CASE REPORT
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Myoepithelial carcinoma of lacrimal gland in HIV patient: A rare case

L.P. Suryantini Septia Dewi[1*], Putu Yuliawati[2], Ni Made Laksmi Utari[2], A. A. A. Sukartini Djelantik[2]

[1]Ophthalmology Registrar of Ophthalmology, Faculty of Medical and Health Science, Universitas Udayana, Sanglah General Hospital Denpasar Bali;
[2]Ophthalmologist of Faculty of Medical and Health Science, Universitas Udayana, Sanglah General Hospital Denpasar Bali;

*Corresponding author:
L.P. Suryantini Septia Dewi,
Ophthalmology Registrar of Ophthalmology, Faculty of Medical and Health Science, Universitas Udayana, Sanglah General Hospital Denpasar Bali;
tiaq.08@gmail.com

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ABSTRACT

Introduction: Lacrimal gland tumors have 1: 1,000,000 cases per year, accounting for one-fourth of lesions that occupy the orbital space. The rarity of lacrimal gland tumors and the many types of tumors with various types of lesions make determining optimal treatment difficult. A myoepithelial tumor is a rare epithelial neoplasm of the lacrimal gland. This paper aims to report a good outcome case of myoepithelial carcinoma.

Case Report: A 30-year-old male complained of protruding his left eye since 2014, accompanied by pain and clear white discharge. The vision was getting blurry and also double vision. Patients with Human Immunodeficiency Virus (HIV) disease since 2013 on Anti Retro Viral (ARV) therapy. Right eye visual acuity was 6 / 7.5, and good eyeball examination. The left eye was 6/10 with no pinhole improvement, non-axial proptosis, and superolateral mass. The movement of his left eye was restricted. Other anterior and posterior segments were promising. The patient was diagnosed with pseudotumor and was given oral methylprednisolone. The eye condition is said to be getting better. The patient complained that the left eye was more prominent and protruding three years later, with blurred vision accompanied by infection. Left eye visual acuity becomes Light Perception Bad Projection with non-axial proptosis, retraction of palpebra and pus. Orbital exenteration was performed, and anatomical pathology examination revealed myoepithelial carcinoma of the lacrimal gland. The patient was planned for radiotherapy.

Conclusion: These rare cases have made setting up a regimen in randomized controlled trials difficult. The rapid development of therapy gives more hope to treating patients with lacrimal gland tumors.

Keywords: Myoepithelial Carcinoma, Lacrimal Gland, Epithelial Neoplasm, HIV.
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INTRODUCTION

Lacrimal gland tumors have an incidence of 1 case per 1,000,000 persons per year and constitute a quarter of the lesions occupying the orbital space, with most tumors being benign. These lesions are of epithelial origin in more than half of cases and lymphoid origin in one-third of patients. The remaining 10–15% are mesenchymal or secondary tumors either as direct invasion or distant metastases.[1] A myoepithelial tumor (MET) is an uncommon epithelial neoplasm of the lacrimal gland. Primary epithelial neoplasms account for 23-70% of histologically lacrimal gland lesions. Seventy percent of lacrimal gland epithelial neoplasms are pleomorphic adenomas, followed by adenoid cystic carcinoma and ex-pleomorphic carcinoma. Rare tumors of the lacrimal gland include basal cell adenocarcinoma, ductal carcinoma, acinar cell carcinoma, mucoepidermoid carcinoma, oncocytoma and oncocytic carcinoma, polymorphic low-grade adenocarcinoma, myoepithelioma and myoepithelial carcinoma.[2] Myoepithelial carcinoma of the lacrimal gland is rare. These lesions have difficulty making the diagnosis because they usually have non-specific clinical and radiological findings. Suspicion of a malignant lacrimal gland tumor requires a biopsy with permanent histologic confirmation.[3] These tumors are also characterized by histological polymorphism. Myoepithelial carcinoma is characterized by nested myoepithelial cells with an atypical nucleus, a mitotic rate of 1 mitosis per 10 high-power fields, and no glandular formation. Immunohistochemical staining results were positive for keratin, irregular positive for S-100 protein, and weakly positive for actin.[4]

