

## Tumors of the Lacrimal Sac: Three case reports

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### Abstract

Lacrimal Sac tumors are extremely rare and potentially life threatening. It may be primary, secondary and metastasis from distant organ. Lacrimal sac tumors may be broadly classified into epithelial (72%), mesenchymal (12%), lymphoproliferative (11%) and melanocytic (04%). About 72% are malignant; tend to be locally invasive with high recurrence rate. To report three cases of rare primary lacrimal sac tumors. We evaluated three rare case reports on primary lacrimal sac tumors in two tertiary eye hospitals in Bangladesh. 46 year old female, 36 year old male, and a 21-year-old young male, patients presented with mass in the lacrimal sac area. Two of them underwent deep incision biopsy and histopathology revealed Low grade extranodal marginal zone lymphoma (ENMZL). One patient was treated with Radiotherapy and another one with 6-Cycles of CHOP chemotherapy (Cyclophosphamide, Hydroxydaunorubicin, Oncovin, Prednisone). One patient underwent excision biopsy and histopathology reported lacrimal sac haemangiopericytoma. In all cases the lesions were resolved completely. Management of lacrimal sac tumors requires a high index of suspicion, as these are fatal tumor and often misdiagnosed as dacryocystitis. Early and appropriate intervention will help to complete resolution of the tumor as well as to reduce the recurrence.

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**Key words:** Dacryocystitis, extra nodal marginal zone lymphoma, haemangiopericytoma, radiotherapy

### Introduction

Lacrimal sac tumors are extremely rare and potentially life threatening. A case series by Shields et al, evaluated 1264 orbital tumors, of which only 2 originated from the lacrimal sac.<sup>1</sup> A French study that evaluated only 2 lacrimal sac tumors out of 1705 malignant tumors of eye & adnexal tumors.<sup>2</sup> It may be primary, secondary and metastasis from distant organ. Secondary tumors of the lacrimal sac may arise by invasion from local structures, including the nose, paranasal sinuses, orbit, conjunctiva, and eyelid tumors.

Metastasis from cutaneous melanoma, hepatocellular carcinoma, and even renal cell carcinoma have been described in the literature.<sup>3,4</sup> Given their rare occurrence, approximately 776 cases of the tumors of LDS in different case report studies and few large case series studies have been published in the literature worldwide from the 1930s to the present day. About 72% are malignant; tend to be locally invasive and high recurrence rate. Benign tumors tend to present in younger adults, where malignant tumors typically

occur in the fifth decade, with age ranges reported 22 to 94 years. Lacrimal sac tumors may be broadly classified into main four categories like epithelial (72%), mesenchymal (12%), lymphoproliferative (11%), and melanocytic (04%). Each category of Loews-Dietz Syndrome (LDS) tumors being further subdivided into benign and malignant. The majority of tumors are, however, primary and epithelial. Among the benign tumors, epithelial origin is 59.5% and

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mesenchymal 30.6%. Of the malignant tumors, close to 72% are of epithelial origin followed by 17% Lymphoma, 6.4% mesenchymal and 3.5% melanocytic. In order of frequency, benign lacrimal sac tumors include squamous papilloma, transitional papilloma, fibrous histiocytoma, oncocytoma, and haemangiopericytoma. Malignant tumors include squamous cell carcinoma, lymphoma, melanoma, transitional cell carcinoma, mucoepidermoid carcinoma and adenocarcinoma, with melanoma and transitional cell carcinoma being associated with high fatality rate.

Squamous and transitional cell papilloma which constituents of 285 of all lesions. The most common malignant epithelial tumor was squamous cell carcinoma which comprised 19% of all tumors.<sup>1-6</sup> We attempt to present three rare case reports on the primary tumors of the lacrimal sac.

#### Case 1:

A 46 years old female patient presented with painless firm, non-tender, mass in the left lacrimal sac area for 7 months duration. There was no abnormality in the examination of anterior segment and posterior segment of both eyes. A computed tomography (CT) scan of the orbit revealed a moderate enhancement homogeneous lesion involving the left lacrimal sac. The clinical and radiological diagnoses were squamous papilloma of the lacrimal sac. Deep incision biopsy was done. Histopathology revealed Low grade ENMZL which was treated with Radiotherapy. Lesion was resolved completely.

Modified dacryocystorhinostomy(DCR) was performed after six months of primary surgery. There was no recurrence within four years of follow up.



