Lacrimal Sac Squamous Cell Carcinoma: From Resection to Prosthetic Rehabilitation. A Case Report

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Abstract

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates. Diagnosis of these tumors is often delayed as they are confused with chronic dacryocystitis. There is a lack of consensus and evidence on standard treatment strategies for advanced lacrimal sac carcinomas. A case of advanced lacrimal sac squamous cell carcinoma treated with wide *en bloc* margin-negative surgical resection with further prosthetic rehabilitation without adjuvant therapy and 38 months of recurrence-free postoperative follow-up is presented.

Keywords: Lacrimal Sac Carcinoma, Squamous Cell Carcinoma, Facial Prosthetic Rehabilitation, Carcinoma En Block Resection, Magnabar Fixation

Key clinical message: There is a lack of consensus and evidence on treatment strategies for lacrimal sac carcinomas. Wide *en bloc*surgical resection with farther prosthetic rehabilitation could be the treatment option in certain cases.

Introduction

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates.[1-6] A total of 539 cases were reported in the literature from 1960 to 2019 among which squamous cell carcinoma, with 296 cases, was the most commonly described.[4]

The clinical presentation of lacrimal sac squamous cell carcinoma (LSSCC) resembles that of chronic dacryocystitis, which does not cause alarm until the appearance of specific symptoms, such as blood-stained tearsora, palpable lump or a progressive mass in the area of the lacrimal sac/nasolacrimal duct.[3,4,7] Thus, diagnosis is often delayed in LSSCC and in some instances, the diagnosis is missed even during routine dacryocystorhinostomy. In these cases, the tumor can grow into the adjacent sinuses and the nasal cavity and cause significant morbidity.[3] Fewer than 15% of cases of LSSCC are diagnosed within 2 months, and treatment is initiated within 12 months in 72% of patients.[1] MRI or CT scan of the orbit or paranasal sinuses is the preferred imaging modality to diagnose lacrimal sac tumors.[6]

Complete surgical excision followed by radiotherapy is the preferred modality of management, and only 18% require orbital exenteration.[5] Extensive surgical *en bloc* resection of lacrimal sac tumors with medial maxillectomy or total maxillectomy is favored with good success rates for local disease control. Orbit

exenteration, resection of the paranasal sinuses, or lymph node dissection is performed in certain advanced cases.[7-9] Lymph node status was found to be a key factor for prognosis.[10]

Because of the anatomic location of the lacrimal sac and nasolacrimal duct and their proximity to the orbital soft tissue, the maxilla and maxillary sinus, and the ethmoid bone and ethmoid sinuses, a multidisciplinary surgical approach is often optimal. Some practitioners may shy away from attempting globe-sparing surgery because of concerns about a higher risk of local recurrence if the eye is spared and because of concerns about ocular damage from radiation therapy.[3]

A case of advanced lacrimal sac squamous cell carcinoma treated with wide *en bloc* margin-negative surgical resection with further prosthetic rehabilitation but no adjuvant therapy and 38 months of recurrence free postoperative follow-up is presented.

Case history/Examination

A 30-year-old man was admitted to the Department of ENT and Maxillofacial Surgery with complaints of a slightly painful progressive mass in the area of the lacrimal sac. He presented a one-year history of unilateral epiphora in the right eye following acute ipsilateral inflammation (dacryocystitis) of the medial canthal region. The patient was treated with short-term systemic antibiotic therapy prescribed by the family doctor and then referred for specialized evaluation. On clinical examination right side extensive reddish bulging mass was revealed on the medial cantus region with lower and upper eyelid involvement (Figure 1). On palpation the mass was firm and moderately painful. An incisional biopsy of the mass was performed under local anesthesia and squamous cell carcinoma of the lacrimal sac was disclosed.

Differential diagnosis, investigations, and treatment

Contrast head and neck CT scan examination revealed a 2.5x 1.6x3.2 cm neoplastic formation with involvement of the lacrimal sac/duct, the medial part of the right orbit, the medial rectus muscle with very close adjustment of the eyeball without visible borders (Figure 2a) There was orbit inferior-medial bone wall and lacrimal bone resorption and invasion of neoplastic formation to the nasal cavity close to the medial nasal concha and maxillary sinus (Figure 2b) Right side carotid group lymph nodes slight enlargement was revealed with a size of 1.3x0.9 cm and submandibular lymph nodes with a size of 1.1x0.8 cm. The orbits contrast CT scan (DLP-1530.0 mg) did not reveal lesion invasion into the eyeball.

Fine needle aspiration of regional sentinel lymph node did not reveal metastatic involvement. Right-side wide *en bloc* resection with orbit exenteration including the ethmoid, lateral nasal bone, lacrimal sac and duct, and maxillary sinus upper wall without neck lymphodissection was performed (Figure 3a and b). The frontal musculocutaneous flap was used for lateral nasal defect closure (Figure 3 c).