Management of epithelial lacrimal gland tumors includes surgery, and in the case of malignancies, adjuvant chemoradiation is added or neoadjuvant intra-arterial chemotherapy, depending on the stage of the disease. Management of benign epithelial lacrimal gland tumors includes resectioning the lacrimal gland mass, including pseudo capsule, especially in pleomorphic adenomas. Incisional biopsy should be considered carefully to avoid recurrence when pleomorphic adenoma is suspected from clinical and radiographic findings.[1]

Exocrine glands produce and secrete fluid into the epithelial surface through ducts. The lacrimal gland is an exocrine gland that produces tears. HIV infection has

many ophthalmic manifestations, ranging from underlying microvasculopathy to opportunistic infections to autoimmune reactions. These complications affect 50-75% of people infected with HIV. The part of the eyeball involved can include any component of the eye, including the orbit and adnexal structures, the anterior segment, and the posterior segment. Posterior segment lesions have more significant morbidity. Anterior segment lesions, such as keratoconjunctivitis sicca, are common and impact patients' quality of life. HIV-1 virus and HIV viral load have persisted in patient tears. The etiology of lacrimal gland abnormalities in HIV patients is usually thought to be due to HIV lymphocytic infiltration of the lacrimal gland. However, many infections are opportunistic and have underlying and worsening symptoms. It also causes damage to the lacrimal acini and the ductal system and damage to the conjunctiva. Keratoconjunctivitis contributes to a chronic inflammatory state, which further increases cytokines' secretion, breakdown, and appearance that causing lacrimal gland dysfunction and loss of tear production.[5] This paper aims to report a good outcome case of myoepithelial carcinoma.

CASE ILLUSTRATION

A 30-year-old male patient came to the eye clinic at Sanglah Hospital on July 19, 2017, with a complaint of a prominent left eye for the previous three years (Figure 1). The left eye has been swollen since 2014. Initially, the lump was small on the upper eyelid and became more significant and prominent. Complaints are also accompanied by pain and clear white discharge. Vision is also getting blurry slowly, and one begins to experience double vision, which improves when one eye is closed.

The patient previously went to the Johannes Kupang Hospital. A CT scan was performed in January 2016. It was said that there was a mass in the left orbital cavity, suspicious of the lacrimal gland (Figure 2). The patient was referred to Sanglah Hospital for further treatment. The patient had a history of HIV disease since 2013, with an absolute CD4 count of 205 at the time of diagnosis. The patient has received antiretroviral therapy with lamivudine, zidovudine, and nevirapine. Currently, his CD4 count reaches 415. History of wearing glasses, allergies, smoking and family history of suffering from the same complaint was denied. The patient worked as an employee in the social service.

The visual acuity of his right eye was 6/7.5, and his left eye was 6/10 and was not improved using a pinhole. Abnormalities found in the left eyelid were non-axial proptosis but no lagophthalmos. There was a superolateral mass on the left conjunctiva, approximately 14 x 5 x 3 mm, well-circumscribed, mobile, dense consistency, color resembling surrounding tissue, minimal pain to touch, and no bleeding. The movement of the left eyeball was restricted to superolateral, superior, and superomedial (Figure 3). Lumps in other areas such as the neck, armpits, and breasts were denied. The patient was diagnosed with Oculi Sinistra proptosis non-axial et causa suspect retrobulbar tumor + suspect Ocular Surface Squamous Neoplasia (OSSN) dd lymphoma. It is planned to do a CT scan. The result was a solid left orbital retrobulbar mass that fused with the left lateral rectus muscle pushing the optic nerve medially and causing left bulbus oculi proptosis, bilateral maxillary sinusitis, and no visible intracranial infiltration.

