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Squamous Cell Carcinoma of the Lacrimal Sac: A Case and Literature Review

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Authors' contributions

This work was carried out in collaboration between all authors. Author DR designed the study, wrote the protocol, and wrote the first draft of the manuscript. Authors BC, BM, ÖS and AI managed the literature searches, analyses of the study performed the spectroscopy analysis all authors read and approved the final manuscript.

Case Study

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ABSTRACT

Lacrimal sac squamous cell carcinoma is rare and often presents with unspecific symptoms of associated chronic dacryostenosis or dacryocystitis and may be falsely diagnosed as dacryocystitis. A chronically ill looking 73-year-oldcaucasian, who is a house wife, presented with excessive lacrimation in the right eye of 15 months duration and a right inner canthal mass of 6 months duration. The finding of a painless 2cm firm rubbery and smooth mass in the right medial canthus in addition to the symptoms prompted a diagnosis of chronic dacryocystitis and antibiotic therapy was initiated. However, there was no improvement with therapy. Further investigations, including magnetic resonance imaging (MR) were performed. Although the imaging suggested an inflammatory or infectious etiology, because of the suspicion of the malignancy, complete excision of the tumour, periosteum of the lacrimal fossa and nasolacrimal duct was performed via a medial orbitotomy. Histopathological examination revealed infiltrative well-differentiated highly keratinized squamous cell carcinoma arising from lacrimal sac. Herein, we report a case of squamous cell carcinoma of the lacrimal sac mimicking chronic dacryocystitis.

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1. INTRODUCTION

The lacrimal sac is a membranous structure that is derived embryologically from ectoderm. It serves as a temporary reservoir for the collection of tears before transmitting them into the nasolacrimal duct to the nose [1]. The lacrimal sac may be involved by inflammatory (or infectious) processes that give origin to acute or chronic dacryocystitis. Malignant lacrimal tumors are rare and occasionally arise in the lacrimal sac [2]. Malignant neoplasms, which occasionally arise in the lacrimal sac, include squamous cell carcinoma, and mucoepidermoid carcinoma [3].

The clinical presentation of a lacrimal sac neoplasm is similar to that of chronic dacryocystitis, however, lacrimal cell tumors often involve the area both below and above the medial canthal tendon, whereas in chronic dacryocystitis, the swelling is usually limited to the area inferior to the tendon. Patients can develop epiphora and a small medial canthal mass as the lesion expands. Spontaneous retrocanalicular bleeding associated with epiphora may be a finding of a malignant lacrimal sac tumor. It can also occur with inflammation [4]. A number of different malignant processes can involve the lacrimal sac, and polyps in this location can undergo malignant degeneration [2]. Carcinoma generally occurs in older patients and is associated with pain or epiphora or both. Deaths from metastasis of lacrimal sac tumors are uncommon and are usually due to local extension [4].

Squamous cell carcinoma can involve the lacrimal sac or the canaliculi as either a primary tumor or as a result of contiguous spread from the eyelids, conjunctiva or sinuses [5,6].

The treatment of lacrimal sac neoplasms depends on the time of diagnosis (preoperative, intraoperative, or postoperative) and the nature of the tumor. Most cases are not suspected prior to surgery and are discovered either during or after performing a dacryocystorhinostomy (DCR). Patients with a bloody canalicular discharge should have an evaluation with computed tomography (CT), using bone windows, and MR prior to surgery. Some patients may have regional adenopathy involving the preauricular, submaxillary or cervical nodes. These later suspicious areas should be evaluated with fine-needle aspiration biopsy (FNAB) prior to surgery. Unlike chronic dacryocystitis, lacrimal sac malignancies often extend downward, with swelling developing above the medial canthal tendon. If a suspicious mass is noted at the time of a DCR, frozen-section biopsy should be performed.

The management of lacrimal sac tumor depends on its histology. Death is rarely seen. If a primary epithelial malignancy or melanoma is present and appropriate evaluations for contiguous extension (orbit, brain, and sinus imaging studies) and metastatic disease (chest and abdominal CT, complete blood count, lactate dehydrogenase, glutamyl transpeptidase, carcinoembryonic antigen [CEA], alkaline phosphatase) are negative, exenteration is sometimes indicated. Some tumors, however, can be controlled with wide excision and wide-field photon irradiation [6,7]. Occasionally, radical combined orbital-sinus exenteration with postoperative irradiation can be performed [8].

We report a case of squamous cell carcinoma (SCC) of the lacrimal sac mimicking chronic dacryocystitis.

