Case of Unrecognised of Maxillary Adenoid Cystic Carcinoma

Slučaj neprepoznatog adenoidnoga cističnog karcinoma u gornjoj čeljusti

Abstract
Adenoid cystic carcinoma is a less commonly diagnosed cancer that may affect the major or minor salivary glands. We present a 70 year old male patient who was admitted to the Department of Oral Medicine, School of Dental Medicine in Zagreb, Croatia due to pain in the right maxilla. In this case we report a case of the patient with unilateral pain in the maxilla & eye which lead to the diagnosis of adenoid cystic cancer without any visible oral lesions.

Introduction
Adenoid cystic carcinoma is a rare malignant tumor with an incidence rate of less than 1% of all head and neck malignancies (1). It constitutes approximately 29.6% of minor salivary gland tumors (2) and affects both males and females, without any sex predilection, and usually occurs after the fifth decade of life (3). The palate is the most common site of intra-oral lesions. In the major salivary glands, adenoid cystic carcinoma most commonly affects the parotid and submandibular glands, while palate is most frequently affected in the area of minor salivary glands, followed by the floor of the mouth, tongue and lip. In some cases it can present in the jaws as a primary intraosseous tumor (4). Characteristic signs of adenoid cystic carcinoma are slow growth pattern, tendency to local recurrences, postponed appearance of the distal metastases as well as neural invasion (5). The most important prognostic factors include tumor size, grade, stage, lymph node involvement, neural invasion and margin status (6). Diagnosis is based on clinical examination, histopathological analysis of a biopsy specimen and imaging techniques.
In this report, we present a case of sharp unilateral pain in the maxilla & eye in whom diagnosis of adenoid cystic cancer was established albeit no visible oral lesions could be seen.

Case report

A 70 year old male patient was admitted to the Department of Oral Medicine, School of Dental Medicine in Zagreb, Croatia in April 2017 due to pain in the right maxilla. In March 2016, he went to an ear, nose and throat (ENT) specialist due to pain in the right maxilla and a CT scan of the paranasal sinuses was obtained. Speckled zones of bone demineralization of the distant part of the right side of the hard palate were found. Since no soft tissue pathology could be seen, the patient was sent to MRI examination of the head which he never did. Our clinical examination revealed a mild asymmetry of the hard palate, therefore a panoramic image was taken. It showed a mass on the right side of the maxilla and the cheek (Figure 1). Furthermore, the patient was admitted to the Emergency Ophthalmology Department due to pain in the right eye. The ophthamologist treated the patient’s glaucoma and recommended the use of ultrasound for diagnostic imaging of the eye, which the patient did not perform. Six months after the first examination at our Department, he was admitted again and tumors thickening of the right maxilla could be noticed (Figure 2). He was immediately referred to a maxillofacial surgeon and a biopsy of palatal swelling was taken. A histopathological analysis revealed a tumor of a minor salivary gland, composed of both cribriform and tubular areas of atypical cuboidal epithelial cells with fossae of central necrosis within the cribriform areas. The final diagnosis was established. It was an adenoid cystic carcinoma (Figure 3).

The MSCT of the head, neck, and thorax examination was performed by standard recording techniques with 3D reconstructions. On the transitions between the head and the neck in the projection of the maxillary antrum to the right, and on the right half of the nasal cavity, a soft neoplastic heterogeneous contrast-absorbed process of about 48 mm in diameter was shown. Cranio-caudal dimension of the lesion was about 70 mm with invasion into the right orbit and the middle skull to the anterior part of the cavernous sinus. A dorsal lesion went to the right half of the sphenoidal sinus (Figure 4). On both sides of the neck, in region II, more oval lymph nodes without pathology were found.

Due to the size of the lesion and structures compromised, the tumor was inoperable, therefore, the patient was treated by radiotherapy. Radiation dose was 70 Gy divided at 35 fractions. After radiotherapy, the tumor has greatly reduced its size (Figure 5).

Discussion

The differential diagnosis of aggressive intraoral lesions should, among others, consider various salivary gland tumors. In case of an aggressive lesion which involves hard palate, differential diagnoses should involve adenoid cystic carcinoma of the minor salivary glands (2). Adenoid cystic

U ovom prikazu predstavljamo slučaj bolesnika s jednostranom oštrom boli u gornjoj čeljusti i oku kojem je postavljena dijagnoza adenoidnog cističnog karcinoma, a bez provjera u usnoj šupljini.

Prikaz slučaja

U Zavodu za oralnu medicinu Stomatološkog fakulteta Sveučilišta u Zagrebu (Hrvatska) u travnju 2017. godine došao je 70-godišnjak zbog bolova u desnoj strani gornje čeljusti. U ožujku 2016. pregledao ga je otorinolaringolog zbog bolova sa desne strane gornje čeljusti i oku. Na OPG-u bila je vidljiva masa na desnoj strani gornje čeljusti i obraz (slika 1.). Nadalje, bolesnik je hitno upućen na oftalmološki odjel zbog bolova u desnem oku. Oftalmolog je liječio bolesnikov glaukom i preporučio ultrazvuk oko koji bolesnik nikad nije učinio. Šest mjeseci nakon prvog pregleda u našem zavodu, bolesnik je ponovno učinio se učio željezni sadežanje desne strane gornje čeljusti (slika 2.). Odmah je poslan maksilo-facialnom kirurgu te je učinjen biopsija onkologije na gornjoj čeljusti. Histopatološka analiza pokazala je tumor malih žlijezda slinovnica sastavljen od kribriformnih i tubularnih područja atipičnih kuboidalnih epitelnih stanica s jamicama sredinjske nekrozne unutar kribriformnih područja. Postavljena je konačna dijagnoza – adenoid cističnog karcinoma (slika 3.).

