**ARC Journal of Clinical Case Reports**
*Volume 9, Issue 1, 2023, PP 1-5*
*ISSN No. (Online) 2455-9806*
*DOI: http://dx.doi.org/10.20431/2455-9806.0901001*
*www.arcjournals.org*

# Adenoid Cystic Carcinoma of The Lacrimal Gland: Case Report and Literature Review

**Rajaa El azzouzi*, Othmane Bouanani, Bouchra DANI and Malik Boulaadas**

*Department of Maxillofacial Surgery Hospital of Specialities-Rabat. Faculty of Medicine and Pharmacy-Rabat Mohamed V University.*

***Corresponding Author:** **Dr Rajaa EL azzouzi** *Department of Maxillofacial Surgery Hospital of Specialities-Rabat. Faculty of Medicine and Pharmacy-Rabat Mohamed V University..*

**Abstract:** *Adenoid cystic carcinoma or cylindroma is a locally invasive and slow growth malignant tumor. The lacrimal gland's localization remains very rare of all intraorbital tumors. We report the case of 44-year-old patient who consulted for painful unilateral exophthalmos as first sign with effect on visual function. A surgical biopsy was performed using a left hemicoronal approach confirmed the diagnosis. Treated with an eyelid-sparing orbital exenteration and an adjuvant external radiotherapy. No tumoral recurrence has been demonstrated after 18 months of follow-up.*

**Key Words:** *Exophthalmos, Cylindroma, Lacrimal gland, Rare Tumor.*

## 1. Introduction

Adenoid cystic carcinoma also known as cylindroma, is a malignant epithelial neoplasm, characterized by it's local invasion and it's high rate of recurrence.[1,2]

The lacrimal gland's localization occur relatively rarely of all intraorbital tumors. The diagnosis is mainly histological and a successful treatment requires a rigorous clinical and radiological evaluation..

## 2. Case Report

We report a case of 44 years old man, referred to our institution by ophtalmologists, with current smoking habit, who consulted for unilateral exophtalmos that has been evolving for 1 year, with general state preservation. The Maxillofacial examination on admission reveals the presence of a painful, unreducible left exophthalmos without inflammatory signs associated, the sensitivity of the face was preserved.

The visual acuity was 1/10 on left and 5/10 on right, with limitation of the left eye motility, and a chalazion of the right upper eyelid. The Fundoscopic examination revealed a superficial punctate keratitis on the left side, the rest of Slit lamp examination, intraocular pressures and fundoscopy were unremarkable.

Computed tomography (CT) scans of brain and orbit with contrast demonstrated a left intraorbital process with soft tissue density and intense homogeneous enhancement that involved the lacrimal gland, and encroached the superior rectus and external oblique muscles with no bony orbital or endocranial expansion. (Figure 1)

