Case Report

Cystic Adenoid Carcinoma of Lacrimal Gland

SAULO BUENO DE AZEREDO[1], NATHALIA BECK CORREA[1], ARTUR EDUARDO MARTIO[2], PAULO MOACIR MESQUITA FILHO[1]

[1]*University of Passo Fundo, UPF, Passo Fundo, Rio Grande Do Sul, Brazil*

[2]*Atitus Eduacação, Passo Fundo, Rio Grande Do Sul, Brazil*

ABSTRACT: Adenoid cystic carcinoma (ACC) is an uncommon malignant tumor that accounts for less than 5% of head and neck cancers. ACC is characteristic for its indolent nature and its propensity for late distant metastases. Late diagnosis, tendency to perineural invasion, periosteal infiltration and local recurrence are factors of poor prognosis. Although studies still discuss the ideal treatment, the initial therapy consists of surgical resection, followed by postoperative radiotherapy.

KEYWORDS: *Lacrimal gland; adenoid cystic carcinoma; head and neck neoplasms.*

Introduction

Adenoid cystic carcinoma (ACC) is a rare type of malignant neoplasm, originating from glandular tissue, usually from the salivary glands, formed by an epithelial and myoepithelial component.

This neoplasm has three histological patterns, often coexistent in the same tumor: tubular, cribriform and solid, the latter being more aggressive than the others.

ACC has a slight predominance in women (6:4) and individuals between the fifth and sixth decades of life, with an age-adjusted prevalence of 4.5 cases per 100,000 individuals in this age group.

Despite its nonspecific symptomatology, this tumor is locally aggressive, with extensive perineural invasion.

The preferred treatment for ACC is surgical excision.

Recurrence is common, occurring in up to 75% of cases despite adequate surgical technique.

The 5-year survival rate is 77%, dropping to 45% at 15 years [1,2,3].

Herein, we report the case of ACC of the lacrimal gland, reviewing the existing literature on the condition.

Case Report

A 59-year-old white male was referred to the Neurosurgery department with progressive proptosis of the right eye, slightly painful on decompression.

No other abnormalities, such as impairment in ocular and systemic motricity, were observed on physical examination.

Magnetic resonance imaging (MRI) of the skull evidenced a solid, expansive lesion, in the superolateral margin of the right orbit measuring approximately 2.6cm × 2cm × 2.1cm.

The mass caused medial deviation of the right eyeball and proptosis, in addition to extrinsic compression of the orbital muscles close to the lesion; however, there was no apparent nerve, vascular or bone invasion (Figure 1).

The oncological staging, performed with computed tomography of the abdomen, chest and neck, did not demonstrate the presence of metastasis.

The patient underwent microsurgical excision of the lesion, through supraciliary access, in which a subtotal resection of the lesion was obtained, due to the loss of a clear cleavage plane between the lesion and the extrinsic ocular muscles (Figure 2).

Anatomopathological examination of the sample identified a malignant epithelial neoplasm, with a tubular and cribriform pattern, composed of cells with hyperchromatic nuclei and scarce cytoplasm.

The immunohistochemical study demonstrated the presence of two cell types: epithelial cells positive for cytokeratin and basal cells positive for p63, calponin and C-KIT (Figure 3); all these features are compatible with the diagnosis of ACC.

The postoperative evolution was satisfactory, with resolution of proptosis and no complications related to the surgical site.

The patient remained symptom free for eight months after surgery, and is still under follow-up.

Due to the subtotal resection of the tumor, the patient is currently undergoing local radiotherapy.

A written informed consent was obtained from the patient for publication of this case and any accompanying images.

