admitted to our hospital because of the excessive accumulation of calculus on the teeth which was noticed during routine dental examination. At the oral examination calcified dental plaque, bleeding by gingival probing, and discoloration of the teeth #31-32 were detected. By radiographic and clinical findings radicular cyst pre-diagnosis was made, and it was decided to enucleate the cyst. However, the patient's medical history revealed that she was diagnosed with C1-INH-HAE at the age of 40. To prevent possible facial and laryngeal edema, 1000 units of C1 inhibitor concentration was given 1 hour before the procedure. Under local anesthesia, the lesion was totally enucleated with its capsule. The risk of having attack after tooth extraction is about 37.5% and nearly 1/3 of these attacks are associated with laryngeal edema in patients with C1-INH-HAE. However, prophylaxis with C1 inhibitor concentration led to a 44.1% reduction in angioedema attacks on a per-patient basis. With this case, we wanted to draw attention to the importance of questioning the patient and his/her family for recurrent nonpruritic skin and mucosa swelling and abdominal pain attacks before surgical interventions to the mouth, neck, and throat area.

Key words: hereditary angioedema, C1 inhibitor prophylaxis, laryngeal edema

Özet

Hereditör anjioödem C1 inhibitör protein eksikliğine bağlı gelişen, tekrarlayan, kaşıntısız, deri ve mukoza şişkinlikleri ile karakterize genellikle, ekstremiteleri, barsak mukozasını, yüzü, genital bölgeyi ve üst hava yollarını tutan otozomal dominant kalıtılan bir hastalıktır. Diş ve diş etine yapılan müdahaleler ile özellikle yüz ve üst solunum yollarına lokalize ataklar oluşabilmekte ve hayatı tehdit edici larinks ödemi gelişebilmektedir. 48 yaşındaki kadın hastanın rutin kontrolünde diş etlerinde anormal tartar birikimi olduğu görüldü. Radyolojik incelemede 31 ve 32 nolu dişlerin apikal köklerinde 2x15 mm boyutlarında radyolüsen lezyon saptandı. Radiküler kist ön tanısı ile kist enükleasyonu kararı verildi. Hastanın anamnezinde 40 yaşında hereditör anjioödem tanısı konulduğu ve yılda 2 kez

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abdominal ve/veya ekstremitte atağı geliştirdiği öğrenildi. Yüzde ve larinkste gelişebilecek olası atağı önlemek amacı ile işlemden bir saat önce C1 inhibitör konsantrasyonu 1000 ünite verildi. Lokal anestezi eşliğinde radiküler kist kapsülü ile birlikte çıkarıldı, patolojik incelemede kapsülün epitel hücresi ile çevrili olduğu ve bu bulgunun da tamıyla uyumlu olduğu görüldü. Hastamızda işlem sonrası atak oluşmadı. Herediter anjioödem hastalarında diş çekimi sonrası atak gelişme riski ortalama %37.5'dur ve gelişen her 3 ataktan biri larinks bölgesini tutmaktadır. Diş çekimi sonrası gelişen ataklar işlemden ortalama 14.3 saat sonra ortaya çıkar (en erken 1 saat en geç 72 saat). C1 inhibitör konsantrasyonu ile yapılan profilaksi atak oluşumunu %44 oranında azaltmaktadır. Yüz, ağız, boğaz bölgesine yapılacak cerrahi girişimler öncesinde hastanın tekrarlayan anjioödem ve karın ağrıları açısından sorgulanması gerekmektedir.