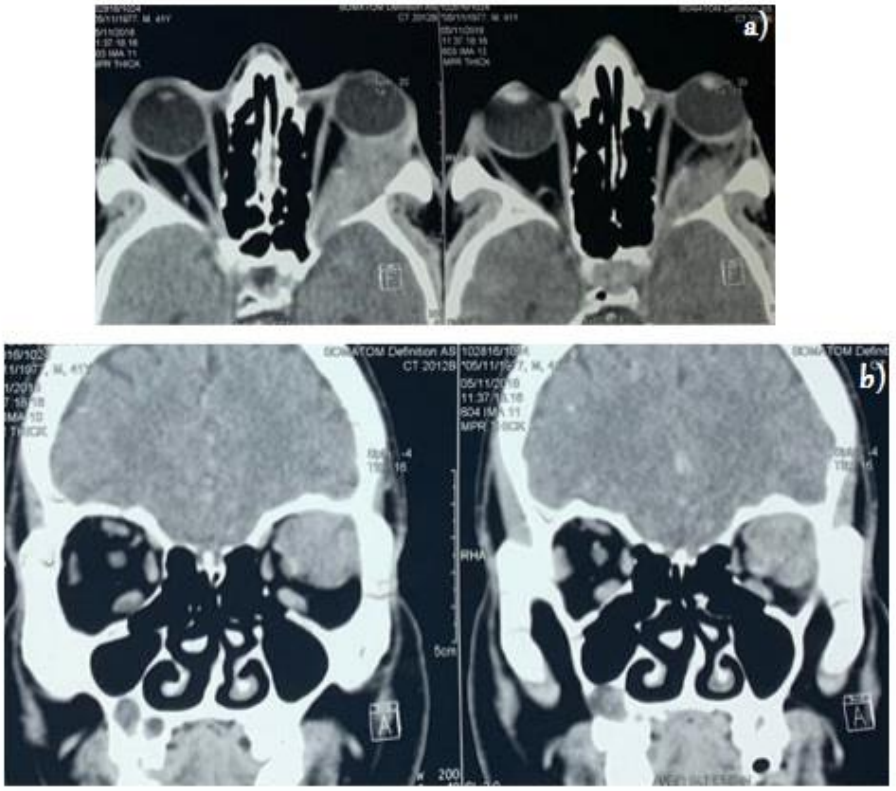


**Figure1.** *(a) Axial (b) Coronal of CT orbital scan of a left intraorbital process.*

T1, and T2-weighted magnetic resonance images (MRI) demonstrated an almost isointense and slightly heterogeneous tumor in the superolateral site of the left orbit, measuring approximately 41*17*26 mm with involvement of the superior rectus and external oblique muscles as well. Extending back to the optic canal and inferior orbital fissure. **(Figure 2)**

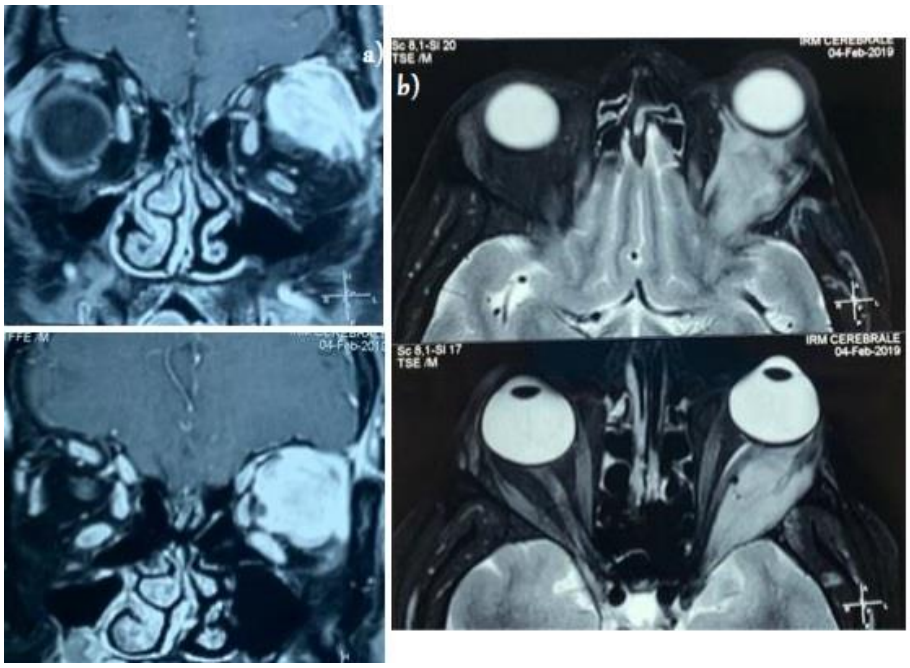


**Figure2.** *a) Coronal b) Axial MR images showing a left lacrymal gland tumor with a local invasion.*

The patient underwent a surgical biopsy using a left hemicoronal approach, (Figure 3) histopathologic evaluation revealed an adenoid cystic carcinoma of the lacrimal gland. In second instance and after patient's explicit free consent, an eyelid- sparing orbital exenteration

was performed using the same approach. The patient has received an external adjuvant radiotherapy.

The postoperative progress was uneventful, and the periodic follow-up examinations, including MRI of brain and orbit performed until 18 months after the surgical procedure showed no tumor recurrence and prepared the cavity for the placement of an orbital epithesis. (Figure 4)