Figures 1-4: Case 1-a. Clinical Photograph of a female patient showing non-tender, firm mass in the left lacrimal sac area, b. Coronal view of CT scan of the orbit viewing the moderately enhanced well defined localized lesion in the left lacrimal sac area, c. Excised left lacrimal sac containing tumor, d. Histopathological microphotographs of ENMZ Lymphoma.

#### Case 2:

A 36 year old patient presented painful swelling in right medial canthal region of 2 months duration and treated with systemic antibiotic considering as acute exacerbation of dacryocystitis. On examination his BCVA was 6/6 in both eyes; anterior and posterior segment was unremarkable. The only ocular finding was firm, tender, nodular swelling in left lacrimal sac region. CT scan showed soft tissue mass in right Lacrimal Sac region without any bone erosion.

As there was dilemma in diagnosis, DCR was deferred and deep incision biopsy from the lesion was done and histopathology and immunohistochemistry were confirmed the diagnosis ENMZL. There was no systemic involvement on evaluation of the patient. Patient was treated with 6-Cycles of CHOP chemotherapy and lesion was resolved completely. There was no recurrence was noted within one year follow up.

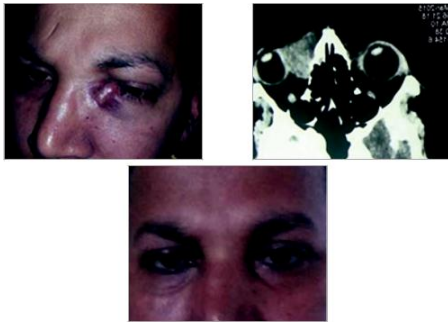


Figure 5-7: Case 2- a. Patients with nodular, firm, tender lesion in the left medial canthal area like as acute exacerbation of chronic dacryocystitis. b. Axial CT scan of the orbit showing soft tissue mass in right lacrimal sac region without any bone erosion. c. Resolved left lacrimal sac tumor after completion of treatment.

**Case 3:** A 21-year-old young male patient presented with a non-tender lesion in the right lacrimal sac area of 2 years' duration. Both eyes were normal functioning. There was no any abnormality in the systemic evaluation. CT scan of the orbit revealed mild enhancement homogeneous lesion in the right lacrimal sac. Clinical diagnosis was squamous papilloma in the lacrimal sac. Excision biopsy was performed and histopathology reported the diagnosis of the lacrimal sac lesion as a haemangiopericytoma.

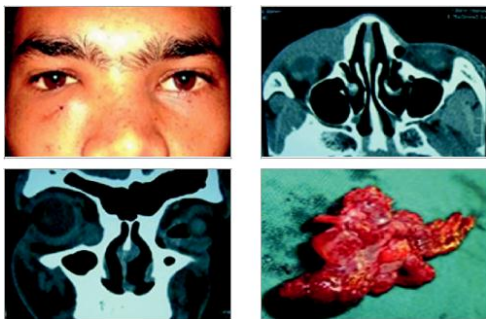


Figure 8-11: Case 3 - a. Young patient with a nontender lesion in the right lacrimal sac area, b-c. axial and coronal CT scan of the orbit showing mild enhanced soft tissue lesion involving the right lacrimal sac area, d. Excised tumor from the right lacrimal sac area.

## Discussion

Lacrimal sac tumours are rare; the majority are primary and of epithelial origin, some are non-epithelial.<sup>6,8,9</sup> Only 400 cases have been reported in the literature, with epithelial tumors to be the most common.<sup>8,9</sup> Non-epithelial mesenchymal tumors of Lacrimal sac are uncommon.<sup>8</sup> Benign lacrimal sac tumors include squamous papilloma, transitional papilloma, fibrous histiocytoma, oncocytoma, and hemangiopericytoma.<sup>6,9</sup> Previous studies showed, among the malignant tumors squamous cell carcinoma to be the most common, followed by lymphoma, melanoma, transitional cell carcinoma, mucoepidermoid carcinoma, and adenocarcinoma, with melanoma and transitional cell carcinoma being associated with high fatality rates.<sup>8,9</sup>

Early treatment of these tumors is necessary because they tend to be infiltrating tumors. Diagnosis of these tumors is often erroneous and delayed because they are confused with dacryocystitis.<sup>6,8,9</sup> The majority of lacrimal sac tumours present with symptoms typical of secondary acquired nasolacrimal obstruction, including epiphora with a medial canthal mass, and thus are often misdiagnosed as dacryocystitis.<sup>6,8,9</sup> Awareness of symptoms, high index of suspicion and proper clinical examination with imaging and biopsy are recommended for confirmation of diagnosis.<sup>8</sup> During DCR, the histological analysis of a lacrimal sac biopsy is essential for confirming the diagnosis and that may guide for adjuvant treatment.<sup>6,9</sup>

