Hyalinizing Clear Cell Adenocarcinoma of the Hard Palate: Case Report

Sert Damağın Hiyalinize Berrak Hücreli Adenokarsinomu

İlker AKYILDIZ,[a]
Zeynep KAPTAN,[a]
Hatice KARADAŞ,[a]
Ethem Erdal SAMİM,[a]
Melahat DÖNMEZ[b]

Clinics of
[a]Otorhinolaryngology Head and Neck Surgery,
[b]Pathology
Ankara Training and Research Hospital, Ankara

Geliş Tarihi/*Received:* 31.01.2014
Kabul Tarihi/*Accepted:* 04.09.2014

Yazışma Adresi/*Correspondence:*
İlker AKYILDIZ
Ankara Training and Research Hospital, Clinic of Otorhinolaryngology Head and Neck Surgery, Ankara, TÜRKİYE/TURKEY
ilkerakyildiz@yahoo.com

ABSTRACT Hyalinizing clear cell adenocarcinoma is a rare tumor originating from minor salivary glands. 48 years old male patient presented with painless swelling on the hard palate. Computerized tomography and magnetic resonance imagination of the maxilla revealed 3 cm by 3 cm sized, heterogenous gadolinium enhancing mass which was originating from the incisive theet gingiva and extending to the bilateral maxillar arch intraorally and to the palatin bone in the nasal cavity. Incisional biopsy report of the mass was hyalinizing adenocarcinoma of the hard palate. In the light of the biopsy result and radiologic examination wide surgical resection and bilateral supraomohyoid neck dissection was performed. Due to low mitotic activity and nuclear pleomorphism it is defined as low grade tumor according to classic adenocarcinoma leading to better prognosis if treated promptly. Given the rarity and lack of reported series, diagnosis and tretament of this tumor was discussed in the light of the literature.

Key Words: Maxillary neoplasms; adenocarcinoma, clear cell

ÖZET Hiyalinize berrak hücreli adenokarsinom minör tükürük bezlerinden kaynaklanan nadir bir tümördür. 48 yaşında erkek hasta sert damağında ağrısız şişlik nedeni ile başvurdu. Maksillanın bilgisayarlı tomografi ve manyetik rezonans ile görüntülemelerinde 3x3 cm boyutunda heterojen gadolinyum tutan, kesici dişlerin bulunduğu gingivadan başlayarak bilateral maksiller arka doğru uzanan, nazal kavite içinde de palatin kemiğe doğru uzanan kitle saptandı. Kitlenin insizyonel biyopsi sonucu hiyalinize berrak hücreli adenokarsinom olarak raporlandı. Biyopsi sonucu ve radyolojik inceleme ışığında geniş cerrahi eksizyon ve bilateral supraomohyoid boyun diseksiyonu uygulandı. Hiyalinize berrak hücreli adenokarsinom düşük mitotik aktivite ve nükleer pleomorfizmi nedeni ile uygun tanı ve tedavisi yapıldığı taktirde klasik adenokarsinoma göre düşük dereceli ve iyi prognozlu bir tümör olarak tanımlanmaktadır. Nadirliği ve yayınlanmış az sayıda serisi olması nedeniyle tanı ve tedavisi literatür eşliğinde tartışılmıştır.

Anahtar Kelimeler: Maksiller tümörler; adenokarsinom, berrak hücreli

Turkiye Klinikleri J Case Rep 2015;23(3):335-8

Hyalinizing clear cell carcinoma (HCCC) is a rare malignant tumor originating from salivary glands and described as a distinct entity from other clear cell salivary gland tumors by Milchgrub et al. in 1994.[1] Clinic behavior of HCCC is not well understood given the rarity of the tumor and the lack of analysis of reported series. It presents as a slow growing sub-mucosal mass without surface ulceration.[2]

doi: 10.5336/caserep.2014-39034

In this paper we present a patient who has HCCC originating from hard palate because of its rarity and to discuss the clinic behavior, surgical management and review of the literature of this uncommon clinic entity.

