

Adenoid cystic carcinoma of the lacrimal sac: A case report

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ABSTRACT

Introduction: Adenoid cystic carcinoma (ACC) of the lacrimal sac is a rare entity characterized by poor prognosis, because of the high rate of local recurrence and metastasis after treatment.

Case Report: A 45-year-old male patient was seen at our institution for epiphora, pain, and swelling of the internal angle of the left eye evolving for three months. The clinical examination revealed a fixed mass above the medial canthus of the left eye with blood discharge from the lacrimal punctum at the pressure of the mass. Orbital magnetic resonance imaging (MRI) showed a left lacrimal sac mass. Incisional biopsy disclosed adenoid cystic carcinoma.

Conclusion: This is a rare case of lacrimal sac malignant tumor. Adenoid cystic carcinoma can develop in the salivary or lacrimal glands; however, its occurrence in the lacrimal drainage apparatus is extremely rare. Imaging plays a key role in the diagnosis, thanks to computed tomography (CT) scan and magnetic resonance imaging (MRI) which allow a detailed analysis of the local and distant extension, as well as in post-treatment follow-up.

Keywords: Adenoid carcinoma, Lacrimal sac, MRI

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INTRODUCTION

The most frequent malignant tumors affecting the lacrimal sac are of the epithelial type, with squamous subtype having the highest incidence in this group. Adenoid cystic carcinoma (ACC) is an extremely unusual entity in the lacrimal drainage system. According to the latest statistics for the year 2016, 11 cases of ACC of the lacrimal sac were reported [1].

Adenoid cystic carcinoma (ACC) is a malignant tumor characterized by local aggressivity and slow evolution. It is a malignant epithelial tumor usually developed in the salivary glands; its orbital localization is very rare, accounting for 1.6% of all orbital tumors [2].

Adenoid cystic carcinoma is known for its slow progression and recurrence despite treatment. The prognosis is poorer than salivary gland ACC despite extensive surgery and postoperative radiation therapy [3].

Computed tomography and magnetic resonance imaging of the orbit play a crucial role in the positive diagnosis of adenoid cystic carcinoma of the lacrimal sac, allowing an analysis of the local extension, in particular perineural, bony, and intracranial, as well as in the post-treatment follow-up for the detection of recurrences and distant metastases which are very frequent [4].

CASE REPORT

We report a case of a 45-year-old male patient, with no past medical history, presenting with epiphora, pain, and swelling of the internal angle of the left eye that had progressively worsened over the preceding three months. The clinical examination revealed a hard, fixed mass above the medial canthus of the left eye with blood discharge from the lacrimal punctum at the pressure of the mass. Visual acuity was 04/10 in left eye and 10/10 for the right eye, visual fields,

intraocular pressure were intact and the fundoscopic examination was normal.

Orbital MRI showed a left lacrimal sac mass, hyperintense on T1-weighted image, heterogeneous T2-hypertintense, enhanced after gadolinium injection; measuring 28×31×37 mm, infiltrating the extraconal fatty tissue and the medial wall of the left orbit with extension toward the nasolacrimal duct to the left nasal fossa (Figures 1 and 2).

The medial rectus muscle appeared enlarged, suggesting invasion by the tumor.

Incisional biopsy disclosed ACC with a cribriform pattern infiltrating the muscle and bone tissue with additional perineural infiltration. In the extension study, there was no evidence of tumor metastasis.

An exenteration was indicated, but the patient refused the procedure. The patient underwent a complete excision of the lacrimal sac as well as the lacrimal drainage system followed by radiotherapy, with no signs of recurrence