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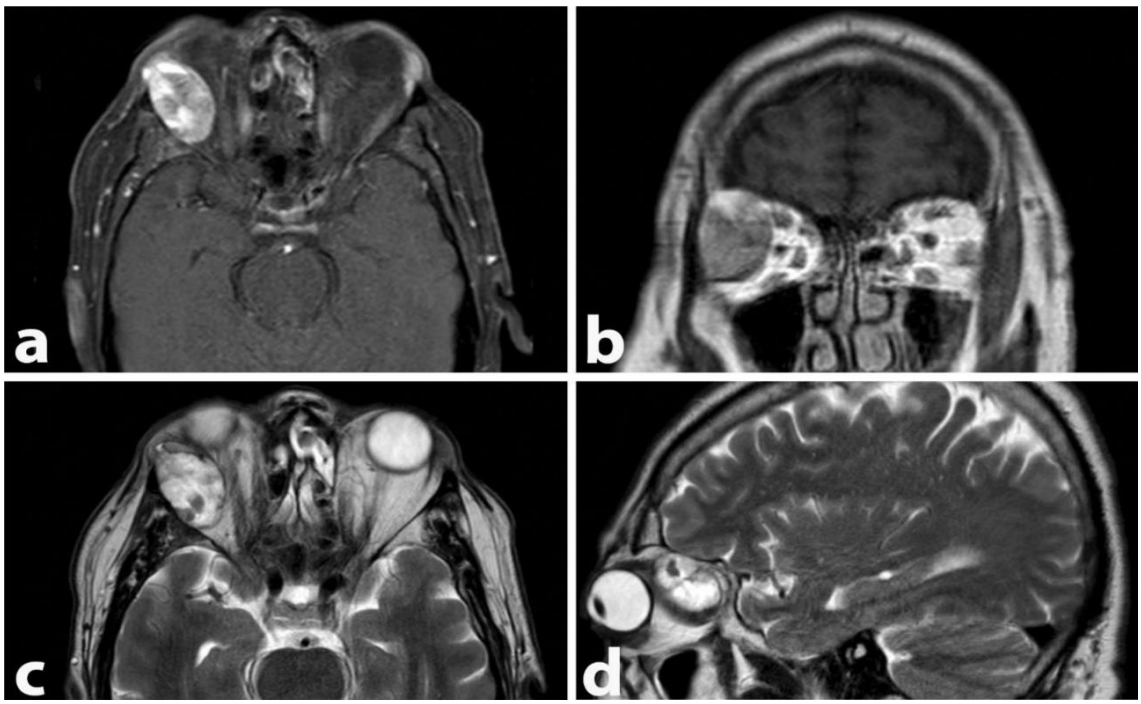

Figure 1. Pre-operative MR imaging. a) Axial T1-weighted gadolinium-enhanced MRI, evidencing a left, well delineated orbital lesion, in the lateral aspect of the orbit, displacing the extraocular muscles, with irregular contrast enhancement. b) Coronal T1-weighted gadolinium-enhanced MRI, evidencing a left, confirming the medial displacement of extraocular muscles and the contact of the tumor with the lateral rectus muscle. c) Axial T2-weighted MRI, exposing the irregular density of the tumor. d) Sagittal T2-weighted MRI, demonstrating the superior and lateral position of the tumor, inside the orbit.