A biopsy was performed on August 4, 2017, at Sanglah Hospital, but there was no pleomorphic adenoma in the biopsy preparation. The patient was

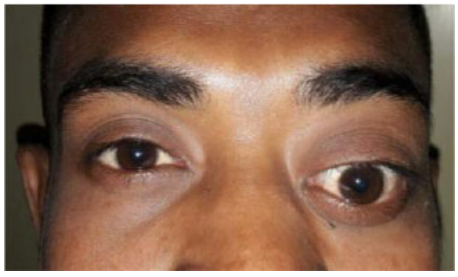

Figure 1. There was an increased mass in the left upper eyelid accompanied by protrusion of the eyeball when it first came on July 19, 2017.

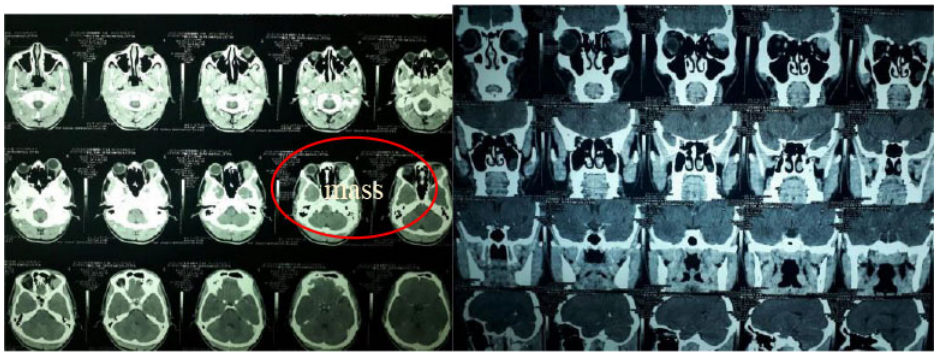

Figure 2. CT Scan from the Johannes Kupang Hospital, CT scan of the head of the axial and coronal slices, without and with contrast when first arrived in 2017.

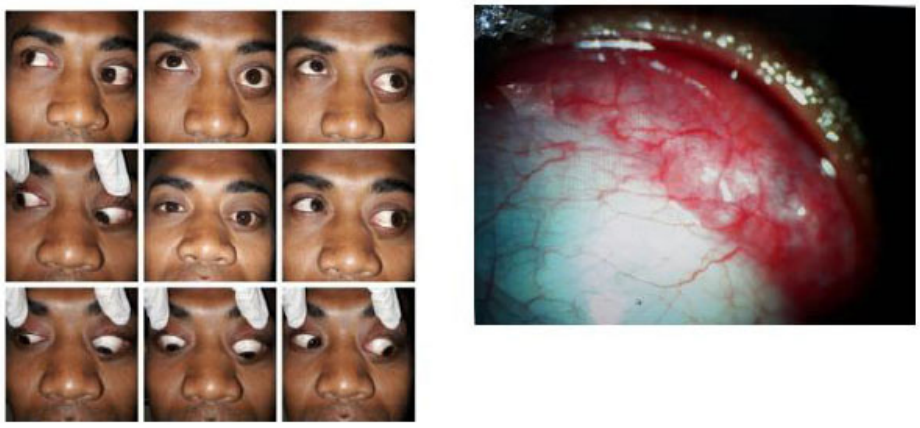

Figure 3. There is an obstruction movement of the left eyeball superiorly, superomedially and superolateral, as well as a mass in the left superior palpebra in the lateral region (in 2017).

diagnosed with a pseudotumor and received oral methylprednisolone. Oral methylprednisolone with an initial dose of 32 mg and reduced by 20% every two weeks, the patient took medicine for three months. The patient's current condition is good, with an absolute CD4 of 638. The patient's eye condition has improved, and the protrusion of the eyeball has decreased. The patient goes back to Kupang.