CASE REPORT

A 48-years-old male patient presented with painless swelling on the front and middle part of the hard palate by a duration of 2 months (Figure 1). There was no history of bleeding and tumor elsewhere in the body. The patient was a heavy smoker by 25 package/year and no alcohol consumption. Physical examination revealed a mass which was 3x3 cm in size, moderate ulcerated and minimally hemoragic. When palpated the mass was fragile, minimally hemoragic, attached to the underlying gingiva and periost tightly. On the physical examination the mass was originating from the gingiva of the incisiva theet and extending bilaterally to the alveolar arch of the maxilla about 2 cm posteriorly. Right maxillary canine, right 1. and 2. premolar, right 3. molar theet were viable. All of the other theet were lost due to decays and tumor invading.

Computed Tomography (CT) scanning of the head and neck revealed a mass of 3 cm by 3 cm in size extending to the alveolar arch intraorally and horizontal plate of palatin bone intranasally (Figure 2). Magnetic Resonance Imagining (MRI) of the maxilla revealed a mass of 3 cm by 3 cm in size which was originating from the anterior part of the maxilla and extending to the palatin bone (Figure 3). MRI showed late heterogenous gadolinium enhancement.

18F-FDG whole body PET/CT sacanning showed an increased 18F FDG uptake with a 12.27 SUVmax value on the anterior part of the maxilla and with a 3.55 SUVmax value on bilateral submandibular region. 12.27 SUVmax value and the appaerance on the CT scanning on the maxilla raise a concern of malignancy.

Pathologic evaluation of the specimen which was acquired by an incisional biopsy from the anterior side of the hard palate was reported as HCCC of the maxilla.

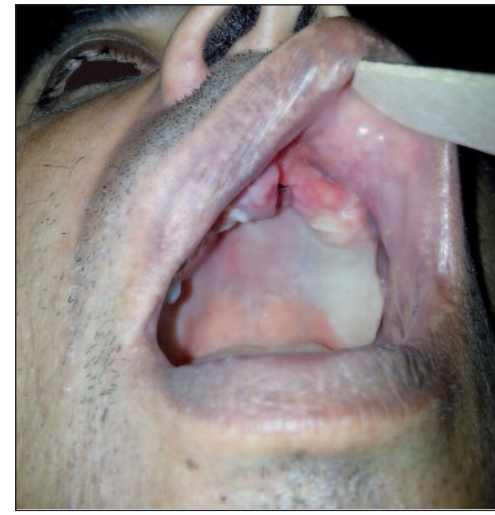

FIGURE 1: Ulcerative mass located to the gingiva.

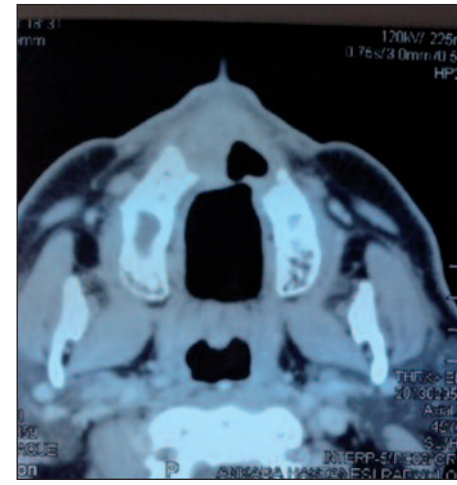

FIGURE 2: Computerized Tomography scanning of the maxilla note the erosion at the front site of the maxilla.

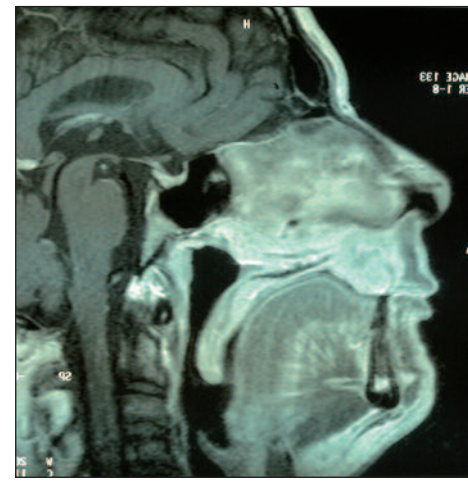

FIGURE 3: T1-weighted Magnetic Resonance imaging of the maxilla.

In the light of these radiologic, pathologic and clinic information wide surgical excision (bilateral inferior maxillectomy) and bilateral zone 1-2-3 elective neck dissection was planned. We obtained an informed consent from the patient including knowledge about the status of the disease, surgical intervention and additive therapies.

