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HYALINIZING CLEAR CELL CARCINOMA OF CHEEK - A RARE DIAGNOSIS

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ABSTRACT

Hyalinizing clear cell carcinoma (HCCC) is an uncommon malignant salivary gland tumor accounting for about 1% of all intra-oral salivary gland tumors. Microscopic diagnosis of clear cell carcinoma may be challenging because of the spectrum of features which frequently overlaps with the other salivary gland tumors that contain clear cells, and thus it may be a diagnosis of exclusion. HCCC is currently classified as a "clear cell adenocarcinoma" by the AFIP and as "clear cell carcinoma, not otherwise specified (NOS)" by the World Health Organization (WHO). Here we, report a case of HCCC in a 55 years old male with detailed histopathological, clinical and management review.

KEYWORD

Hyalinizing Clear Cell Carcinoma of check - A rare diagnosis.

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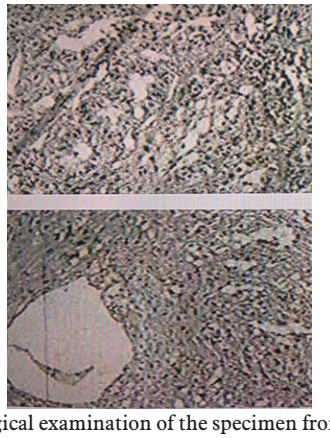
Background -Hyalinizing clear cell carcinoma (HCCC) was first described by Milchgrub et al. in 1994 as a rare minor salivary gland carcinoma made up of clear cells forming cords and nests in a hyalinized stroma [1] Since then, 141 cases have been reported in the English literature (up to 2016). HCCC shows a female predominance (72.7%) and older age at diagnosis (68 years mean), with the vast majority of cases (81.8%) occurring in the oral cavity, most commonly the tongue and hard palate, and less common locations include the parotid gland, nasopharynx, hypopharynx and orbit/lacrimal gland.[2] Microscopically the diagnosis is challenging because the cohort of clear cell salivary gland neoplasms is heterogeneous.

CASE REPORT - A 55 years old male patient reported to mahatma Gandhi medical college and hospital, Jaipur in oct. 2018, with a painless swelling with ulceration over right check, which gradually increased in size over a period of 6 months. Clinical examination revealed a mass measuring of about 4 × 4 cm in size over mid face with central ulceration. [Figure 1].

On palpation, the mass was tender, smooth, firm, and movable over underlying maxilla. Central ulcer of size 3 × 3 cm with sharp margins, punched out edges and necrotic indurated base. Radiographic computed tomographic examination suggest heterogeneously enhancing lobulated ulcerated mass measuring 20X25 mm seen involving skin and subcutaneous soft tissues in left premaxillary region. No evident infiltration into adjacent maxilla seen. Nasopharynx, oropharynx, hypopharynx, glottis and subglottic larynx appears normal. Bilateral major neck vessels and salivary glands were normal. No significant cervical lymphadenopathy is seen. Multiple well defined nodular lesions seen in visualized right lung upper lobe s/o metastasis.

The lesion was clinically diagnosed as basal cell crcinoma, and incisional biopsy was performed. The routine processing and hematoxylin and eosin (H and E) staining findings showing squamous mucosa infiltrated by a tumour composed of nests, infiltrating cords or single file clear cell consisting of glycogen with moderate atypia. The nests and cords were separated by hyalinised stroma. There was stromal desmoplasia and associated inflammation. Overall histomorphology favouring hyalinizing clear cell carcinoma.



Histopathological examination of the specimen from the lesion of the patient in the case report revealing islands of of epithelial cells and clear cells arranged in cords or nests in the connective tissue (H and E stain)

DISCUSSION - Clear cells create diagnostic dilemmas and controversies in the classification of clear cell salivary gland neoplasms, and hence correct classification is a challenge in their presence. It is important to differentiate HCCC from other tumors with clear cell features because of their differences in treatment and clinical outcome.

HCCC usually arises in patients in their 60s or older; however, a broad age range is noted.[3] The most common sites of origin are the palate and tongue base, followed by the mobile tongue and

other minor salivary gland locations. These include the lip, retromolar trigone, buccal mucosa, floor of mouth and gingiva. These latter cases may sometimes invade bone and the distinction between a mucosal and primary bone tumor may be difficult, especially since "central" examples of HCCC have been described [4] The tumor size ranges between 1.0 and 4.5 cm (mean 2.0–3.0 cm) and has a white-tan cut surface grossly. [5] HCCCs may appear relatively circumscribed but lack encapsulation and they have a tendency to show infiltration of the surrounding tissues, despite this grossly circumscribed appearance.

Histologically, there is a relatively wide range of appearance of HCCC tumor cells. The predominance of clear cells is seen in a minority of cases and the tumor cells actually often have pale eosinophilic cytoplasm rather than clear cytoplasm, or they may have a mixture of both.[1]. The clear cells show granular PAS positivity and diastase sensitivity, indicative of glycogen.

Grading, Treatment and Prognosis - There is no formal grading system and HCCC is considered low-grade by definition [3]. The vast majority of HCCC have had good outcome. Only one documented mortality due to disease has been described [6], although an additional "palliative" case has been reported [7]. The tumors have a propensity for locoregional recurrence in the order of 12–17 % [8] and occasional lymph node [1] or distant metastases. [8]. have been noted. Treatment usually involves primary resection if the tumor arises in an amenable location. Tongue base tumors may have primary radiation due to morbidity of resection. Many cases have post operative radiotherapy; however, there is no standard of care for these tumors as they are rare and respond well in most instances irrespective of treatment.

Conclusions - HCCC should be considered in the differential diagnosis of tumors with a histology marked by monomorphic clear cells arranged in cords, trabecular or in clusters, and surrounded by hyalinized stroma. Special stains and IHC tests help in diagnosing this type of carcinomas. Hence HCCC is a diagnosis of exclusion. HCCC has an excellent overall behavior although a growing number of cases are showing high-grade features [9] or unusual behavior. There is no rationale at this time for a grading system.

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. O'Sullivan-Mejia ED, Massey HD, Faquin WC, Powers CN. Hyalinizing clear cell carcinoma: report of eight cases and review of literature. Head Neck Pathol. 2009;3:179–85.
3. Solar AA, Schmidt BL, Jordan RC. Hyalinizing clear cell carcinoma: case series and comprehensive review of the literature. Cancer. 2009;115(1):75–83.
4. Manoharan M, Othman NH, Samsudin AR. Hyalinizing clear cell carcinoma of minor salivary gland: case report. Braz Dent J. 2002;12(1):66–9.
5. Okón K, Modrzejewski M, Strék P. Hyalinizing clear cell carcinoma of the salivary gland. A case report. Pol J Pathol. 1996;47(1):33–6.
6. Maiorano E, Altini M, Favia G. Clear cell tumors of the salivary glands, jaws, and oral mucosa. Semin Diagn Pathol. 1997;14(3):201–12.
7. Solar AA, Schmidt BL, Jordan RC. Hyalinizing clear cell carcinoma: case series and comprehensive review of the literature. Cancer. 2009;115(1):75–83
8. O'Regan E, Shandilya M, Gnepp DR, et al. Hyalinizing clear cell carcinoma of salivary gland: an aggressive variant. Oral Oncol. 2004;40(3):348–52.
9. Weinreb I, Seethala RR, Perez-Ordoñez B, et al. Oncocytic mucoepidermoid carcinoma: clinicopathologic description in a series of 12 cases. Am J Surgical Pathol. 2009;33(3):410–6.