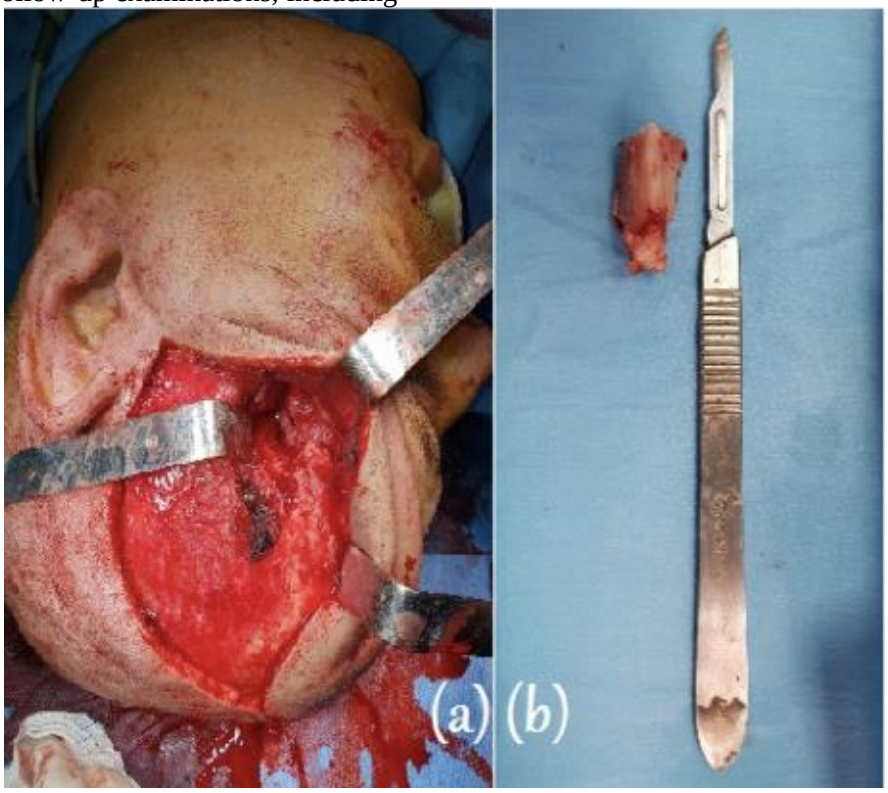


**Figure3.** *(a) Left hemicoronal approach after osteotomy of lateral wall of the orbit. (b) fragment of osteotomy.*

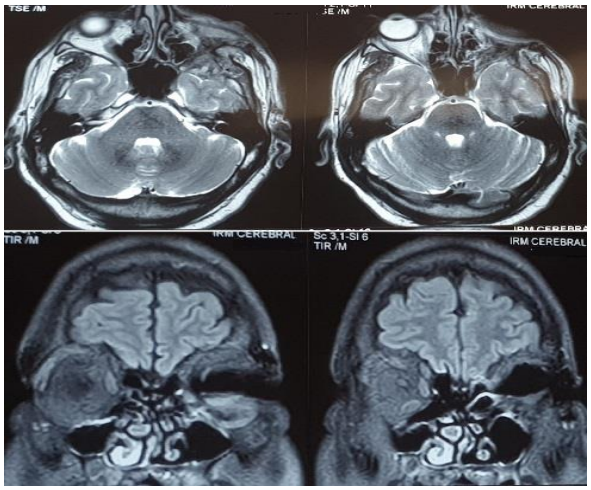

**Figure4.** *10 months post-operativly MRI of brain and orbit showing an empty orbital compartment*

## 3. DISCUSSION

Adenoid cystic carcinoma is a malignant epithelial neoplasm, originating in the exocrine glands from the major and minor salivary glands. It can also arise from the lacrimal gland, accounting for approximately 1.6% of all orbital tumors, 4.8% of primary orbital tumors, despite the rarity of this disease, it is the most common malignant epithelial cancer of the lacrimal gland, representing 35% of all epithelial tumors of the lacrimal gland [3].

The mean age occurrence is around 40 year-old with a peak incidence in the fourth decade and a second peak, in the sixth decade. A discreet female predilection is observed [1,4].

Painful unilateral exophthalmos, associated with hypoesthesia in the territory of the frontal nerve is the most frequent clinical sign. Pain is an important symptom of lacrimal gland malignancy and might be thought to indicate perineural infiltration with a more advanced stage of malignant disease. Most patients seek care within 6 months of onset of their symptoms. the Evolution is marked by the rapid onset of visual function disorders [5].

The CT scan is the modality of choice specially for evaluating the extent of the tumor. It reveals the tumor as an isodence process of lacrimal gland, allows the study of the measurement, the extent of the lesion and mass effect on adjacent structures, including the osseous orbital lesions and endocranial extension wich is described at an incidence of 4 to 22% in the cylindroma of the lacrimal gland [6]. The MRI has the advantage of an early detection of small tumors and of objectifying the extent of the tumor, but so as the CT scan, it can't confirm the nature of the tumor.

The diagnosis of certainty is histological. The histopathologic examination describes the adenoid cystic carcinoma as being well circumscribed tumors, without capsule, infiltrating the tissues of proximity, in particular the bone, the vascular and lymphatic axes and nerve endings. There are five histological types: Cribriform, solid or basaloid, sclerosing, comedocarcinomatous, tubular. All types can coexist within the same tumor, but in general only one histological type predominates.

