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Clinical Characteristics of Lacrimal Sac Tumors: Report of Ten Cases

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Background and Objectives: To review our experience with lacrimal sac tumors in an effort to identify features, to evaluate the results of various methods of management, and to compare our data with previous studies.

Methods: We reviewed the medical records of all patients with lacrimal sac tumors who were managed in our institution between January 1990 and December 2015. The pre-operative clinical data, imaging, operation notes, and follow-up records were reviewed for each patient.

Results: The study group consisted of four men and six women with a mean age of 47.6 years. Most patients experienced long-standing epiphora, for a mean period of 20 months. Two of the tumors were benign, and eight of them were malignant. The benign tumors were treated with dacryocystectomy. All but one malignant tumor were treated with medial or total maxillectomy. Adjuvant radiotherapy was administered to four patients with malignant tumors. In the eight patients with malignant tumors, the mean follow-up period was 65 months.

Conclusions: Important characteristics of lacrimal sac tumors include dacryocystitis, epiphora, and in some cases, a palpable medial canthal area mass. Wide en bloc resection via medial or total maxillectomy and/or postoperative radiotherapy are proper treatments for malignant lesions of the lacrimal sac.

KEY WORDS: Nasolacrimal system · Lacrimal sac tumor · Lacrimal sac carcinoma.

INTRODUCTION

Tumors of the lacrimal sac are rare. Approximately 700 cases of lacrimal sac tumors have been reported in the literature.[1-7] Stefanyszyn *et al*. reviewed six series of lacrimal sac tumors, which included 255 cases, and added 115 new cases in 1993.[5] Since the report of Stefanyszyn *et al*., there has been only two series report of 15 and 10 lacrimal sac tumors.[8)9)] Thus, only limited information for this disease entity is available.

Symptoms of lacrimal sac tumors are not easily distinguishable from chronic dacryocystitis or dacryostenosis. Commonly tumors of the lacrimal sac are detected inadvertently at the time of dacryocystorhinostomy for presumed dacryocystitis.[5] Previous studies have reported a high malignancy rate for lacrimal sac tumors.[1-3)5)10)] All these factors, including a lack of information due to the rarity of lacrimal sac tumors, clinical similarity with primary obstruction of the nasolacrimal system, and high malignancy rate, make lacrimal sac tumors life-threatening, and the early diagnosis and management difficult.

Even though rhinologists are specialists who are familiar with the anatomy around the lacrimal sac and duct, there are few studies involving lacrimal sac tumors authored by rhinologists. The purposes of this study were to review our experience with lacrimal sac tumors in an effort to identify

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Table 1. Clinical characteristics of lacrimal sac tumors

Case No.	Age/ gender	Symptoms (duration)	Prior DCR history (times)	Pathologic diagnosis	Treatment	Initial pathologic staging	Recurrence	Follow-up period (months)
1	39/F	Medial canthal mass (2 years)	1	Squamous papilloma	Dacryocystectomy	D (−), LN (−), DM (−)	(−)	239
2	57/M	Epiphora (3 years), Medial canthal Mass (1 year)	1	Squamous cell carcinoma	Dacryocystectomy and wide excision	D (−), LN (−), DM (−)	Local recurrence after 6 months and lung metastasis after 15 months	Died at 18 months
3	40/M	Epiphora (1 year)	10	Squamous cell carcinoma	Total maxillectomy with orbital exenteration	D (+, Maxillary sinus, orbit), LN (−), DM (−)	(−)	204
4	26/F	Epiphora (2 months), Medial canthal mass (2 months)	0	Squamous cell carcinoma	Medial maxillectomy	D (−), LN (−), DM (−)	(−)	217
5	32/M	Epiphora (2 years), Medial canthal mass (1 year)	0	Solitary fibrous tumor	Dacryocystectomy	D (−), LN (−), DM (−)	(−)	181
6	55/M	Submandibular area mass (2 months)	0	Malignant melanoma	Medial maxillectomy with supraomohyoid neck dissection	D (+, Frontoethmoid, maxillary sinus), LN (+, Level Ib), DM (−)	Spinal metastasis after 6months	Died at 10 months
7	61/F	Epiphora (2 years)	3	Squamous cell carcinoma	Medial maxillectomy and radiotherapy	D (−), LN (−), DM (−)	(−)	143
8	65/F	Epiphora (1 year), Epistaxis (6 months)	0	Malignant melanoma	Medial maxillectomy and radiotherapy	D (−), LN (−), DM (−)	(−)	Loss after 3 months
9	47/F	Epiphora (5 years)	3	Undifferentiated carcinoma	Medial maxillectomy and radiotherapy	D (−), LN (−), DM (−)	(−)	22
10	50/F	Epiphora (1 year)	3	Squamous cell carcinoma	Medial maxillectomy and radiotherapy	D (−), LN (−), DM (−)	(−)	12

