Mucoepidermoid Carcinoma Of The Lacrimal Sac - Case Report

Maria Dref 1, Ayman Ismail1, Imane Boujguena1, Rachid Oukassou 2, Ibtissam Hajji 2, Abdeljalile Moutaouakil 2, Hanane Rais1

1Department of Pathology, FMPM-UCA, CHU Mohammed VI, Marrakech, Morocco
2Department of Ophthalmology, FMPM-UCAM CHU Mohammed VI, Marrakech, Morocco

ARTICLE INFO

Mucoepidermoid carcinoma (MEC) is a malignant epithelial neoplasm containing both mucous and epidermoid cells (WHO2018). It is the most common primary carcinoma of the major salivary glands, but has also been noted rarely in other sites. We report an unusual clinical case of a 14-year-old with Mucoepidermoid carcinoma of the lacrimal gland with orbit invasion, diagnosed at the department of Pathology CHU Mohammed VI Marrakech. The diagnosis is made initially on a biopsy and then on a piece of surgical exenteration.
INTRODUCTION:
Mucoepidermoid carcinoma (MEC) is a malignant epithelial neoplasm containing both mucous and epidermoid cells (WHO2018). It is the most common primary carcinoma of the major salivary glands, but has also been noted rarely in other sites like respiratory tract [1,2]. It is extremely uncommon in the ocular region, where it can arise in the lacrimal gland, lacrimal sac (LS) or conjunctiva [3]. Lacrimal sac and conjunctival MECs have been reported to invade the eye or orbit, and enucleation or exenteration may be necessary. We report the case of a patient diagnosed with MEC of the lacrimal gland with orbit invasion.

CASE REPORT:
Our female patient is 14 years old. She had a rapidly progressive orbital tumor of the left superolateral angle. A biopsy was performed showed an epidermoid carcinoma with a mucinous contingent. The patient underwent left orbital exenteration. The macroscopic examination showed a budding neoplasm of 3X3x2.5 cm originating at the outer corner of the eye that infiltrates the entire eyeball associated to an extensive cutaneous ulceration over 2X2cm at the inner corner of the eye. Multiple sections showed that the neoplasm was white, firm, the seat of hemorrhagic changes. Hematoxylin-eosin–stained tissue sections revealed an invasive high-grade tumoral proliferation. The tumour was composed of varying proportions of atypical Squamous cells, glandular cells, and intermediate cells forming cords, sheets and clusters. The carcinomatous cells were contiguous of medium to large size, ballooned in places. The nuclei were anisokaryotic, with vesicular chromatin and a high cyto-nuclear ratio. These cells were strongly nucleated with numerous mitoses estimated at 40 mitoses per 10 high power fields. Cytoplasm was abundant eosinophilic vacuolized by location. Tumour stroma was desmoplastic and the mucinous contingent accounted less than 10 percent of tumor proliferation. This proliferation infiltrates the entire eyeball and dissociates the periorbital muscle. The immunohistochemistry study showed that tumour cells were labelled with anti-p63 , anti-CK7 , anti-CK5/6 and ki67 antibodies. Our final diagnosis was a high-grade infiltrating mucoepidermal carcinoma.

![Figure 1: HE x 40 MEC: Squamous cells, glandular cells, and intermediate cells](image)
Figure 2: HEx 200 MEC: Little differentiated glandular component

Figure 3: P63 x 200 MEC: Squamous component

Figure 4: Ck7 x 200 MEC glandular component
DISCUSSION:
The lacrimal gland is a bilobed eccrine secretory gland located in the superotemporal orbit. Most of the neoplasms of the lacrimal gland are originated from epithelial tissues, of which 55% are classified as benign and 45% as malignant [2]. Mucoepidermoid carcinoma accounts for only 3.6% of all malignant epithelial lacrimal gland tumours [mucoepider lacr gland]. MEC generally arises in major salivary glands and minor salivary glands of the oral cavity and the respiratory tract. It is acknowledged to occasionally occur from the minor and major lacrimal glands of the orbit and in the nasolacrimal system.