2. CASE REPORT

A 73-year-old chronically ill looking caucasian, who is a house wife, is presented to us with excessive lacrimation in the right eye of 15 months duration and a right inner canthal mass of 6 months duration. Medical history was unremarkable and the patient had no history of previous ocular surgery. Physical examination revealed a painless 2cm firm rubbery and smooth mass in the right medial canthus not attaching to the underlying tissue or skin (Fig.1). It was observed that the nasolacrimal canal was occluded during irrigation. Visual acuity was 10/10 in both eyes. Ocular movements were intact and there was no sign of exophthalmos or diplopia. There was no bleeding from the nose or eyes but the side of the lesion was erythematous. Biomicroscopic examination findings were also normal. There was no sign of regional adenopathy involving the preauricular, submaxillary or cervical nodes.

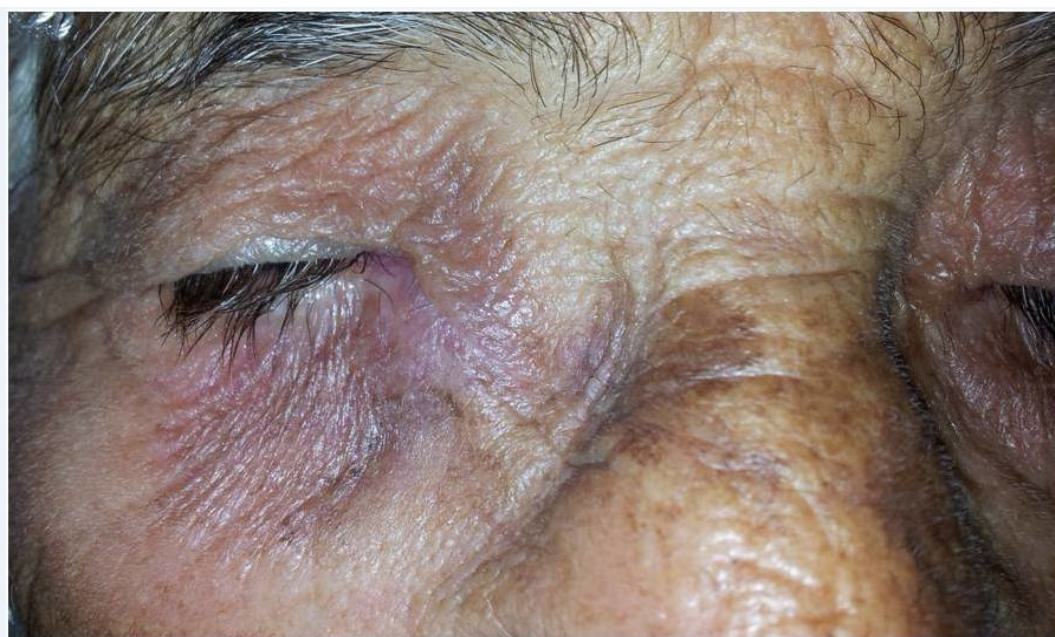


Fig. 1. A firm rubbery and smooth mass in the right medial canthus

With these clinical findings, an initial diagnosis of chronic dacryocystitis was made and antibiotic therapy was commenced. However, there was no improvement with therapy. MR imaging was therefore performed and a well-defined mass lesion in the left lacrimal sac extending to the nasolacrimal duct was detected. On T1-weighted images, the lesion was hypointense and enhanced minimally and heterogeneously after contrast administration. The lesion was hyperintense on T2-weighted images suggesting an inflammatory or infectious etiology. Orbital, brain, sinus imaging studies and systemic evaluations were performed for contiguous extensions and metastatic disease (chest and abdominal CT, complete blood count, lactate dehydrogenase, glutamyl transpeptidase, carcinoembryonic antigen [CEA], alkaline phosphatase). They were all negative. Complete excision of the tumour, periosteum of the lacrimal fossa and nasolacrimal duct was performed via a medial orbitotomy (Fig.2).