Anahtar kelimeler: herediter anjioödem, C1 inhibitör profilaksisi, laringeal ödem

Introduction

Hereditary angioedema (HAE) due to C1-INH deficiency (C1-INH-HAE) is an autosomal dominant inherited disease characterized by recurrent skin and mucosa swellings that commonly involve the extremities, intestines, face, genital area and upper airways.¹ C1-INH-HAE is resulting from a mutation of the *SERPING1* gene encoding for C1-INH. In C1-INH-HAE, the reduced inhibitor effects of C1-INH on plasma contact system leads to increased liberation of bradykinin from high-molecular-weight kininogen (HK). There are two types of C1-INH-HAE; in type I mutations resulting in the production of truncated or misfolded proteins that are not secreted efficiently localized throughout the gene lead to low C1-INH concentrations and therefore low C1-INH function, in type II mutations resulting in the production of the dysfunctional C1-INH protein occur commonly at exon 8 that decode the reactive center and two critical hinge region lead to low C1-INH function despite normal levels of antigenic C1-INH.² Dental procedures are important attack triggers in C1-INH-HAE patients. Attacks following dental procedures are

commonly localized to the face and larynx.³ Laryngeal edema can cause death by asphyxiation.¹

Case report

A 48-year-old female patient was admitted to our hospital because of the excessive accumulation of calculus on the teeth which was noticed during routine dental examination. At the oral examination calcified dental plaque, bleeding by gingival probing, and discoloration of the teeth #31-32 were detected, however there was no periodontal pocket and teeth mobility. By radiographic examination a radiolucent lesion with a defined border of 2x15 mm at the apical roots of teeth #31 and #32 was discovered (Fig. 1). In the electrical pulp test, teeth #31 and #32 were found to be devital. Patient had pre-existing root canal treatment on tooth #32. Root canal treatment for tooth #31 was recommended as soon as possible.

The region on the vestibular gingiva of teeth #31 and #32, was compatible with the radiological location of the lesion, could be fluctuated, and bone expansion was detected. All these findings led us to the pre-diagnosis of radicular cyst, and it was decided to enucleate the cyst. One week prior to the operation, the patient was trained in cleaning teeth and gums.

However, the patient's medical history revealed that she was diagnosed with C1-INH-HAE at the age of 40. Her diagnostic laboratory values were concordant with type I C1-INH-HAE with low levels of C1-INH, 10 mg/dL (normal, 21-39 mg/dL); C1-INH function, 18.3% (normal, 70-130%) and near normal C4, 10 mg/dL (normal, 10-40 mg/dL) but normal C1q, 248 µg/mL (normal 100-300 µg/mL). Her angioedema symptoms began at 27 years old and until now, she has usually experienced biannually peripheral angioedema attack, but just one genital edema attack and one abdominal attack but no facial or laryngeal attack.

The enucleation of cyst procedure was explained to the patient. To prevent possible facial and laryngeal edema, 1000 units of C1 inhibitor concentration, (Cinryze, Takeda, Switzerland) was given 1 hour before the procedure. 2% articaine HCL solution was used for local an-