The histological examination revealed cohesive atypical squamous cell tumor complexes composed of cells with moderately atypical nuclei and eosinophilic cytoplasm. Few foci of keratinization and necrosis were found. There were mitoses, including atypical ones. A moderately expressed mixed inflammatory infiltrate with a predominance of eosinophils was observed in the stroma (Figure 4). The tumor grew into the adjacent soft tissues and upper bone. There were no lymphovascular or perineural embolisms. The cutting edges were ablastic. The final histological diagnosis was lacrimal sac/duct squamous cell carcinoma G2 pT4a pNx LO VO PnO RO.

The post-exenteration wound healing was performed under iodoform gauze. It takes approximately one month for secondary intention wound healing under gauze.

During the first postoperative year, quarterly control head and neck contrast CT scan examinations were performed. No signs of recurrence were observed (Figure 5).

Therefore prosthetic rehabilitation was planned for the patient.

Under local anesthesia two dental implants were installed in the lateral zygoma (Figure 6). After four months healing caps were adopted through the small skin incision.

A silicone implant-supported epithesis was constructed for the patient with a right zygo-orbital defect for a total esthetic rehabilitation. The magnabar retention system was selected as the method of choice for the patient due to the depth and volume of the defect. The main magnet was situated on a casted framework bar at the lateral perimeter of the prosthetic field. The ocular was made of crystal clear acrylic (non-free-monomer methyl-met-acrylate, class 3 of ANNEX IX classification) in the scleral portion, and the iris was painted in conventional methods of iris painting used in ocular prosthetics to achieve a good esthetic result in comparison with the left eye (Figure 7a and b).

Silicone material of the orbital epithesis was made of a VTR platinum silicone of Technovent M511 with 25 Shore. Magnet keepers for the matrix magnet were made of a self-cure Vilacryl methacrylate. The silicone coloring system used in this epithesis was intrinsically pigmented silicone layering with negative painting of the stone cast.

Outcome and follow-up

The 38 month postoperative follow-up did not reveal signs of recurrence in the presented case.

Discussion

Primary epithelial malignancies of the lacrimal apparatus, comprising the lacrimal gland, duct, and sac, are extraordinarily and uncommon rare tumors with significant recurrence rates.[1-4,11-14] As noted by Singh and Ali (2021)[5] among primary malignant epithelial tumors, squamous cell carcinomas were the most common (61%), followed by transitional cell carcinomas (15%) and mucoepidermoid carcinoma (7%). Because of their rarity, no extensive clinical data on their management and prognosis exist.[7] Mortality rates for malignant tumors depend on tumor stage and type, and the mean rate is 38%.[1] Lacrimal drainage system carcinoma is a disease of the middle-aged and elderly population. Studies have reported that malignant lacrimal sac tumors often occur in the fifth decade with a slight male predominance.[4,10,13,15] Symptoms of lacrimal sac carcinoma might resemble those of benign disease of the lacrimal drainage system. In combination with the rarity of these tumors, there is often a prolonged diagnostic latency as the diagnosis of these tumors is often delayed because they are confused with dacryocystitis.[1,4,6,7,12,13] Swelling in the medial canthal region, epiphora and pain are the most common presenting features.[6,13] LSSCC can involve the lacrimal sac and grow through the nasolacrimal duct to invade its peripheral organs and structures.[7] These patients are often referred later after the discovery of a malignant tumor on biopsies of the lacrimal sac taken when dacryocystitis recurs.[1,16]

In the presented case the 30 -year-old male patient had a one year history of right eye epiphora and redness and swelling in the medial cantus area for the preceding 5 months. Dacryocystitis was diagnosed by the family doctor and antibiotics were prescribed. After three months of unsuccessful conservative treatment the patient was directed to hospital treatment where the deep incisional biopsy was performed and a diagnosis of LSSCC was clarified.

Thorough clinical workup and computed tomographic-dacryocystography, computed tomography (CT) scan of the orbit, or magnetic resonance dacryocystography can help in diagnosing a lacrimal sac tumor. Imaging is essential for identifying the location, size, and extent of the lesion, assessing the disease severity, and differentiating tumors from inflammatory and infectious lesions.[17] CT scans of the orbit or paranasal sinuses with axial, coronal, or sagittal images are used to diagnose lacrimal sac tumors and to assess osteolytic changes as well as the invaded surrounding tissues.[3,6,11]

Kumar VA with coauthors recommends performing a thin-section (1.25 mm) CT with contrast as the first-line imaging study to evaluate malignant lacrimal sac and nasolacrimal duct tumors at the time of initial staging. MR imaging can be performed if CT cannot distinguish sinonasal tumor extension from postobstructive secretions.[11]

In the present study head, neck and chest CT scans with contrast and isolated CT scans of orbits were performed.