In all cases of lacrimal sac mass with epiphora, a complete workup should comprise a detailed history and a full ophthalmic exam with lacrimal probing, irrigation and examination of the nose. However it is clinically difficult to differentiate from chronic dacryocystitis. Imaging is therefore essential in diagnostic evaluation of lacrimal sac tumors and chronic inflammatory conditions, which can masquerade as neoplasms.<sup>9</sup>

CT scan of orbits and sinuses would show a lacrimal sac mass and the extent of bony erosion and invasion into surrounding structures. Both T1- and T2-weighted magnetic resonance imaging sequences with gadolinium contrast would provide a better tumour delineation and also providing information of involvement of neighboring soft tissue structures, including orbital fatty tissue.<sup>6,9</sup>

CT dacryocystography of the lacrimal drainage system would reveal a possible lacrimal sac filling defect due to a space-occupying lesion or delayed disappearance of contrast and show extent of obstruction. Radiological imaging is also valuable in postoperative follow-up.<sup>9</sup> Treatment of lacrimal sac tumors depends on the histological type, malignancy, size, extension and the patient's general health.<sup>6,9</sup> The aim of treatment is complete tumor removal. Wide resection is the treatment of choice for most malignant lacrimal sac tumors. Complete resection of the lacrimal drainage ducts is recommended, sacrificing the superior and inferior tear ducts, the lacrimal sac, the nasolacrimal duct, the lacrimal fossa, and the adjacent lacrimal ethmoid cells.<sup>6</sup>

Careful histopathological assessment in all cases is essential to confirm diagnosis and tumor type and thus plan for further treatment.<sup>9</sup> These tumors are potentially life-threatening due to proximity to vital structures and skull base.<sup>8</sup> Wide resection is the choice of treatment for most malignant lacrimal sac tumors.<sup>6</sup> Multidisciplinary management comprising wide surgical excision followed by radiotherapy and targeted adjuvant chemotherapy is optimal to prevent recurrences and metastases.<sup>8</sup> Lacrimal sac lymphoma should be suspected in any patient with systemic lymphoma with epiphora and/or mass above the medial canthal tendon. Treatment is usually a combination of surgery, irradiation, and/or chemotherapy, but no commonly agreed treatment regimen for periocular lymphoma exists because of the limited number of cases seen.<sup>9</sup>

In our present study, two cases were diagnosed as lacrimal sac ENMZL.

There was no systemic involvement in both patients. One patient was treated with Radiotherapy and another one with 6-cycles of CHOP chemotherapy and lesion was resolved completely. No recurrence was noted during the follow up period.

Radiotherapy can be the exclusive treatment when the tumor is deep-rooted, when surgery is refused by the patient or is contra-indicated. The immediate toxicity of radiotherapy is limited to conjunctival or cutaneous hyperemia. One of the risks of radiation is reduction in visual acuity, due to the involvement of optic nerve and the retina. A protective shell can be used to limit these risks.<sup>6</sup>

Recurrence and mortality rates for lacrimal sac tumors are varies. Benign tumours of the lacrimal sac have a good prognosis if completely excised but benign papillomas with inverted pattern have a tendency to recur 10 to 40%. The recurrence rate of invasive squamous cell and transitional cell carcinoma appears to be about 50%.

The malignant potential of hemangiopericytoma can be unpredictable. Lymphoid lesions respond to radiotherapy and chemotherapy and have a variable prognosis, depending on the extent of the disease and the type of tumor. The most terrible prognosis is that of malignant melanoma despite aggressive treatment.<sup>1,5-7</sup>

The best management for the suspected tumors confined to the lacrimal sac, is total excision of the tumor (Dacryocystectomy without osteotomy). A planned DCR or reconstruction will take place at a later date after histological confirmation. Adjuvant therapy (Radiotherapy and/or chemotherapy) can be an effective to reduce the rate of recurrence and for incomplete resection. Treatment should be decided within a multidisciplinary team and close, long-term monitoring is essential.<sup>6</sup>

## Conclusion

Successful management of lacrimal sac tumors requires the following- a high index of suspicion, as these are fatal tumor and often misdiagnosed as dacryocystitis. Early and appropriate aggressive intervention will help to complete resolution of the tumor as well as to reduce the recurrence. It is an important tool for the surgeon to carefully inspect the lacrimal sac during DCR to prevent missing a neoplasm. Careful long term follows up is required, because recurrence and/or metastases can be occurred many years after primary treatment.

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