The treatment is essentially surgical; The American Joint Committee on Cancer (AJCC) rules for classification of adenoid cystic carcinoma of the lacrimal gland include definitions for TNM wich correlates with the type of treatment chosen.The conservative treatment including globe-preserving surgery followed by RT is reserved only for < T3 tumors (tumor <2.5 cm in greatest dimension). For reserved prognosis tumors ≥ T3(tumor ≥ 2.5 cm in greatest dimension), the recommended treatment is orbital exenteration followed by RT, wich seems to be associated with less risk of local-regional recurrence [4].

Other authors propose in the absence of an endocranial extension a surgical approach in which the tumor and adjacent adnexa are removed in a one-stage procedure, that is an « en bloc resection » of the neoplasm, its periorbital base, and surrounding bone, by a superolateral of frontotemporal approachs. This technique will minimize seeding of the tumor, provide a more complete eradication of the neoplasm without loss of the eye, and assure either a cure or a longer interval free of local recurrences or metastasis than is presently attainable [7,8].

Radiotherapy alone seems to be insufficient for ACC. Although ajduvant radiotherapy of local resection ,delay significantly the recurrence of tumour and is associated with a slightly longer survival, especially when there is a residual microscopic tumor after surgery. According to a study by (Rapidis et al 2005) 83.3% of patients with positive lesion margins, had developed a local recurrence, radiotherapy probably contributed to the achievement of local control in the remaining 16.7% of tumors with positive margins [9].

The risk of local recurrence was higher in patients treated with local resection and who did not receive an adjuvant radiotherapy [4]. 50% of patients survive two and a half years after diagnosis [3], local bone invasion, lymph node and lung metastasis , explain this reserved prognosis. Adenis estimates the five-year survival rate at 45% [1].

In our case the tumor measured more than 4 cm in greatest dimension with extension to the orbital apex, an eyelid-sparing orbital exenteration was performed using a left hemicoronal approach, completed with an adjuvant external radiotherapy. No tumoral recurrence has been demonstrated after 18 months of follow-up.

## 4. Conclusion

Adenoid cystic carcinoma of lacrimal gland, is a malignant epithelial neoplasm, characterized by its local invasion and it's high rate of recurrence. Revealed by a painful unilateral exophthalmos, the biopsy orient the treatment which requires a close collaboration between surgeons and radiotherapists. Extended follow-up beyond 5 years is essential.

## References

[1] Adenis, J. P., and P. Saint-Blancat. "Tumeurs de la glande lacrymale." Pathologie orbito-palpébrale. Masson Paris, 1998.

[2] Garrity, James A., John Warren Henderson, and J. Douglas Cameron. Henderson's orbital tumors. Lippincott Williams & Wilkins, 2007.

[3] Lee, David A., et al. "A clinicopathologic study of primary adenoid cystic carcinoma of the lacrimal gland." Ophthalmology 92.1 (1985): 128-134.

[4] Ahmad SM, Esmaeli B, Williams M, Nguyen J, Fay A, Woog J, Selvadurai D, Rootman J, Weis E, Selva D, McNab A, DeAngelis D, Calle A, Lopez A. American Joint Committee on Cancer classification predicts outcome of patients with lacrimal gland adenoid cystic carcinoma. Ophthalmology. 2009; 116(6): 1210-15.

[5] Bernardini FP, Devoto MH, Croxatto JO. Epithelial tumors of the lacrimal gland: an update. Cur Opin Ophthalmol. 2008;19(5):409-13.

[6] Terasaki, Mizuhiko, et al. "High-grade adenoid cystic carcinoma originating from the lacrimal gland." Brain tumor pathology 17.3 (2000): 159-163.

[7] Wright, John E., Geoffrey E. Rose, and Alec Garner. "Primary malignant neoplasms of the lacrimal gland." British Journal of Ophthalmology 76.7 (1992): 401-407.

[8] Henderson, John W., and Roger W. Neault. "En bloc removal of intrinsic neoplasms of the lacrimal gland." *Transactions of the American Ophthalmological Society* 74 (1976): 132.

[9] Rapidis AD, Givalos N, Gakiopoulou H, Faratzis G, Stavrianos SD, Vilos GA, Douzinas EE, Patsouris E. Adenoid cystic carcinoma of the head and neck: clinico pathological analysis of 23 patients and review of the literature. Oral Oncol. 2005; 41(3):328-35.

**Citation:** *Rajaa El azzouzi, et.al., " Adenoid Cystic Carcinoma of The Lacrimal Gland: Case Report and Literature Review " ARC Journal of Clinical Case Reports (AJCCR). vol 9, no. 1, 2023, pp. 1-5 DOI: http://dx.doi.org/10.20431/2455-9806.0901001, www.arcjournals.org*