Fig. 2. Gross appearance of the lacrimal sac tumor

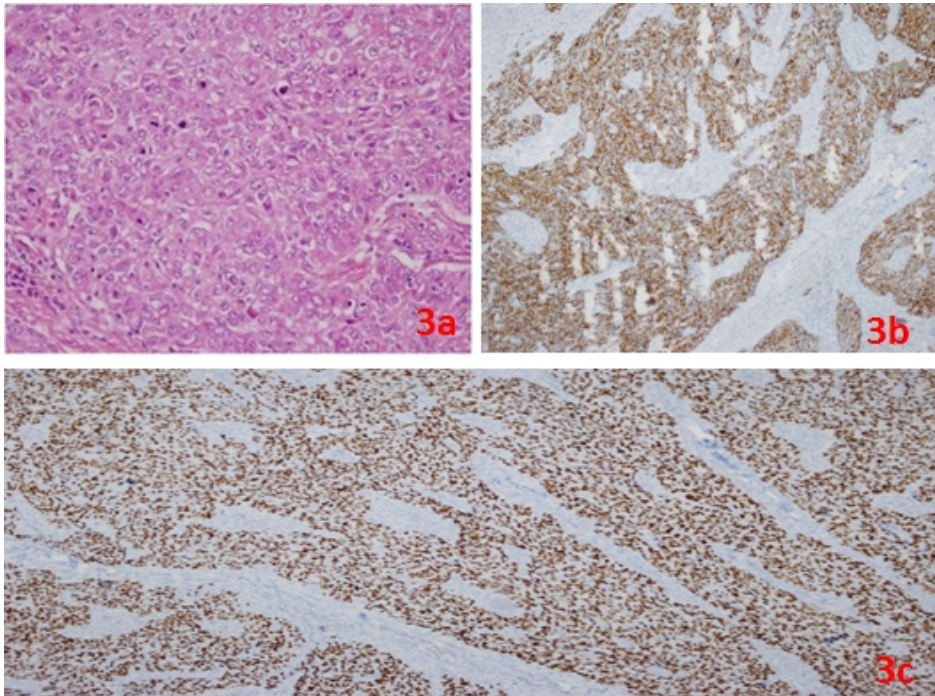


Fig. 3. Squamous cell carcinoma with atypical squamous cells within abundant eosinophilic cytoplasm, pleomorphic nuclei, prominent nucleoli

Histopathological examination of the specimens revealed infiltrative well-differentiated highly keratinizing SCC arising from the lacrimal sac. Pathological examination of the excised sac revealed squamous cell carcinoma with atypical squamous cells within abundant eosinophilic cytoplasm, pleomorphic nuclei, prominent nucleoli (Fig. 3a above). The specimens were stained with high molecular weight cytokeratin antibody and p63 (Figs. 3b and 3c above). The borders of the lesion were positive.

The patient did not accept postoperative radiotherapy . However, during postoperative follow up, no local recurrences were detected by orbital tomography at 6th, 12th and 24th months.

3. DISCUSSION

Malignant tumors of the lacrimal sac are rare. Approximately 400 cases have been reported in the literature, as case series or isolated cases [9,10]. Histopathologically these tumors can be categorized-as epithelial and nonepithelial types with primary epithelial tumors accounting for 75.6% of the cases [11-12].

Squamous cell carcinoma of the lacrimal sac occurs with equal frequency in both sexes and peaks during the fifth decade of life. Most lacrimal sac tumors present with nonspecific signs and symptoms including a slowly growing palpable mass in the medial orbital canthus and epiphora.

The clinical presentation of lacrimal sac tumors resembles that of chronic dacryocystitis or mucocele which also usually present as masses in the medial canthus. So the patients can be initially misdiagnosed as a case of dacryocystitis. These patients are often referred later after discovery of a malignant tumor on biopsies of the lacrimal sac taken when dacryocystitis recurs. Montalban et al reported that 2 of their 7 cases were diagnosed as dacryocystitis such as our patient [4]. The duration of symptom progression before diagnosis varies. For some, fewer than 15% of patients are diagnosed within 2 months. On the other hand, for 72% of the patients, treatment begins within 1 year [13]. The duration of the symptoms was 15 months in our case.

The most frequent symptom is tearing. Symptoms of regurgitation of blood-stained tears and epistaxis raise the possibility of underlying malignancy. The paranasal sinuses should be examined, because carcinomas from the maxilloethmoid complex may also present as medial canthal masses.

Squamous cell carcinoma of the lacrimal sac spreads mainly by direct invasion of the orbit, paranasal sinuses, and cranium. Metastases to lymph nodes are late occurrences and generally involve the preauricular, submandibular, jugo lodigastric, and cervical nodes. If the tumor is localized to the orbit at diagnosis, complete surgical excision before and/or after radiation therapy offers the best chance for survival [14].