The patient returned to Sanglah Hospital after three years, on August 3, 2020, complaining that his left eye was getting more prominent. Complaints have started to worsen, and the lump has been growing faster since March 2020. Blurred vision has also been felt since April 2020, accompanied by infection in the eyeball and white discharge. Double vision has also persisted since 2017. Eye movement has worsened since March 2020 with pain when moving the eyeball. The patient is currently on ARV treatment, and the absolute CD4 count is 627. Visual acuity in the left eye deteriorated to Light Perception and Bad Projection with non-axial proptosis, eyelid retraction and pus. Found a mass on the superior eyelid measuring 7x15 mm solid, fixed, color resembling the surrounding skin and no tenderness, conjunctival chemosis, corneal edema, anterior and posterior segments were difficult to evaluate. Examination of the right eye was good with 6/12 pinhole and 6/7.5 vision. (Fig. 4). No other lumps on the neck, armpit, and chest were found.

Head CT Scan with and without contrast at Prof. Hospital WZ Johannes Kupang on May 12, 2020, found a lobulated solid, heterogeneous contrast-enhanced mass, approximately 4.3x3.4x3.2 cm in the retroorbital superolateral left orbital cavity. The mass appeared attached to the posterolateral side of the left orbit and was seen pressing the left orbit anteriorly and pressing the optic nerve to the media. No erosion or destruction of bone around the eyes was seen. The right orbit, right optic nerve and right extra oculi muscle did not show any abnormalities. The impression is that there is a solid, heterogeneous contrast-enhanced mass, lobulated, measuring approximately 4.3x3.4x3.2 cm in the retroorbital superolateral to the left orbital cavity with a suspected lacrimal gland tumor (Fig. 5).

The patient has been diagnosed with oculi Sinistra malignant tumor of the orbital lacrimal gland with suspected adenoid cystic carcinoma and suspected adenocarcinoma, malignant mixed tumor + HIV based on the history, physical examination and the latest CT scan in 2020. Orbital exenteration was carried out on August 25, 2020. Intra-orbital soft tissue, periorbita and eyelids were excised entirely, and anatomical pathology was examined. An evaluation was performed intraoperatively. It was found that the sphenoid bone was eroded. The mass obtained after the extraction process was 11x10x5 cm (Figure 6). The