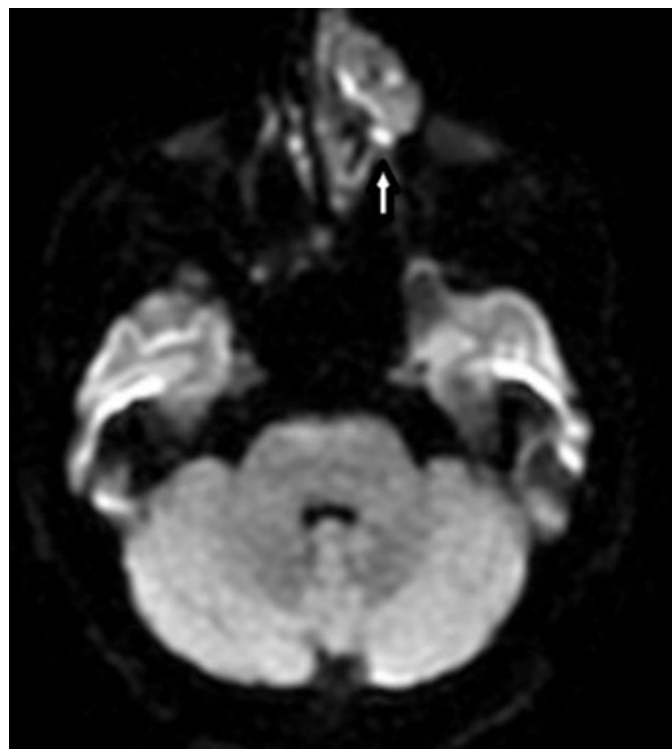


Figure 1: Orbital MRI on diffusion WS showing a left lacrimal sac mass (arrow) hyperintense measuring 28×31×37 mm, infiltrating the extraconal fatty tissue and the medial wall of the left orbit with extension toward the nasolacrimal duct to the left nasal fossa.

observed after a follow-up for six years.

DISCUSSION

Primary malignant tumors of the lacrimal sac are extremely rare, with epithelial malignancies being the most common, including squamous cell carcinomas,

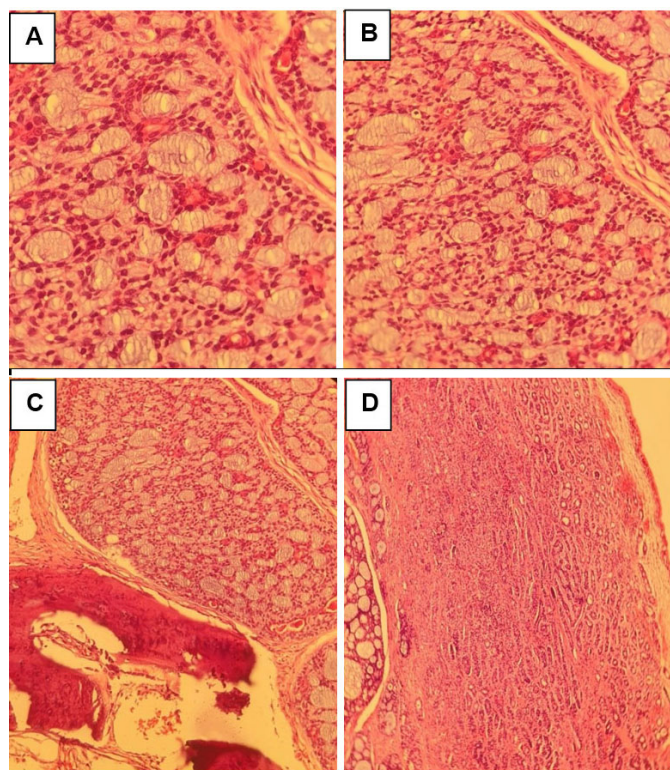


Figure 2: Histological examination images. (A) Eosin-Hematein ×40: tumoral proliferation of tubular and cribriform architecture. We recognize the conjunctival surface coating. (B) Hematein-Eosine ×40: tumor proliferation infiltrating bone tissue. (C) Hematein-Eosine ×200: higher magnification of tumor proliferation showing cribriform character. (D) Hematein-Eosine ×400: detail of small tumor cells, monomorphic, placed around cavities housing a basophilic material.

transitional cell carcinomas, adenocarcinomas, mucoepidermoid carcinomas, oncocytic adenomas, and ACC “adenoid cystic carcinoma”) [5].

Adenoid cystic carcinoma is a malignant tumor of epithelial origin that can develop in the lacrimal glands or salivary glands and it is also found in the mucous glands of the respiratory tract. The orbital localization usually derives from the lacrimal gland and more rarely from mucous glands of lacrimal sac. It is a slow-growing tumor but has a tendency to perineural invasion and spread to adjacent tissues, such as bone. Local recurrence is frequently seen several years after surgical excision. However late, hematogenous metastases are possible and most often involve the lungs [1, 4].

