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RESEARCH ARTICLE

PRIMARY ADENOCARCINOMA OF THE LACRIMAL GLAND: CASE REPORT

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Abstract

A patient aged of 43-years-old presented a left exophthalmia, was diagnosed with adenocarcinoma of the lacrimal gland. Loco regional assessment in the CT and MRI showed a tumor of 3 cm diameter in the superolateral angle of the left orbit infiltrating the left lateral rectus muscle. He had exenteration of the left eye. Given the positive margins, adjuvant radiotherapy was indicated. The radiotherapy was delivered using intensity modulated radiation therapy (IMRT) technique in order to overcome anatomical complexity of the region, cover the tumor bed and preserve the organs at risk.

Introduction:-

Primary adenocarcinoma of the lacrimal gland (PALG) is a rare malignant tumor that develops in the epithelial cells of the lacrimal gland. It represents only 5 to 7% of epithelial tumors of the lacrimal gland[1].

Given thesupratemporal location of the lacrimal gland, the most frequent clinical sign is exophthalmia. Computed tomography (CT) and magnetic resonance imaging (MRI) are very useful for loco regional extension but histology makes the diagnosis of certainty.Its histopathological classification is the same as the salivary glands due to its histological similarity with the salivary glands[2].

Treatment is based on multidisciplinary management but is not yet standardized given the rareness of the disease. Primary adenocarcinoma of the lacrimal gland can have a very aggressive character similar to salivary carcinoma with a poor prognosis[3].

We report a case of adenocarcinoma of the lacrimal gland with a review of the literature.

The case:

Our patient is a 43-years-old man, a chronic smoker for over 20 years with no pathological history.

The symptomatology goes back to nine months before his consultation in our department for a unilateral left exophthalmia non axial, non-pulsatile and painless, with progressive worseningand preserved visual acuity(figure 1).

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Figure 1:- Front picture of our patient showing exophthalmia of the left eye.

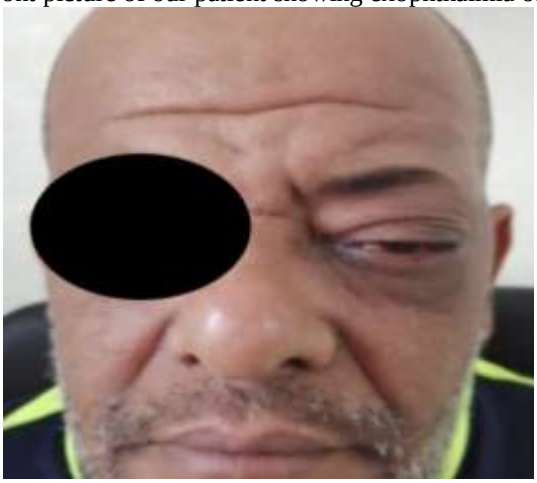


Figure 2:- Cerebral-orbital CT scan injected in axial section of our case, the arrow in red corresponds to the tumor.

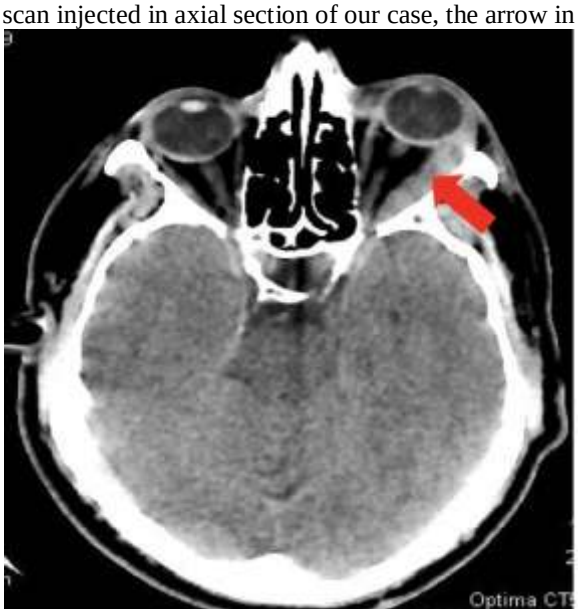


Figure 3:- Orbital MRI in axial and coronal section of our patient.