In adults, the differential diagnoses of masses occurring in the medial orbital canthus include mainly inflammatory lesions and tumors [15]. Dacryocystitis (dilatation and inflammation of the lacrimal apparatus) is usually accompanied by signs of periorbital inflammation [15]. Pseudo tumor may occasionally present as a painful and isolated mass in the medial canthus. Characteristically, pseudotumor is of low signal intensity on T2-weighted MR images (true inflammation is of high signal intensity on long repetition-time/long-echo-time sequences [15]. Anterior ethmoidal mucoceles may also present as medial canthus lesions. MR or computed tomography shows the site of origin, benign bone expansion, and a rim of

enhancing mucosa, all of which suggest a benign process. Sarcoidosis involving the lacrimal sac is rare and is clinically and radiographically indistinguishable from other lesions [15]. The most common primary tumors of the lacrimal sac include squamous cell, transitional, and muco-epidermoid carcinomas. Melanoma, lymphoma, and various metastases have also been reported [15]. Primary tumors of the sinonasal cavities may also occasionally present initially as masses in the medial canthi.

In our case, the initial clinical diagnosis was dacryocystitis. As typical in patients with malignancies of the lacrimal sac, antibiotic treatment resulted without any improvement. MR imaging was performed and a well-defined mass lesion in the left lacrimal sac extending to the nasolacrimal duct was detected (Fig. 4). On T1-weighted images, the lesion was hypointense and enhanced minimally and heterogeneously after contrast administration. The lesion was hyperintense on T2-weighted images suggesting an inflammatory or infectious etiology. MR imaging of lacrimal tumors usually shows an intermediate signal lesion on T1-weighted sequences and a hypointense signal on T2-weighted images [4,16]. In some cases such as our case with hyperintense signal on T2-weighted sequences, MR images may not distinguish lacrimal sac tumors from dacryocystitis [4].

This lesion takes up the contrast agent moderately after gadolinium injection, sometimes heterogeneously. This imaging technique stages the tumor locally given that it differentiates the tumor from retained secretions and evaluates invasion of the adjacent structures as orbital fatty tissue, the extraocular muscles, the periorbita, and the meninges. It is also valuable in postoperative follow-up.

Treatment of malignant tumors of the lacrimal sac depends on the histological type, the tumor extension, and the patient's general health. In all cases, it is advised to analyze the tumor biopsy. Biopsies should be done deeply because chronic inflammation develops in the tumor periphery and can result in misdiagnosis of inflammatory pseudotumor. When necessary, wide resection is the choice treatment of most malignant lacrimal sac tumors [17]. Where epithelial and mesenchymal tumours are localized to the lacrimal sac fossa, intact excision of the tumour (and the periosteum of the fossa and as much of the nasolacrimal duct as possible) may be performed through a dacryocystorhinostomy incision [2-8]. 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 cells¹⁸ We also tried to remove the lacrimal sac, nasolacrimal duct, lacrimal fossa, and adjacent lacrimal ethmoid cells. Orbital exenteration is recommended in cases of major invasion of the orbital cone or of tumor recurrence after adjuvant radiotherapy. Resection of the adjacent orbital and lateral nasal wall may be required with more extensive tumours, it may be necessary to perform orbital exenteration, resection of paranasal sinuses, or cervical lymph node dissection [15].

Postoperative radiotherapy is recommended for malignant epithelial tumours, although irradiation of benign papillomas might induce malignant transformation. Recurrent lesions may be treated with further surgery or radiotherapy [15]. Radiotherapy can be the first treatment when the tumor is deeply located or when surgery is refused by the patient or is contra-indicated [12]. The immediate toxicity of radiotherapy is low and is limited to conjunctival or cutaneous hyperemia. One of the risks of radiation is reduction in visual acuity, which can occur with doses of 50 Gy delivered on the optic nerve or the retina. Delayed toxicity leads to xerophthalmia, radiation retinopathy, glaucoma, cataract, or keratitis with corneal ulcerations [12,17]. A protective shell can be used to limit these risks.