Three basic cellular elements that compose mucoepidermoid carcinoma as it’s revealed by histology: mucous-secreting, epidermoid-Squamous and intermediate cells. Moreover, there may be clear, admixed columnar and occasionally oncocytic cells. The use of special stains makes the Intracytoplasmic mucin obvious in some tumors, such as mucicarmine or Alcian blue. Mucosal cells, regardless of their form, constitute <10% of most mucoepidermal carcinomas. When a tumor contains both neoplastic mucin producing and epidermoid cells it claimed to be a Mucoepidermoid carcinoma (4).Mucoepidermoid carcinoma of the LS is in fact very rare. Ni et al (5) postulate that it may arise from either the LS wall serous gland epithelium or the columnar epithelium of the conjunctiva, with its globet cells. The first case of MEC arising in the LS was described in Brazil in 1981 by Bambirra et al(6) An LS biopsy is the most important tool (7), biopsies are performed either to establish the diagnosis of a suspicious mass or to identify an abnormal lesion seen in the LS mucosa during DCR(8).To diagnose MEC, the coexistence of 3 types of cells is highly required :epidermoid, intermediate and mucin secreting cells.(9) Hence the importance of properly sampling the tumor in order to find the different cell quotas.

MECs are ranked histologically on the grounds of the prevalence of mucosal cells: low, intermediate and high. Low-grade tumors are clearly differentiated and made up of over 50% of mucus-secreting elements and Squamous epithelial cells. High-grade tumours are poorly differentiated and are composed mainly of Squamous epithelial and intermediate cells, containing less than 10% mucus-secreting cells. The histological characteristics of intermediate-grade tumours lie between those of low-grade and high-grade tumours.

Histological classification of MEC is the most predictive prognosis, it can be used to formulate a therapeutic plan ( 2).

Multi-planar CT imaging of the paranasal sinuses and orbit it is vital to define the extent of the tumor and to find evidence of bony destruction of the lacrimal fossa(10).Magnetic resonance imaging is useful for distinguishing the lesion form adjacent retained secretions, hypertrophic inflammatory mucosa and adipose tissues(11). The treatment of LS tumors is often multidisciplinary and modalities can include surgery, chemotherapy, immunotherapy, and radiotherapy, depending of the histologic type and anatomic extent of the tumor (8). Most patients
underwent exenteration with postoperative radiation therapy when the soft tissue margins were positive for tumor infiltration.(6,7).

CONCLUSION
MEC of the lacrimal sac is a rare and locally aggressive tumor. Positive diagnosis is histological. Prognosis and treatment depend on histological grade and degree of tumor extension.

REFERENCES
4. JoAnna D. Williams, MD, Amit Agrawal, MD, and Paul E. Wakely, Mucoepidermoid Carcinoma of the Lacrimal Sac Annals of Diagnostic Pathology, Vol 7, No 1 (February), 2003
8. Sonia T. Brar and Dale Meyer. Diagnosis and Management of Mucoepidermoid Carcinoma of the Lacrimal Duct. Informa Healthcare USA, Inc. 2011
11. Lavleen Singh, Shuchita Singh1, Deepali Jain, Suresh C Sharma, Mucoepidermoid carcinoma of eyelid: A usual tumor at an unusual site Cancer journal. 2015.

How To Cite This Article:
Maria Dref, Ayman Ismail, Imane Boujguena, Rachid Oukassou, Ibtissam Hajji, Abdeljalile Moutaouakil, Hanane Raiss

Source of Support: Nil
Conflict of Interest: None declared

Your next submission with British BioMedicine Publishers will reach you the below assets
- Quality Editorial service
- Swift Peer Review
- E-prints Service
- Manuscript Podcast for convenient understanding
- Global attainment for your research
- Manuscript accessibility in different formats (Pdf, E-pub, Full Text)
- Unceasing customer service

Track the below URL for one-step submission
http://www.britishbiomedicine.com/manuscript-submission.aspx