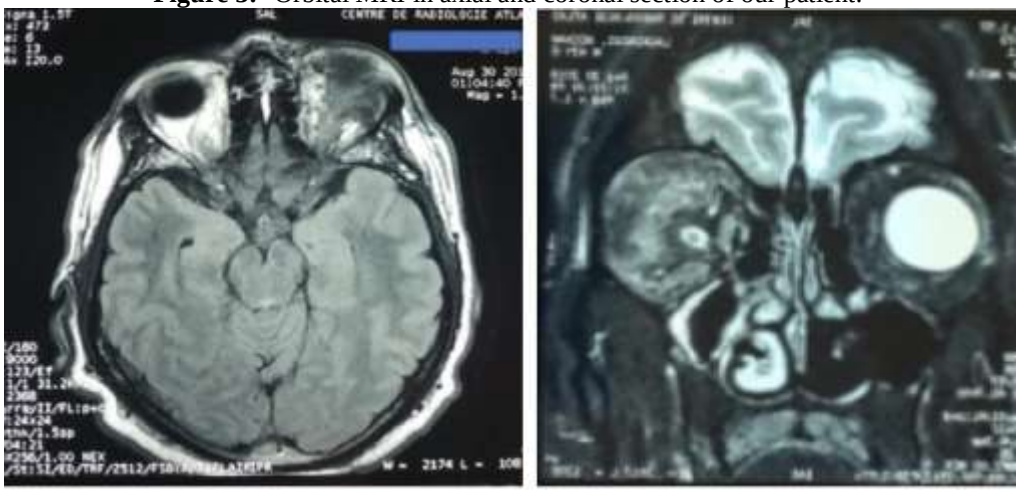
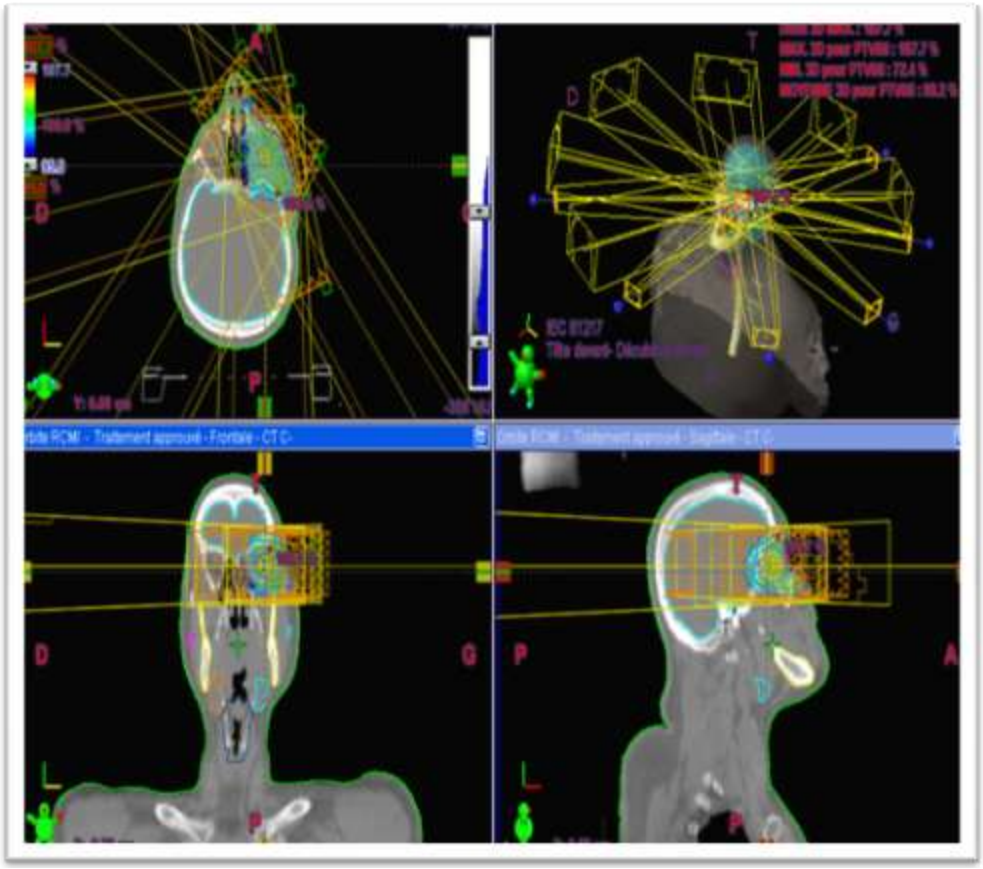


Figure 4:- Pathological images of the surgical specimen with the electronic microscope (x400) (A) and IHC (B).