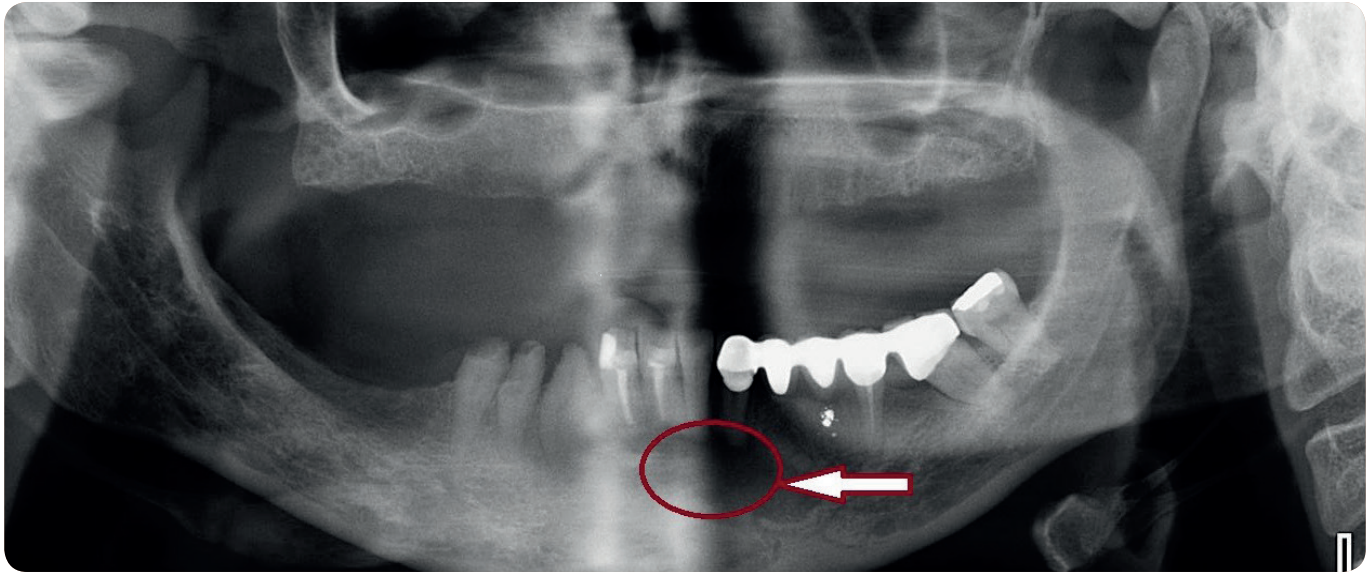


Fig. 1. The preoperative orthopantomograph radiograph showed large periapical radiolucency associated with teeth #31-32

esthesia, and then the lesion was totally enucleated with its capsule and wound margins were primary closed. Radicular cysts are commonly paved with non-keratinized squamous epithelium.⁴ Compatible with this information postoperative histopathological examination revealed that the cyst capsule was covered with squamous epithelium. In the postoperative period, the patient was treated with amoxicillin/clavulanic acid 1000 mg bid and diclofenac potassium 50 mg bid treatment for 7 days long. On the 7th day, it was observed that the operation area improved in a healthy way and sutures were removed. Radiological follow-up was recommended every 3 months.

Discussion

Hereditary angioedema (HAE) is an important disease that dentist should have knowledge about; because surgical interventions to the mouth and throat area may trigger skin and mucosa swellings that bear the risk of laryngeal edema.³ Seven cases with fatal outcomes have been reported after tooth extraction in patients with HAE.⁵⁻⁷ However, not all dental surgery procedures, including tooth extraction, are followed by an acute attack. The risk of having attack after tooth extraction is about 37.5% and nearly 1/3 of these attacks are associated with laryngeal edema. The mean time between tooth

extraction sessions and the onset of symptoms was 14.3 hours (range 1-72 hours).³ In the literature, it has been reported that fresh frozen plasma, antifibrinolytics, attenuated androgens, and C1-INH concentrate can be used in prophylaxis.⁸⁻¹¹ However HAE symptoms were reported despite prophylaxis in 20.8% patients.³

Radicular cysts are inflammatory cysts of the jaws and are the most common form of odontogenic cysts. The progression of the caries cavity to pulp and formation of chronic inflammatory tissue can cause radicular cyst formation. Epithelial cell rests of Malassez are stimulated via several inflammatory mediators and cyst formation is started following degeneration of the cells in the center of the expanding epithelial tissue. Most of the radicular cysts are asymptomatic, however especially infected cysts can cause pain.⁴ In the literature, there are no reported cases of C1INH-HAE who underwent dental cyst enucleation. We successfully performed enucleation procedure via 1000 Unites C1 inhibitor concentrate prophylaxis therapy an hour prior. Because of no facial and laryngeal edema history of our patient, prophylaxis therapy may not be considered necessary. However, it was decided that prophylaxis should be performed since the risk of laryngeal edema increases especially after dental procedures and since the patient may have the first attack of laryngeal edema. On a per-pa-

tient basis, prophylaxis with C1 inhibitor concentration led to a 44.1% reduction in angioedema attacks.³

With this case, we wanted to draw attention to the importance of questioning the patient and his/her family for recurrent nonpruritic skin, mucosa swelling and abdominal pain attacks before surgical interventions to the mouth and neck and throat area. Prophylaxis treatment of hereditary angioedema patients can prevent the risk of fatal laryngeal edema after dental intervention.

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