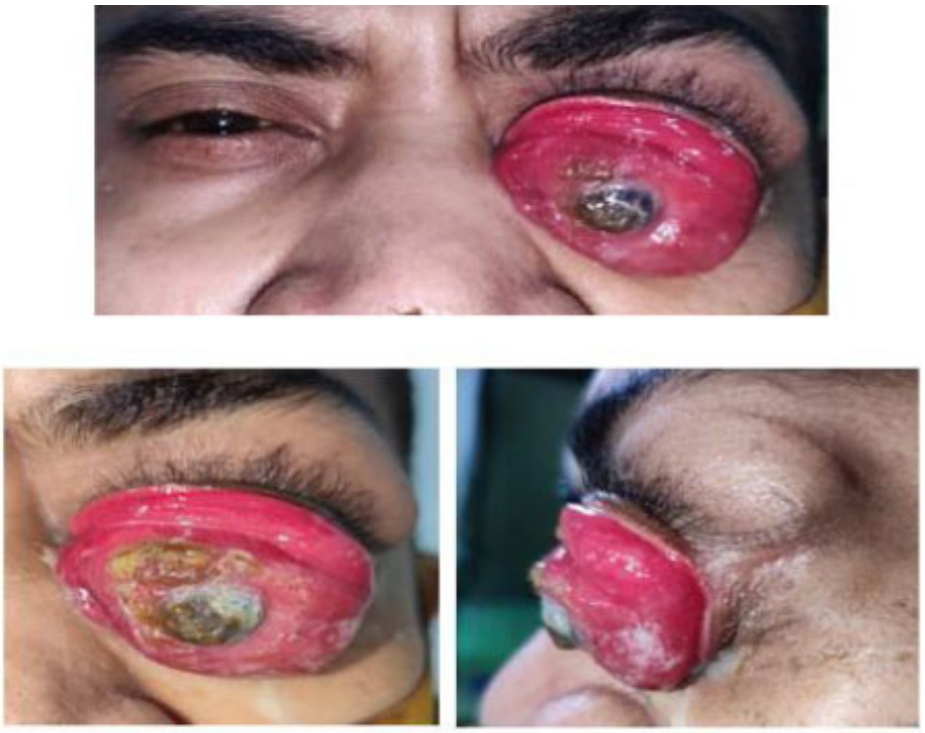

Figure 4. Development of the left eye after three years when the patient returned (3/08/2020).

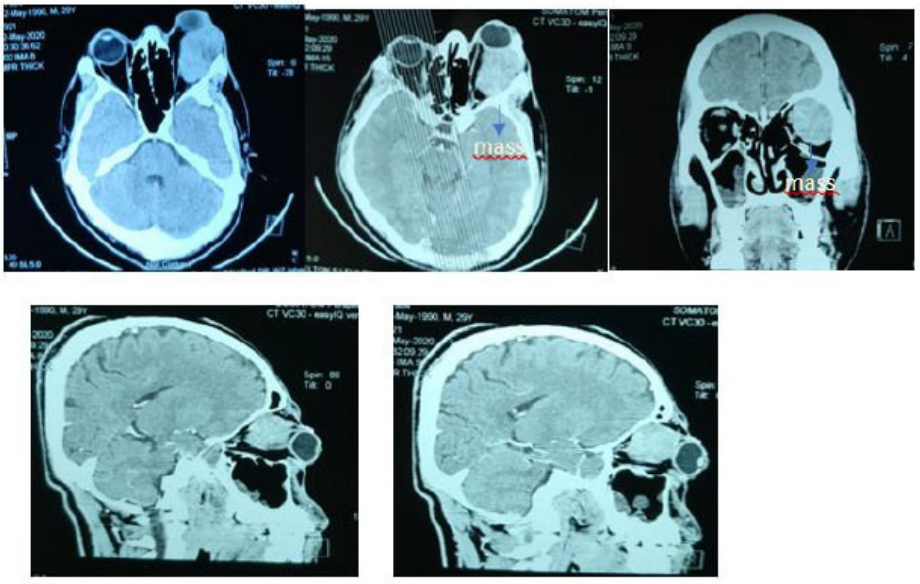

Figure 5. Head CT Scan with and without contrast at Prof. Hospital. WZ Johannes Kupang on May 12, 2020, after experiencing a worsening.

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orbital bones are left after extraction to undergo spontaneous granulation and epithelization.

Anatomical and pathological examination was carried out, and the results were no invasion of tumor cells in the eyeball and optic nerve (Fig. 7). Immunohistochemistry showed an immunohistochemical staining pattern, indicating that the tumor cells were of epithelial origin (carcinoma). The specific carcinoma subtype has not yet been determined (Fig. 8).

The patient was diagnosed with post-exenteration anophthalmia left oculi et causa myoepithelial lacrimal gland carcinoma regarding the results of anatomical pathology examination. The patient planned for radiotherapy and got scheduled six months later. The patient was checked in Sanglah Hospital on September 29, 2020, for the last time, with no active bleeding in the socket and no signs of infection. The patient said he started to experience a lump in his left breast six months later. The swelling was initially small and said to be getting more prominent. The patient said he would check the lump at Hospital in Kupang, but this has not been done.