Učinjen je MSCT glave, vrata i toraka standardnim tehnikama snimanja s 3D rekonstrukcijom. Na presjecima glave i vrata u projekciji antruma maksile desno, te desne polovine nosne šupljine, vidljiv je mekotovski neoplastički heterogeno kontrastno imbiniran proces od oko 48 mm u promjeru. Kranio-kaudalna veličina lezije bila je otrpilike 70 mm, s invazijom u desnu orbitu i srednju lubansku jamu do prednjeg dijela kavernoznog sinus. Dorzalno se lezija pruža do desne polovine sfenoidnog sinus (slika 4.). Na obje strane vrata u regiji II vidljivo je više ovalnih limfnih čvorova bez patoloških procesa.

Posljedično veličini lezije i komponentarnim strukturama, tumor je bio inoperabilan te je bolesnik liječen zračenjem. Doza zračenja iznosila je 70 Gy i bila je raspodijeljena u 35 frakcija. Nakon liječenja zračenjem tumor se značajno smanjio (slika 5.).

Rasprava

Diferencijalna dijagnoza agresivnih intraoralnih lezija, između ostalog, uključuje različite tumorne žlijezda slinovnica. Ako agresivna lezija obuhvaća tvrdo nepce, diferencijalna dijagnoza mora uključivati adenoidni cistični karcinom malih žlijezda slinovnica (2). Ta novotvorina sporo i podmuklo ra-
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Slika 1. OPG je pokazao masu na desnoj strani gornje čeljusti
Figure 1 OPG showed a mass on the right side of the maxilla

Slika 2. Tumorsko zadebljanje na desnoj strani gornje čeljusti koje zahvaća alveolar greben i tvrdo nepce od regije 11 do 18; na mekom nepcu mogu se uočiti teleangiektazije
Figure 2. Tumorous thickening of the right maxilla which involves alveolar ridge and the hard palate extending from the region 11 to 18. Teleangiectasia can be noticed on the soft palate.

Slika 3. Adenoidni cistični karcinom sastavljen od kribriformnih i tubularnih područja atipičnih kuboidalnih epitelnih stanica (HE x 100)
Figure 3 Adenoid cystic carcinoma composed of both cribriform and tubular areas of atypical cuboidal epithelial cells (HEx100).

Slika 4. MSCT glave i vrata; lezija na nepcu širi se u meko tkivo obraza, u desnu orbitu i prednji dio kavernoznog sinusa te u sfenoidni sinus
Figure 4 The MSCT of the head and neck. The palatal lesion extends to the soft tissue of the cheek, into the right orbit and into the anterior part of the cavernous sinus as well as into the sphenoid sinus.

Slika 5. Smanjenje tumora nakon liječenja zračenjem
Figure 5 Reduction of the tumor after radiotherapy.

carcinoma has a slow and often insidious growth pattern and most patients present at advanced stages of the disease. Based on tumor location, patients might have different signs and symptoms. Pain is a common clinical finding due to an early perineural invasion of the neoplastic cells (7). Adenoid cystic carcinoma develops frequently in minor salivary glands in comparison to the sublingual and parotid glands. Sometimes it starts as a jaw lesion (2).

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It is a well known fact that the pain arising from non-dental causes might be confused as pain arising from the tooth, which leads to misdiagnosis. Park et al., reported an adenoid cystic carcinoma of maxillary sinus misdiagnosed as chronic apical periodontitis (8). Therefore, pain associated to salivary gland malignant tumor may be misdiagnosed as pain from odontogenic origin.

In a recent study of 4004 periapical lesions which were diagnosed as endodontically associated pathoses, nine were found to be malignant processes, seven of which were squamous cell carcinoma, one Langerhans cell histiocytosis and one adenoid cystic carcinoma located in the mandible (9).

Perineural spread of the head and neck cancer was studied in a recent review (10). The results have shown that different types of head and neck tumors, including adenoid cystic carcinoma, have a high tendency to spread perineurally. This is especially true for tumors arising in the maxillary sinus, as they can spread to adjacent structures and cause pain in the face and orbit.

Therefore, it is crucial to consider the possibility of a neoplastic cause when a patient presents with persistent pain in the absence of a clear dental cause. Early diagnosis and appropriate treatment are essential to improve outcomes for patients with adenoid cystic carcinoma.

In conclusion, adenoid cystic carcinoma is a rare but aggressive tumor that can present with perineural pain. It is important for clinicians to have a high index of suspicion for this diagnosis in patients presenting with persistent pain, especially in the absence of a clear dental cause. Early diagnosis and treatment are crucial to improve outcomes for these patients.

References:
1. Park et al., reported an adenoid cystic carcinoma of maxillary sinus misdiagnosed as chronic apical periodontitis (8).
2. Dobro is poznato da se bol koja potječe od nedentalnog uzorka može zamijeniti s boli koja potječe od zuba, što rezultira pogrešnom dijagnozom. Park et al., reported an adenoid cystic carcinoma of maxillary sinus misdiagnosed as chronic apical periodontitis (8).
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Sažetak
Adenoidni cistični karcinom rjeđe je dijagnosticirani tumor koji može zahvatiti velike i male žlijezde slinovnice. Prikazan je slučaj 70-godišnjeg bolesnika koji je upućen u Zavod za oralnu medicinu Stomatološkog fakulteta u Zagrebu zbog boli u desnoj strani gornje čeljusti. U ovom slučaju opisujemo kako jednostrana oštra bol u gornjoj čeljusti i oku može ukazivati na dijagnozu adenoidnog cističnog karcinoma, iako bolesnik nema u usnoj šupljinu vidljivih promjena.

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Ključne riječi
adenoidni cistični karcinom; tumori žlijezda slinovnice; jednostrana bol gornje čeljusti i oka

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