Correct diagnosis and appropriate therapy require a multidisciplinary management approach. First and

foremost in the treatment of these malignant epithelial tumors is complete surgical removal with wide excision.[1,12]

However, there is no standard surgical treatment strategy. Multidisciplinary therapy, including surgery, chemotherapy, and radiotherapy, is the primary treatment modality[3,4]. Extensive surgical *en bloc* resection of lacrimal sac tumors with medial maxillectomy or total maxillectomy is favored with good success rates for local disease control.[7,9] Orbit exenteration, resection of the paranasal sinuses, or lymph node dissection is performed in certain advanced cases.[8,18]Song et al.[10] reported that the outcomes of comprehensive treatment were quite encouraging, and the 5-year overall survival rate and 5 year progression-free survival rate were 87.6±4.8% and 76.3±6.4%, respectively.

The combined sinus-orbit approach is an effective method of managing lacrimal sac tumors to achieve optimal tumor clearance from the orbit and nasal cavity.[9]

Decreased recurrence rates were observed in patients who underwent lateral rhinostomy and wide excision compared to those without rhinostomy.[6] Aggressive malignant lesions may require the removal of the entire lacrimal drainage system, including the canaliculi, lacrimal sac, and entire nasolacrimal duct with lateral rhinotomy.[5,6] Orbital exenteration and resection of the paranasal sinus may be needed for extensive primary or secondary malignant lesions.[6,8,12,19] The balance of surgical radicality and preserving quality of life is similar to trade off thinking for these cases.[14] In the presented case the neoplastic lesion was invaded the medial rectus orbital muscle, ethmoid, nasal cavity, medial concha and maxillary sinus. Wide *en bloc*surgical removal could be more life-lengthening for this case than organ-preserving operation tactics and radiation therapy. Notably, lymph node status was a key factor in determining outcomes.[1,7]

Radiation therapy has been considered an alternative to surgery, but there is no consensus on its use for advanced lacrimal sac carcinoma. Song et al.[7] in their study of 17 cases concluded that radiation therapy alone achieved excellent long-term clinical outcomes and could be a viable treatment option for patients who refused surgery or had unresectable tumors. There is also no high-quality evidence on the use of chemotherapy in advanced lacrimal sac carcinoma to date. In sum, there is a lack of consensus and evidence on standard treatment strategies for advanced lacrimal sac carcinoma.[14]Adjuvant treatment modalities include external beam radiation therapy, local radiation therapy (plaque brachytherapy), chemotherapy (CHOP regimen) or immunotherapy.[6,20]

Recurrence and mortality rates for lacrimal sac tumors vary from case to case.[3,4,6-10,16,18,20] In the present study the patient did not receive any postoperative adjuvant treatment and no recurrence was revealed during 38 months of follow-up.

To our knowledge this is the first report of a case of facial prosthetic rehabilitation after advanced lacrimal sac squamous cell carcinoma wide*en bloc* resection without adjuvant therapy.

Conclusion

Malignant epithelial lacrimal sac tumors are rare cancers with high recurrence rates. Diagnosis of these tumors is often delayed because they are confused with dacryocystitis. Multidisciplinary therapy, including surgery, chemotherapy, and radiotherapy, is the primary treatment modality. Wide surgical *en block* resection with orbit exenteration is recommended as a surgical tactic for advanced cases. Facial prosthetic rehabilitation could be an effective method in the recurrence free postoperative period.

Author contributions

Anna Yu. Poghosyan—drafted the manuscript for important intellectual content and implemented the clinical work.

Armine F. Gharaqeshishyan –implemented the clinical work, approved the version to be published

Martin S. Misakyan—made substantial contributions to the concept and design of the work. David K. Minasyan—was responsible for performing an accurate literature review and implementing the clinical work.

Parandzem S. Khachatryan—made contributions to the design of the histological examinations and was responsible for the histological data, design and literature review.

Artavazd Kharazyan- drafted the manuscript for intellectual content and implemented the prosthetic clinical work.

Karen Mashinyan-made contribution to the design of prosthetic rehabilitation

Sergo Hovhanisyan- was accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work were appropriately investigated and resolved.

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Figure legends

Figure 1 . External view of the patient's face—extensive reddish bulging mass on the medial cantus region with the lower eyelid involvement

Figure 2 . Post-contrast -enhanced CT shows tumor extension from the lacrimal sac/duct into the medial canthus, medial rectus muscle, ethmoid, nasal cavity, medial concha and maxillary sinus: axial view (a), coronal view (b)

Figure 3. Operating field after *en bloc* resection (a), macroscopic view of resected block from outside (b), lateral nasal defect closure with frontal musculocutaneous flap (c)

Figure 4. Nests of squamous cell carcinoma (blue arrow) with surrounding inflammation (yellow arrow) ×40, H@E

Figure 5. Coronal CT scan of patient after one year follow-up

Figure 6. Two dental implants were installed in the lateral zygoma.

Figure 7. External view of the patient's face: magnabar retention system fixed on implants (a), a silicone implant-supported epithesis fixed on a magnabar retention system (b)