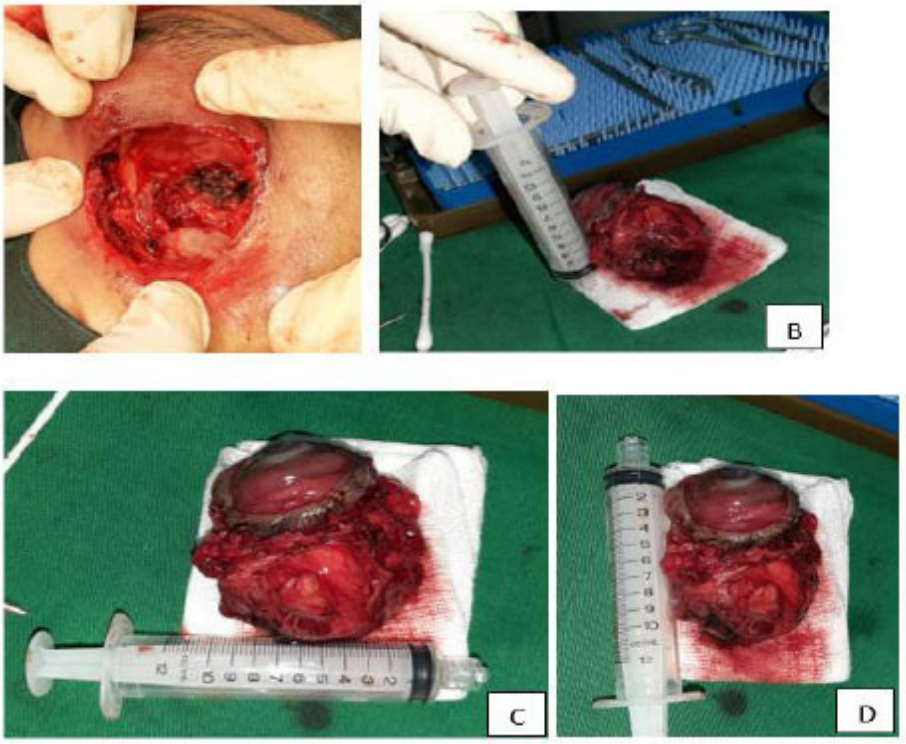

Figure 6. Post exenteration. (A) shows erosion of the sphenoid bone. (B, C, D) shows the mass size of 11x10x5 cm.

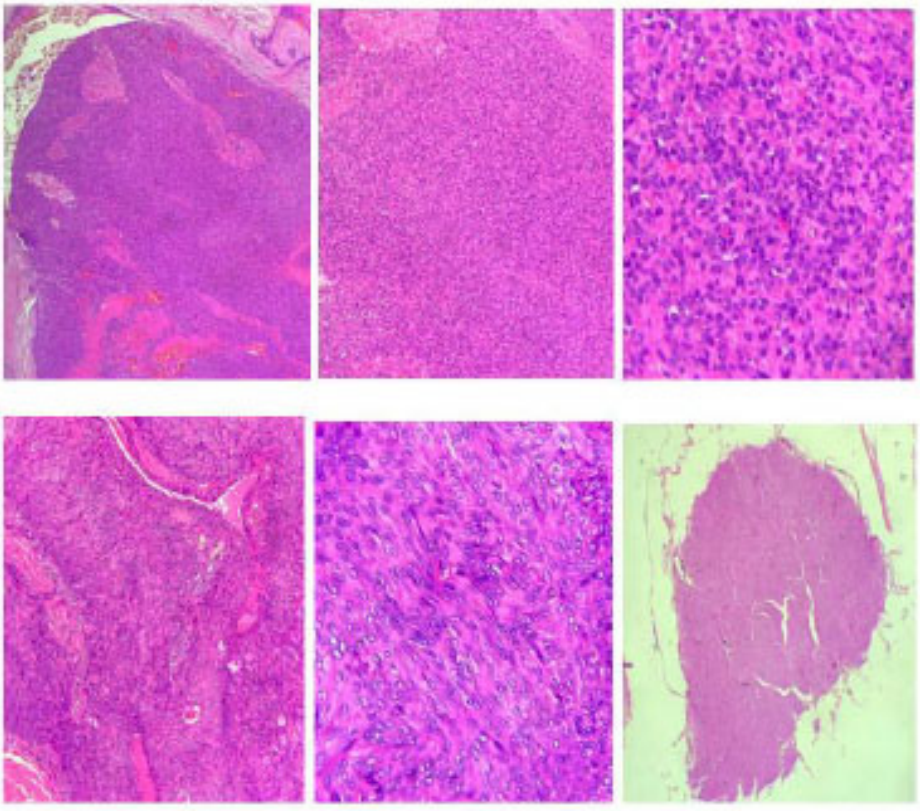

Figure 7. Histological results from hematoxylin and eosin staining.