Heconsulted in the ophthalmology department of the University Hospital of FES where an imaging was performed.

CT scan showed a hypodense tumor of 3 cm diameter in the superolateral angle of the left orbit infiltrating the left lateral rectus muscle responsible of an exophthalmia. the CT characteristics of thetumorare non-specific (figure 2).

Therefore, an MRI was performed, that showed a left intra-orbital inflammatory tumor of the lacrimal gland, suspecting a malignant tumor (figure 3).

A needle Core biopsywas performed showing at the pathological report: a poorly differentiated CK7+ carcinomatous tumor which could be of the lacrimal gland or secondary metastatic origin.

To eliminate a metastatic lesion; a work-up looking for a primitive tumor was performed including urological examination, PSA (prostate specific antigen) and a CT scan of the chest and abdomen; showed no abnormalities.

In the Head & neck MDT Board upfront surgery was decided.Total exenteration was performed, which concluded to be a CK7+ moderately differentiated adenocarcinoma of the lacrimal gland (figure 4). A temporal muscle flap reconstruction was performed with a total skin graft.

The pathology report showed that the nearest excision limits are less than 0.5mm, presence of perineural invasion and without ocular invasion; therefore, adjuvant radiotherapy was indicated.

Radiotherapy was delivered using an intensity modulated technique (IMRT) with a total dose of 66 Gy in 33 fractions, at a daily dose fraction of 2 Gy over 6.5 weeks.

Conforming to ICRU 83, we were able to keep the tolerance doses of organs at risk within normal limits and at the same time deliver the intended dose of radiation to the tumorbed (Figures 5& Figure 6). The patient received

treatment according to plan and the tolerance was good. He was reviewed weekly during the treatment. There were no significant side effects observed except moderate skin reactions on the irradiated skin.

Figure 5:- Disposition of multiple IMRT beams.

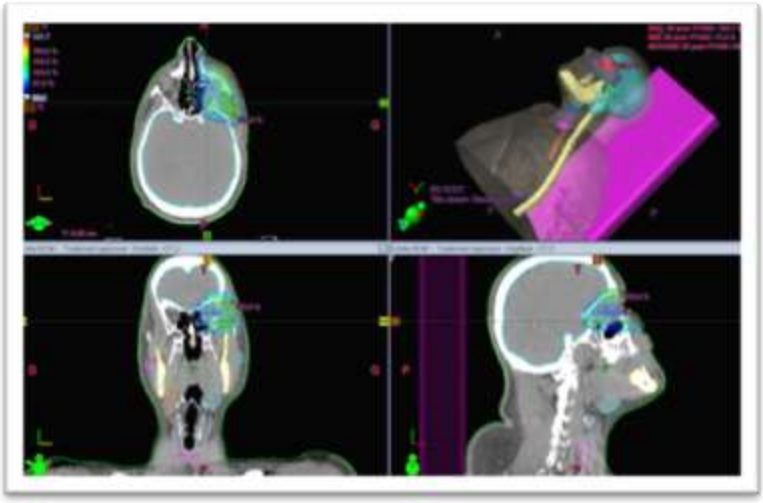
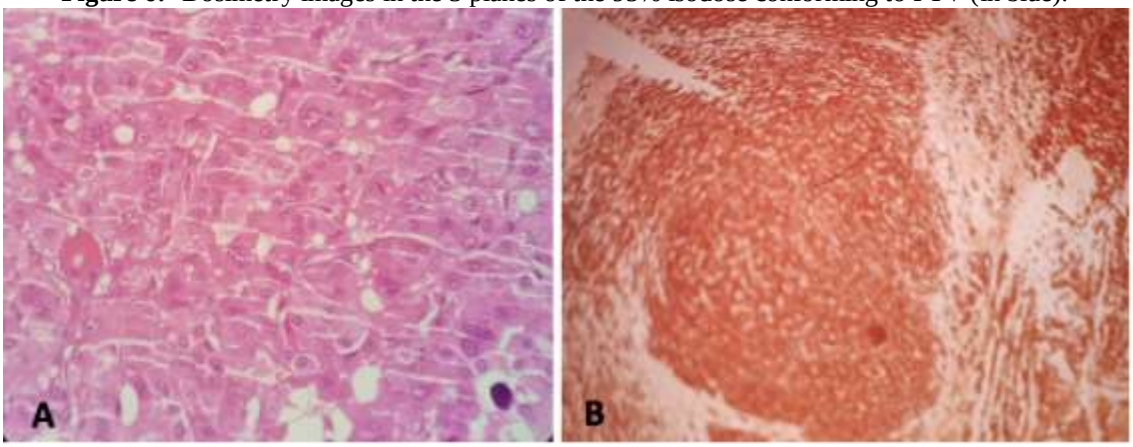