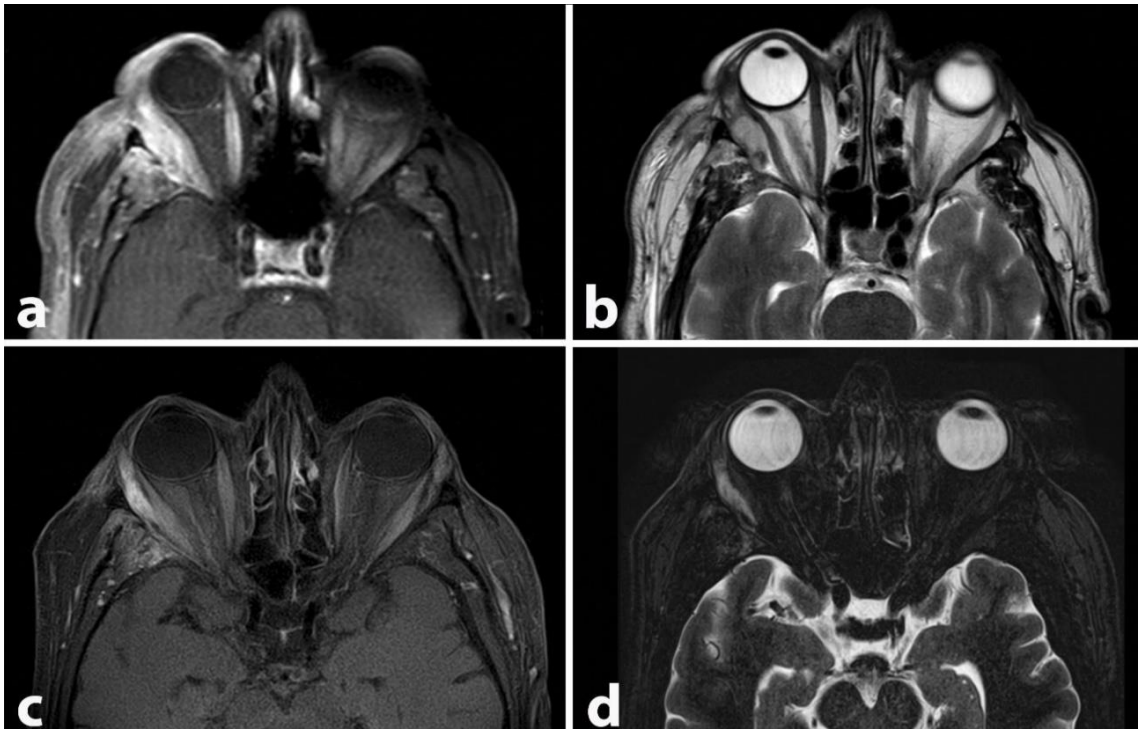

Figure 2. Post-operative MR imaging. a) Immediate axial T1-weighted gadolinium-enhanced MRI, evidencing the subtotal resection of the orbital lesion, with reminiscent area of irregular contrast enhancement. b) Immediate axial T2-weighted MRI, evidencing the small reminiscent tumor, in contact with the displaced lateral rectus muscle. c) Follow-up axial fat-suppression MRI, evidencing important reduction on tumor size, after radiotherapy. d) Follow-up axial T2-weighted MRI, demonstrating the important reduction in tumor size.

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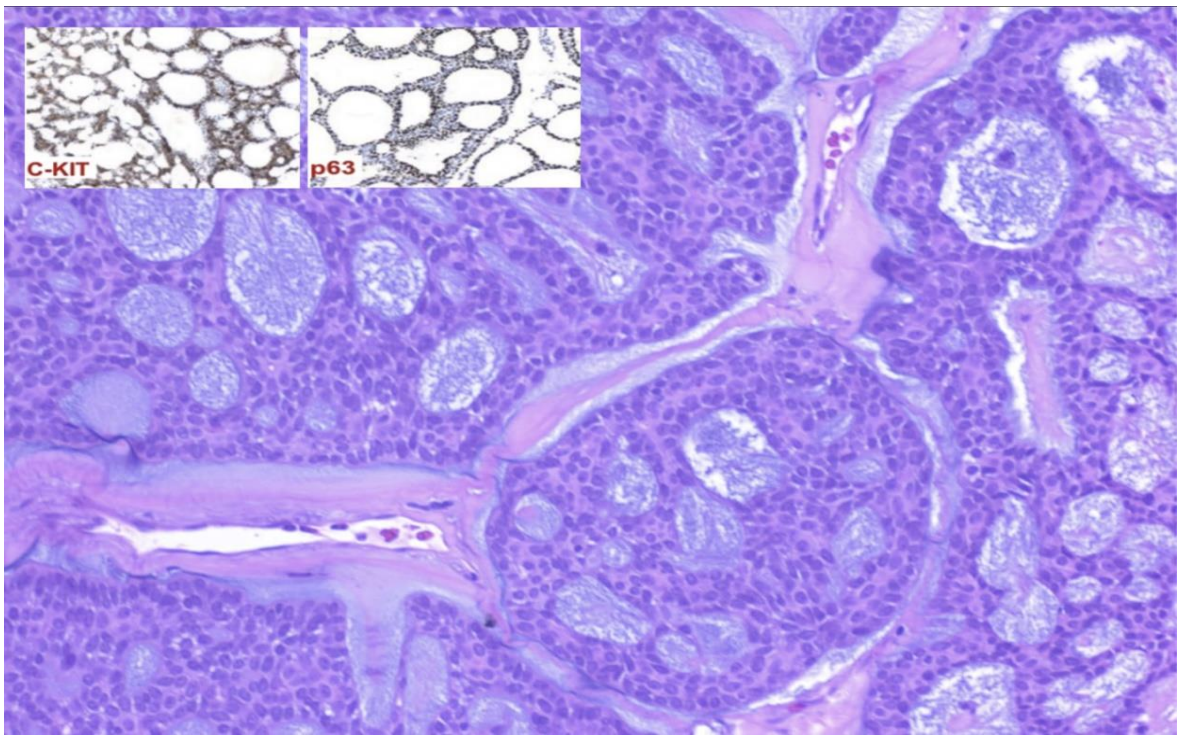

Figure 3. Pathology findings. H&E (10X)-Neoplastic tissue constituted by glandular structures, with cribriform aspect, and the presence of amorphous material inside the glandular tissue, and atypical nuclei. Immuno-histochemical stain confirming positivity to c-kit and p63.