DISCUSSION

Lacrimal gland lesions represent 9% of all orbital lesions. Primary epithelial tumors account for less than 30% of lacrimal gland lesions. Mahdi et al. Mentioned the symptoms that are usually complained of by patients with myoepithelial lacrimal gland carcinoma in the form of proptosis, decreased visual acuity, a mass in the superolateral area, pain around the orbit and diplopia.[6] The patient in this case report showed symptoms of a lump on the eyelid and pushing the eyeball. Visual acuity decreases slowly, and complaints of double vision are accompanied by pain. The diagnostic algorithm of lacrimal gland mass can be described as follows, CT scan or MRI imaging is performed first. After an imaging diagnosis is made, further consideration is whether to perform an incisional biopsy or total surgical resection of the mass. Wright et al. recommend an approach based on the duration of symptoms and bone changes in radiographic findings to reduce the need for unnecessary biopsies such as pleomorphic adenomas with the risk of transformation after incisional biopsy of pleomorphic adenomas not followed by

total surgical resection.[7,8,9]

The diagnosis cannot be determined before a pathological examination is carried out. The diagnosis can be neoplasm, pseudotumor, hemangioma, lymphoma, sarcoma or metastatic process. The lacrimal and salivary glands have similar histological and pathological structures. Neoplastic cells can take various morphological forms such as spindle, epithelioid, plasmacytoid or net. A case study conducted by Mahdi et al. found a highly cellular tumor consisting of spindle-shaped cells resembling a sarcoma. Tumor cells are often arranged in a solid, trabecular or reticular pattern. Central necrosis and pseudocyst formation may occur. Infiltrative and destructive growth is the main histological feature associated with malignant behavior.[6]

Anatomical, pathological examination in this patient showed a well-defined tumor mass with some parts appearing infiltrative into the surrounding stroma. The tumor cells are solidly arranged. Some are epithelioid with firmer boundaries, eosinophilic cytoplasm, round to spindle cell nuclei, and coarse granular chromatin. Some of the daughter nuclei are difficult to observe. Mitosis 3-4/10 HPF. There is also cystic degeneration of the tumor. In some parts, some glands show precursor lesions. These are consistent with the histomorphology of myoepithelial carcinoma.[10]

Further examination, namely immunohistochemistry, is needed to establish the diagnosis. MET is characterized by histologic patterns that myoepithelial cells assume, such as epithelioid, clear, hyaline (plasmacytoid), spindle, and mixed, with most neoplasms having two or more patterns.[10] Diagnosis requires reactivity to cytokeratin and at least one myoepithelial marker: Smooth Muscle Actin (SMA), glial fibrillary acidic protein (GFAP), calponin, protein S-100, p63 and CK 5/6. Some case studies were said not to have been carried out, and no genetic study examination was required. The patient in this case report underwent immunohistochemical analysis of his tumor tissue with the results of CK being diffusely stained on tumor cells and SMA being stained weakly on some tumor cells (focal). It was concluded that the outward pattern in this patient indicated that the tumor cells were of epithelial origin (carcinoma).[6]

