
CASE REPORT**Clear Cell Variant of Mucoepidermoid Carcinoma-Maxillary Region:
A Case Report**

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Abstract:

The clear cell variant of mucoepidermoid carcinoma (MEC) has been described in the major and minor salivary glands, oral cavity, skin and the bronchus. We report a case of clear cell variant of MEC of maxillary region in a 60 year old male who presented with a huge, disfiguring, lobulated maxillary mass. To our knowledge, this variant has not been previously reported in the maxillary region.

Keywords: Clear Cells, Mucoepidermoid Carcinoma, Maxillary Region

Introduction:

Mucoepidermoid carcinoma (MEC) was first described by Masson and Berger in 1924. These are low grade tumors constituting 5-13% of salivary gland neoplasms [1-6]. They have been described at various atypical sites including nasal cavity, paranasal sinuses, respiratory tract, breast, and skin [2]. MEC is histologically characterised by an admixture of varying proportion of intermediate cells, epidermoid cells and mucocytes. Clear cells, oncocyctic cells and spindle cells may be seen focally. When they predominate in large areas they are termed as clear cell, oncocyctic and spindle cell variants respectively.

Case Report:

A 60 year old male presented to ophthalmology department with a painful swelling below his right eye for the previous 2-3 months. On examination, the infraorbital swelling was tender, fixed, nonpulsatile and extended over the right maxillary region. Computed Tomography of Paranasal Sinuses (CT PNS) coronal/axial study showed a large, heterogenous contrast enhancing solid mass measuring 5.5x4cm arising in the right maxillary region extending into the infraorbital region, right frontal sinus, posterior infratemporal region and obliterating right ethmoidal sinus. The nasal septum showed no deviation. With the CT findings suggestive of carcinoma right maxilla, he underwent right maxillectomy with orbital exentration. We received right maxillectomy specimen with orbital exentration which showed a maxillary nodular mass with a bosselated appearance covered by unremarkable skin (Fig1). The cut section showed a grey white tumour measuring 7.2x6x5cm with gross involvement of medial surgical margin and erosion of orbital plate and underlying maxilla. Histologic examination showed tissue lined by pseudostratified columnar epithelium with underlying tumour showing

intricate admixture of three types of cells-epidermoid cells, intermediate cells and mucinous cells (Fig 2).



Fig 1: Gross Photograph Showing Eyeball and a Large Multilobulated Maxillary Mass

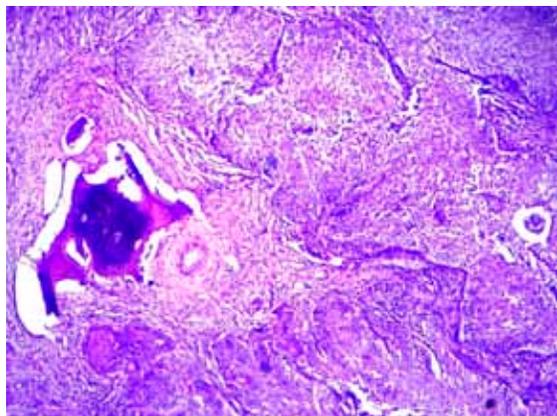


Fig 2: Photomicrograph Showing Bone Infiltrated by a Tumor Composed of Admixture of Squamous Component and Intermediate Cells with Central Cells Showing Clear Cell Predominance (H and E, X40)

The epidermoid component was composed of polygonal cells with hyperchromatic nuclei and abundant eosinophilic cytoplasm arranged in

sheets, pseudoalveolar and papillary pattern. Occasional pearl formation was noted (Fig 3).

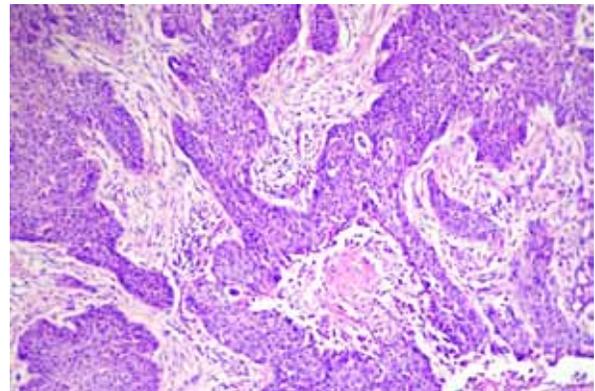


Fig.3: Epidermoid Component (H and E, x100)

Intermediate cells with mild nuclear atypia, moderate amount of eosinophilic cytoplasm were arranged in sheets with a predominant clear cell component showing large polygonal cells with nuclear atypia and moderate amount of clear cytoplasm (Fig 4).

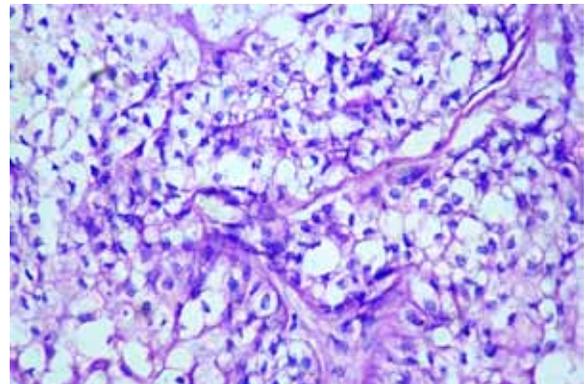


Fig 4: Clear Cells with Centrally Placed Round Nuclei, Marked Nuclear Atypia and Abundant Clear Cytoplasm (H and E, X400)

Admixed mucinous component showed mucocytes lying in clusters (Fig 5) and cystic spaces lined by mucin secreting tall columnar cells. Six to seven mitotic figures/10 high power fields were seen.