In this case we performed a bilateral inferior maxillectomy and bilateral zone 1, 2, 3 elective neck dissection. Resection margins were decided to obtain approximately 1 cm safety zone. Histopathologic evaluation of intraoperative frozen sections of the surgical margins and bilateral zone 2 lymph nodes were tumor free (Figure 4, 5). Elective neck dissection was not mandatory but we performed neck dissection because of the presence of submandibular 18F-FDG uptake with a SUVmax value of 3.55. Post operative adjuvant radiotherapy was planned due to perineural invasion of the post operative histopathologic evaluation of the maxillectomy specimen.

Histopathologic evaluation of the maxillectomy specimen was reported as hyalinizing clear cell carcinoma of the hard palate (Figure 6, 7).

Postoperative radiotherapy was performed to the patient due to perineural invasion of the tumor.

DISCUSSION

Clear cell carcinoma of the head and neck may be seen in a wide variety tumors such as mixed tumors, salivary gland tumors, myoepitheliomas, oncocytomas, mucoepidermoid tumors, adenoid cystic carcinomas, clear cell carcinomas metastizing

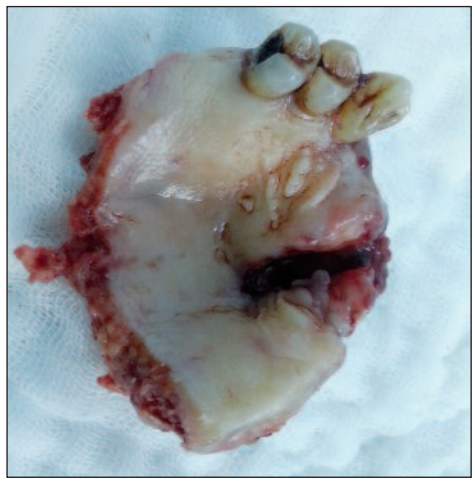

FIGURE 4: Inferior maxillectomy specimen.

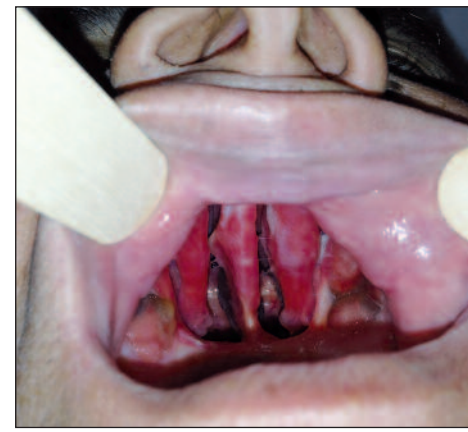

FIGURE 5: Post operative 3 months of the patient.

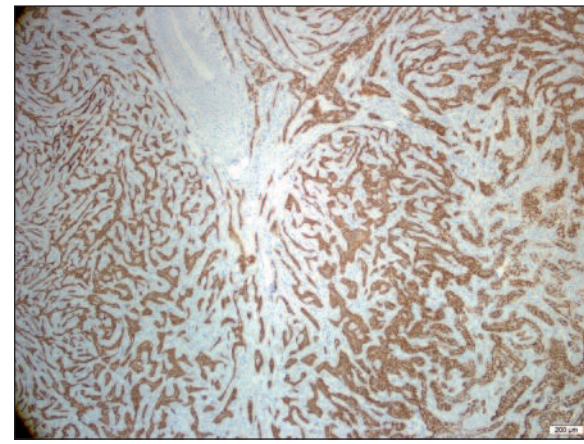

FIGURE 6: Neoplastic epithelial cells are positive with CK14 antibody in immunohistochemical study (X200).

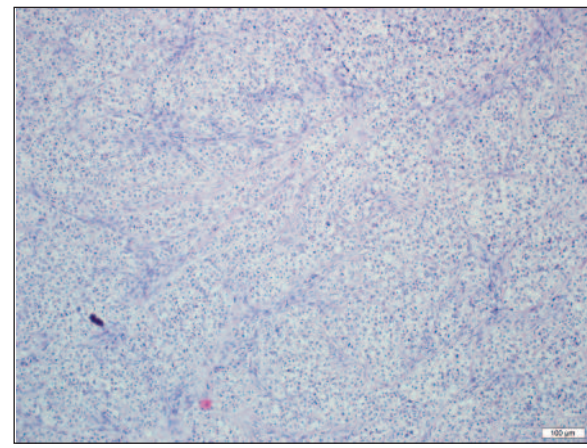

FIGURE 7: Hyalinizing connective tissue at the ground of the tumor is positive but neoplastic epithelial cells are negative with SMA antibody (X200).