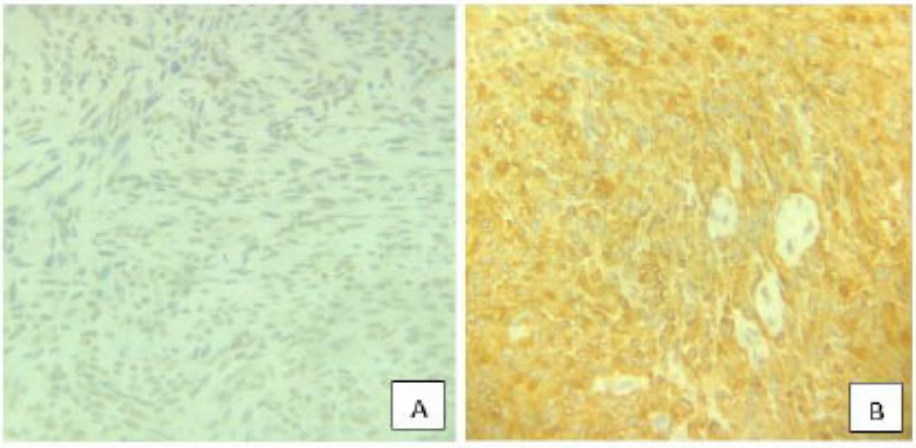

Figure 8. Immunohistochemistry of SMA (A). Immunohistochemistry of cytokeratin appearance (B).

Management such as resection is the primary therapy in cases of myoepithelial carcinoma. The type of resection can be wide local excision or orbital exenteration. This patient has a mass size of 11x10x5 cm, and an eyeball that is no longer functioning properly has caused erosion of the orbital bone and has experienced distant metastases (T4bN0M1), so that orbital exenteration is considered followed by radiotherapy to prevent perineural worsening and extension. Esmaeli et al. mentioned that lacrimal gland carcinomas measuring less than 2.5 cm could be regarded as tumor excision followed by radiation therapy to maintain the eyeball and vision.[1,11,12,13,14]

The recurrence and mortality rates have been high in patients with lacrimal myoepithelial carcinoma. More case reports are needed to characterize better primary lacrimal myoepithelial carcinoma. Complete immunohistochemical examination with positive epithelial and myoepithelial markers helps classify tumors of the poorly differentiated type.[6] The case report reported by Mahdi et al. mentioned that no distant metastases were found in a lacrimal gland epithelial carcinoma patient. Three cases were said to have local or locoregional recurrence diagnosed between 3 to 24 months. One of these three patients received palliative radiotherapy and died eight months after diagnosis. Two other patients died within months of being diagnosed without receiving definitive treatment. One patient had myoepithelial carcinoma arising from a pleomorphic adenoma.[6,7]

The lacrimal gland has few lymph nodes. The primary tumor that often metastasizes to the lacrimal gland is breast cancer. The incidence of metastases is infrequent and hematogenous. The scarcity of lacrimal gland tumors and many tumor types with different types of lesions make it difficult to determine optimal treatment, so the determination of regimens in randomized controlled trials is difficult. The many biological characteristics of lacrimal gland tumors provide the advantage that they are morphologically similar to tumors of another anatomy, e.g., with salivary glands and breast for epithelial lesions and lymph nodes for lymphoid tumors. The rapid development of non-lacrimal gland therapy provides more hope for treating patients with lacrimal gland tumors.[1,2,15,16]

CONCLUSION

A myoepithelial tumor (MET) is an uncommon epithelial neoplasm of the lacrimal gland. The many biological characteristics of lacrimal gland tumors provide the advantage that they are morphologically similar to tumors of another anatomy, such as with salivary glands and breast for epithelial lesions and lymph nodes for lymphoid tumors. These rapid developments in the treatment of non-lacrimal glands provide more hope for the treatment of patients with lacrimal

gland tumors. The scarcity of lacrimal gland tumors and many tumor types with different types of lesions make it difficult to determine optimal treatment, so the determination of regimens in randomized controlled trials is difficult.

DISCLOSURES

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Conflict of Interest
The authors declare no conflict of interest.

Consent for Publication
Written informed consent was obtained from the patient to publish this case report.

Author Contribution
All authors contributed equally in the writing of this article.

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