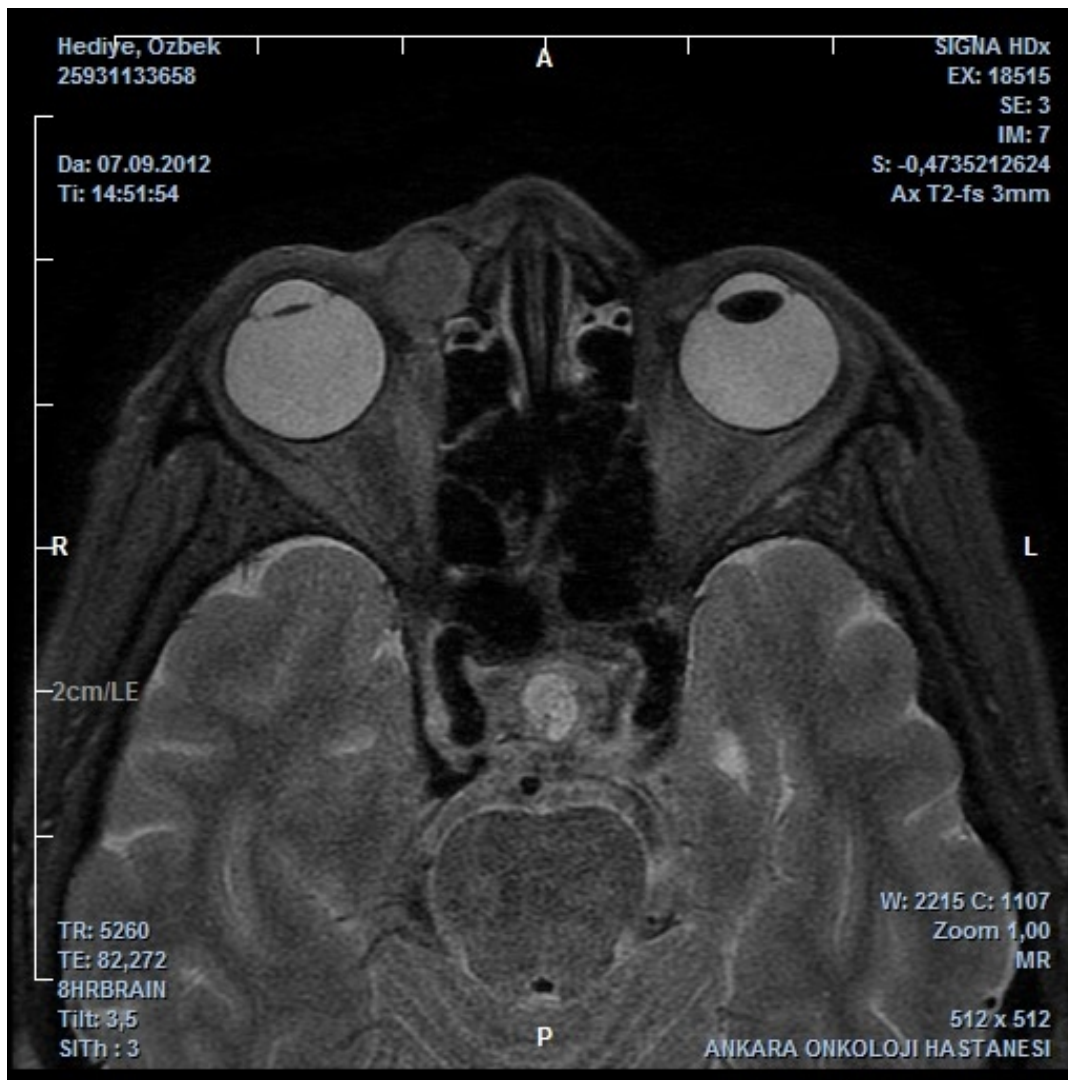


Fig. 4. Magnetic resonance image of the lacrimal sac tumor

Recurrences most often occur locally or reveal lately. For Ni et al, the recurrence rate is directly related to initial tumor excision, with 12.5% recurrence for wide excision versus 43.7% recurrence in cases of isolated lacrimal sac resection [13].

Metastases are rare and most frequently to the lungs. In the series reported by Ni et al. comprising 74 malignant tumors, 28% of the patients presented with lymph node invasion and six presented with metastases (four to the lungs and two to the esophagus) [13]. Fortunately, there was no sign of metastasis according to orbital, brain, sinus imaging studies and systemic evaluations in our study. Orbital and sinus CT is important when tumour is suspected and will provide evidence of expansion or erosion of the lacrimal sac fossa, or invasion into neighbouring structures. Systemic evaluation, both clinically and with chest X-ray and haematological tests, should be undertaken and a diagnostic tissue biopsy is necessary prior to definitive treatment.

4. CONCLUSION

In conclusion, it should be remembered that malignant tumors of the lacrimal sac are rare and often initially diagnosed as dacryocystitis because of the common clinical features. In order to prevent misdiagnosis, a lacrimal sac tumor should be suspected in patients presenting with chronic dacryocystitis and MR imaging should be performed. However, it should also be kept in mind that in some cases MR imaging may not distinguish lacrimal sac tumors from dacryocystitis, so in case of strong suspicion of malignancy surgical excision and histopathological examination should be the approach. Treatment should be decided by a multidisciplinary team and close, long-term monitoring is indispensable.

CONSENT

Informed consent was obtained from the individual Medical Officer who participated in the study.

ETHICAL APPROVAL

The study was approved by the Federal Medical Centre ethics committee and have therefore; been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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