from the kidney.[3] Hyalinizing form of the clear cell carcinoma is a distinct entity which was described in 1994 by Milchgrub.[1] This rare malignancy consist of less then 1% of all salivary gland tumors.[4] HCCC is mostly seen in female population and has a tendency to be seen in the older ages. In the oral cavity HCCC originates from minor salivary glands and the most seen region is tongue, hard palate, floor of the mouth, buccal mucosa, retromolar trigone and alveolar arch. Parotid gland, nasopharynx, hypopharynx and lacrimal gland are the seldom places where HCCC may also arise.[5]

Hyalinizing form of clear cell carcinoma arising from minor salivary glands of the oral cavity is new forms described. This form consist of individual tumor cells and infiltrative borders but has minimal nuclear pleomorphism and low mitotic activity microscopically.[6] This low mitotic activity and nuclear pleomorphism is regarded that hyalinizing form of clear cell carcinoma is a low grade and curable tumor if treated appropriately. So that wide surgical excision of the pimary tumor and with or without radiotherapy is accepted as a treatment modality in spite of its limited reported experience.[1,7] Performing neck dissection depands on positive surgical margins, high grade histology, perineural/perivascular invasion and positive palpabl nodes on the neck examination.[8] Reconstruction of the surgical defect may be done by microvascular radial forearm fascioucutaneous free-flap or basic hard palate prosthesis.[6]

Regarding the rarity of HCCC-reported only 34 cases in the English literature,[1-9] and appearing of the clear cells in most of the head and neck tumors, the differential diagnosis of HCCC should be made accurately by the use of clinic, radiologic, histopathologic and immunohistochemical studies.

Scanning of the maxilla, neck and the whole body was done by the use of CT, MRI and 18F-FDG whole body PET/CT studies in this case. Histopathologic and immunohistochemical staining studies were used for the specimen which was obtained by an incisional biopsy from hard palate for accurate diagnosis. CK 14, P 63 were positive and the proliferation index was 20% with Ki 67.

Immunohistochemical profile should be studied to obtain an appropriate differantial diagnosis. Metastasis of renal cell carcinoma and sarcoidosis are the two diseases that may be seen in the oral cavity so these two diagnosis should be ruled out.[3]

Maxillectomy defect was reconstructed with a basic hard palate prosthesis.

REFERENCES

1. Milchgrub S, Gnepp DR, Vuitch F, Delgado R, Albores-Saavedra J. Hyalinizing clear cell carcinoma of salivary gland. Am J Surg Pathol 1994;18(1):74-82.
2. Baghirath PV, Kumar JV, Vinay BH. Hyalinizing clear cell carcinoma: A rare entity. J Oral Maxillofac Pathol 2011;15(3):335-9.
3. Angiero F, Stefani M. Hyalinizing clear cell carcinoma arising on the anterior palatoglossal arch. Anticancer Res 2007;27(6C):4271-7.
4. Balakrishnan R, Nayak DR, Pillai S, Rao L. Hyalinizing clear cell carcinoma of the base of the tongue. J Laryngol Otol 2002;116(10):851-3.
5. O'Sullivan-Mejia ED, Massey HD, Faquin WC, Powers CN. Hyalinizing clear cell carcinoma: report of eight cases and a review of literature. Head Neck Pathol 2009;3(3):179-85.
6. Saleh KA, Nurishmah MI, Firouzeh GN, Goh BS. Primary clear cell carcinoma of minor salivary gland of the soft palate: a case report. Med J Malaysia 2012;67(3):335-6.
7. Ereño C, Grande J, Alija V, Yarnoz J, Bilbao FJ. Hyalinizing clear cell carcinoma of the hypopharynx metastasizing to the lung: a case report. Histopathology 2000;37(1):89-91.
8. Vander Poorten VL, Balm AJ, Hilgers FJ, Tan IB, Keus RB, Hart AA. Stage as major long term outcome predictor in minor salivary gland carcinoma. Cancer 2000;89(6):1195-204.
9. Suzuki H, Yamauchi G, Hashimoto K. Clear cell carcinoma of the mandibular gingiva 'minor salivary gland': a case report with immunohistochemical study. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2007;103(3): e36-40.