Discussions

ACC is uncommon in the head and neck, with only 1% of cases occurring in this region.

When we talk about the orbital region, it represents only 1.6% of all tumors.

Even so, it is the most common primary malignant tumor of the lacrimal glands, being an important differential diagnosis to be considered in cases of painless lumps, whether or not associated with facial paresthesia [1,2].

Epiphora is the most common symptom of lacrimal gland tumors in general, followed by proptosis, ptosis, epistaxis, edema and skin ulceration.

Pain is a cardinal symptom in ACC, due to its known local aggressiveness, which generates early bone and perineural invasion.

Its invasive behavior makes rapid investigation and treatment necessary in order to improve prognosis.

However, the waiting time until the first appointment for evaluation is 6 months on average, and the diagnosis and initiation of treatment usually occur one year after the onset of symptoms [1].

Our patient only presented with proptosis, which is relatively uncommon and delayed diagnosis.

The delay allowed for greater tumor growth, which generated greater difficulty during the surgical procedure, as evidenced by the subtotal resection.

The initial evaluation is based on imaging exams. MRI is the method of choice; however, it is unable to differentiate ACC from other orbital masses with good specificity, making anatomopathological and immunohistochemical evaluation essential for a definitive and accurate diagnosis.

The differential diagnosis for ACC includes benign and malignant entities, such as pleomorphic adenoma, basal cell adenoma and adenocarcinoma, and low-grade pleomorphic adenocarcinoma [2].

Microscopy of this tumor type reveals three histological growth patterns: the cribriform, solid and tubular forms.

The cribriform pattern is the most common, while the solid pattern is the least frequent.

However, there may be a mixture of patterns within a single neoplasm [4].

In immunohistochemistry, c-kit receptor tyrosine kinase (CD117) is expressed in more than 90% of ACCs.

Another marker, p63, when positive and associated with CD117+, is associated with a higher histological grade and worse prognosis [5].

The initial treatment of ACC is surgical excision [6].

Once diagnosis is confirmed by anatomopathological and immunohistochemical evaluation, the patient must be staged.

When there are no signs of metastases, surgical excision can be complemented with adjuvant therapy, such as proton radiotherapy, external beam radiotherapy and brachytherapy [3].

Comparative case series have found no difference in survival for lacrimal gland carcinoma treated with cranio-orbital resection or tumor excision with eye preservation and radiotherapy.

Adjuvant radiotherapy is recommended after surgical resection, regardless of its extent, especially for patients at high risk of recurrence, including those with advanced tumor stage, positive surgical margins, or perineural invasion.

Adjuvant radiotherapy proved to be effective in preventing locoregional recurrences with local control rates of approximately 50%-80% at 5 years, despite no benefit in survival.

Furthermore, the role of postoperative chemotherapy for this tumor type remains unclear [6].

Metastases to lymph nodes occur only in about 4-9% of cases, rendering regional lymphadenectomy unnecessary in most cases. Subtotal resection is an important factor for disease progression and decrease in survival, which again emphasizes the importance of early diagnosis and intervention.

Subclinical metastases and perineural and bone invasion are related to higher rates of recurrence, distant metastases and mortality [2,3].

ACC can metastasize via hematogenous spread to the lungs, brain and bones, in decreasing order of frequency [6].

Recurrence is common, occurring in up to 75% of cases, despite adequate surgical technique.

The 5-year survival rate is 77%, while at 15 years there is a drop to 45%.

These values are influenced mainly by the late discovery of distant metastases [1,2,3].

Conclusion

ACC is a rare, slow-progressing disease, associated with poor local control, distant metastases, and significant long-term morbidity and mortality rates.

Delay in diagnosis, tendency to perineural and periosteal invasion, and high rates of local recurrence are factors that result in poor prognosis.

To date, the ideal treatment is still debated, with surgical excision being the first line.

Close follow-up after treatment should be performed to assess recurrence and need for reoperation and adjuvant therapy.

Larger multicenter studies may provide additional information and should be carried out in the future.

Conflict of interests

None to declare

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Corresponding Author: Saulo Bueno De Azeredo, University of Passo Fundo, UPF, Passo Fundo, Rio Grande Do Sul, Brazil, e-mail: saulodeazeredo@yahoo.com.br

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