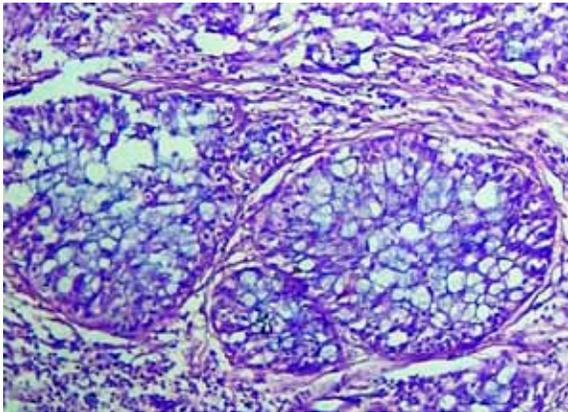


Fig. 5: Photomicrograph Showing Mucocytes with Central/Eccentrically Placed Nuclei and Intracytoplasmic Mucin (H and E, X400)

Comedonecrosis, perineural invasion (Fig 6) and vascular emboli were evident along with skeletal muscle and bone infiltration. A histological diagnosis of high grade mucoepidermoid carcinoma-clear cell variant with involvement of medial, posterior, superior surgical margins and infiltration of maxilla and orbital bony plate was rendered.

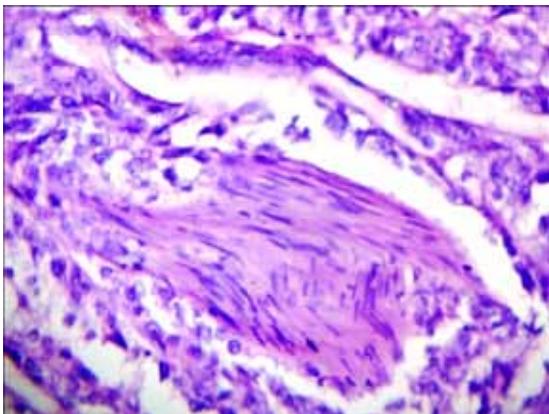


Fig 6- Perineural Invasion by Tumor Cells (H and E, X400)

Discussion:

Clear cell variant of MEC does not vary in clinical setting from conventional MEC and shows a similar female predominance (3:2), the mean age of onset is 5th decade of life. They usually form a painful mass of varying duration and invade the underlying bone. Our case presented at the age of 60 years with a huge maxillary swelling with destruction of maxilla and the infraorbital plate. Histologically, the MECs are characterised by frequent admixture of intermediate cells which are small and basophilic, mucocytes which occur singly or in clusters with pale cytoplasm, a distinct cell boundary and small, peripherally placed nuclei and focally distributed epidermoid cells which may show keratin pearl formation [2-6]. The abundance of clear cells in our case made it a diagnostic challenge. Clear cells in mucoepidermoid carcinoma of the salivary glands may constitute up to 10% of the cell population [5]. The differential diagnosis of salivary gland tumors with a dominant population of clear cells include clear cell myoepithelioma, acinic cell carcinoma, clear cell adenocarcinoma, and metastatic renal cell carcinoma [6]. It was only through identification of mucoid and epidermoid cells and clear cells with PAS positive diastase sensitive material that a diagnosis of mucoepidermoid carcinoma-clear cell variant was reached in our patient. MEC arising and confined to bone commonly affecting mandible are termed as central MEC.

In our case we could see normal salivary gland with tumor infiltrating the bone and soft tissue

around it without confinement to bone. Several microscopic grading systems based on a numerical score have been advocated as means of predicting the outcome. MEC are classified histologically as low, intermediate, and high-grade types depending on the morphologic characteristics, presence of the cellular atypia, number of mitotic figures, nuclear pleomorphism, perineural invasion, necrosis, and its invasive characteristics [1-7]. Low-grade tumors are well differentiated without atypia and are predominantly made up of intermediate and mucus cells. High-grade tumors are poorly differentiated, show predominance of epidermoid component with evidence of cytologic atypia, high mitotic activity, areas of necrosis and neural invasion. The histologic features of intermediate-grade tumors fall in between the two. Dulguerov *et al* [1] studied 220 cases and found correlation between the grade and prognosis. The survival ranged from 0 to 43% for patients with high-grade MEC, 62 to 92% for patients with intermediate-grade tumors, and 92 to 100% for patients with

low-grade tumors. The presence of clear cells does not seem to relate to its prognosis but it has been noted that clear cells usually predominate in high-grade tumors [3, 4, 6]. Our index case was of high grade with a dominant clear cell population and with invasion of maxillary and infraorbital bone. The pathogenesis of the clear cells has been debated. It has been proposed that they may be a result of hydropic degeneration of the epidermoid cells, but may also result from fixation artifacts or the presence of lipidells [6].

Conclusion:

This case highlights the histologic findings of an unusual variant of mucoepidermoid carcinoma which is termed as high-grade, due to its morphologic characteristics, predominance of clear cells and nature of invasion. Other tumours with clear cells come under differential diagnosis. Thorough sampling for search of mucocytes and epidermoid component helps to diagnose this variant of MEC.

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