The clinical presentation evolves through three stages. Stage one includes a simple dacryocystitis with epiphora. In the following stages, the formation and extension of the tumor is witnessed, with the appearance in some situations of sanguineous discharge from the lacrimal punctum, epistaxis, and pain which are considered signs of malignancy. More rarely, skin ulceration and lymphadenopathy are observed.

Although distant metastases have been described, local spread to bone and eventual intracranial extension is generally the rule [5].

Accurate pretreatment imaging diagnosis and initial imaging evaluation in the ACC of the lacrimal sac are necessary in the guidance of surgical resection and definition of target volume in radiotherapy. Imaging is also important in the diagnosis of distant metastases (lungs, bone, etc.) [6].

Computed tomography scan of an adenoid cystic carcinoma of the lacrimal sac may show a characteristic infiltrative mass involving the medial canthus with possible bony remodeling and widening of the lacrimal bone [7].

Magnetic resonance imaging allows a more accurate study of the tumor's relationship with adjacent structures and its intracranial extension and perineural extension, but it is less sensitive than CT in the detection of bone invasion. On T1-weighted images, ACC of the lacrimal sac shows an ill-defined isointense signal, a homogeneous hyperintensity on fat suppressed T2-weighted images with a diffuse enhancement after gadolinium injection [4, 8].

Computed tomography combining with MRI can improve the diagnostic accuracy of this disease, but the histological study is necessary for a definitive diagnosis. Histopathology of adenoid cystic carcinoma reveals it as malignancy of modified myoepithelial (abluminal) and ductal (luminal) differentiated cells. The five histological patterns are: Basaloid (worst prognosis), sclerosing, cribriform "Swiss cheese-like" (most frequent pattern), tubular, and comedo carcinoma, seen in varying combinations and dominance [8].

Treatment of lacrimal sac ACC requires complete excision of the lacrimal sac as well as the lacrimal drainage system (canaliculi and nasolacrimal duct) followed by radiotherapy and/or adjuvant chemotherapy. Adjuvant radiotherapy has been shown to improve survival, especially in cases of perineural invasion [5]. Due to the aggressive nature of ACC, local recurrence is very common and occurs in about 50% of patients within two years with soft tissues and bone as the most frequent site [9].

CONCLUSION

The ACC of lacrimal sac is a rare malignant tumor with a high aggressiveness requiring multidisciplinary management, involving the ophthalmologist, radiologist, oncologist, and radiotherapist.

Imaging combining CT and MRI can improve the diagnosis accuracy of this disease, and show a capital importance in the post-treatment follow-up, which must be for life, to detect local recurrence and metastasis.

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Author Contributions

Kaoutar Imrani – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Tlaite Oubaddi – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Hounayda Jerguigue – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final

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Rachida Latib – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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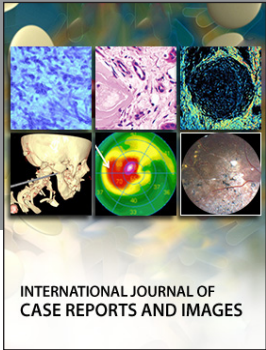
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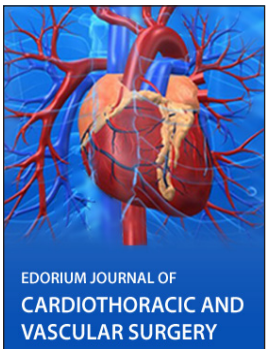
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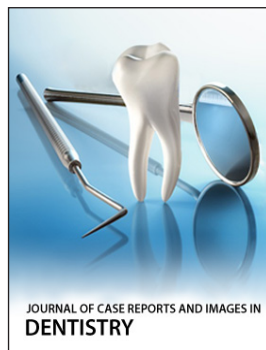
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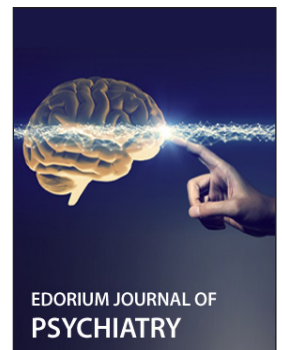
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