Figure 6:- Dosimetry images in the 3 planes of the 95% isodose conforming to PTV (in blue).



During the follow-up, the patient was seenevery three months alternately with his surgeon with an imaging (CT scan) every six months. After 18 months; the patient is still alive with no evidence of disease (local nor distal).

Discussion:-

Primary adenocarcinoma of the lacrimal gland (PALG) is much less common than in salivary glands and represents 5% to 7% of epithelial malignancies in this location and the most common epithelial malignancy is primary adenoid cystic carcinoma [1].

It affects older patients (18–80 years of age, mean 58 years of age) and is more common in the male population[4]. There appears to be no predilection for race or sex for lacrimal gland carcinoma but there is a suggestion that adenoid cystic carcinoma is slightly more common in women[5].

The initial sign is usually inferior and nasal displacement or exophthalmia of the globe and proptosis, owing to the supera- temporal location of the lacrimal gland in the anterior aspect of the orbit. Pain can be secondary to bone or nerve involvement and is often thought to suggest malignancy[2].

The treatment of PALG is challenging and includes surgery and additional adjuvant chemoradiotherapy or radiotherapy depending on disease stage, histological subtype, the presence or absence of perineural invasion,

incomplete surgical resection, involvement of regional lymph nodes, and extracapsular spread[6]. The mean purpose is early locoregional control[3].

In a retrospective study by Lin et al[7], included patients with lacrimal gland cancer who underwent either eye-preserving surgical treatment followed by adjuvant RT with or without chemotherapy, or needle biopsy followed by definitive RT with or without chemotherapy (in cases involving an inoperable tumor); two-year OS, LPFS, and DMFS rates were 69.0, 76.7, and 71.4%, respectively. Patients with early-stage (T1–T2) lacrimal gland cancer had significantly better outcomes than those with advanced-stage disease (T3–T4). Two-year OS, LPFS, and DMFS rates were each 100% in patients with disease stages T1–T2, and 37.5, 50, and 37.5%, respectively, in those with disease stages T3–T4 ($P < 0.05$). Orbital complications were well tolerated [7].

They conclude that adjuvant RT can achieve satisfactory results in patients with T1–T2 lacrimal gland carcinoma. Disease stage T3 and above was associated with poor outcomes even with post-operative RT, likely due to distant metastasis[7].

Given the radio-resistance nature of these tumors, adjuvant RT is always preferred to exclusive RT.The efficacy and safety of carbon-ion radiotherapy (CIRT) for patients with lacrimal gland carcinomas (LGCs) with extra-orbital extension was evaluated in retrospective study by Hayashi et al. in 33 patients [6].They conclude that definitive CIRT is effective for LGCs with extra-orbital extension and with acceptable toxicity.

For prognostic and in general, it appears that with shorter duration of symptoms before treatment, metastases are less likely to develop and the chance for long-term survival is greater [8].

Conclusion:-

Primary adenocarcinoma of lacrimal gland is rare and not inevitably fatal if diagnosed early and treated appropriately. Our patient is alive without evidence of recurrence at least 18 months after treatment. However, the best treatment and overall prognosis, although unknown, are dependent on stage and early diagnosis. Further studies are needed to standardize the best treatment option.

Consent:

The examination of the patient was conducted according to the Declaration of Helsinki principles.

Conflicts of interest:

The authors do not declare any conflict of interest

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