DCR: dacryocystorhinostomy, D: direct extension, LN: lymph node metastasis, DM: distance metastasis

characteristics essential for earlier diagnosis, to evaluate the result of various management methods, to compare our data with previous studies, and to determine the role of rhinologists in the treatment of lacrimal sac tumors.

MATERIALS AND METHODS

We reviewed the medical records of all patients with lacrimal sac tumors who were managed at our institute between January 1990 and December 2015. The study included patients with lacrimal sac tumors which were localized in the lacrimal sac. Patients who had tumors that invaded the lacrimal sac from other organs were excluded, as were patients with inflammatory pseudotumors.

Pre-operative clinical data, imaging studies, operative notes, and follow-up medical records, were reviewed for each patient. The collected data were tabulated and categorized according to the following categories: age, gender, symptoms and duration, prior history, primary tumor pathologic diagnosis, pre-operative imaging studies, initial treatment, staging, recurrence, and outcomes.

RESULTS

Between January 1990 and December 2015, only ten patients had lacrimal sac tumors (Table 1). The study group consisted of four men and six women ranging in age from 26−65 years (mean age, 47.6±12.9 years; median age, 50 years). One patient with benign squamous papilloma was 39 years of age at the time of diagnosis. Another patient with a benign solitary fibrous tumor was 32 years of age. The largest pathologic subgroup included five patients (two men and three women) with squamous cell carcinoma (Fig. 1A); the ages ranged from 26−65 years, with a mean age of 50 years. One 55-year-old man and one 65-year-old woman had malignant melanomas (Fig. 1B). One 48-year-old woman had undifferentiated carcinoma (Fig. 1C).

All patients, except a patient with a squamous papilloma, presented with an epiphora from two months to five years duration (mean duration, 20 months). Six patients had history of dacryocystorhinostomy for dacryostenosis. The average numbers of dacryocystorhinostomy was 3.5. Only one patient with malignant melanoma complained of epistaxis; she was the only patient who came to the otorhinolaryngologic department rather than the ophthalmologic department at the first visit. On physical examination, nine patients had a palpable mass over the lacrimal sac region. Among the nine patients, one had a squamous papilloma and two had squamous cell carcinomas with tenderness. One patient with malignant melanoma who did not complain of epistaxis presented with a mass in the submandibular gland area which had confirmed involvement with the submandibular lymph nodes. Six patients had a history of chronic dacryocystitis.

Eight patients had a pre-operative CT scan only, and two patients had a CT and MRI (Fig. 2). In most cases, the CT showed expansion or erosion of the lacrimal sac and invasion into the surrounding structures. For treatment, dacryocystectomy was performed for squamous papilloma and solitary fibrous tumors; one patient with a squamous cell carcinoma had a subsequent radical excision in the ophthalmologic department. An incisional biopsy was initially performed in seven patients with malignant tumors. Thereafter, medial maxillectomies were performed, as follows: three squamous cell carcinomas; one undifferentiated carcinoma; one malignant melanoma (Fig. 3). A total maxillectomy with orbital exenteration was performed on one patient with squamous cell carcinoma, and a medial maxillectomy

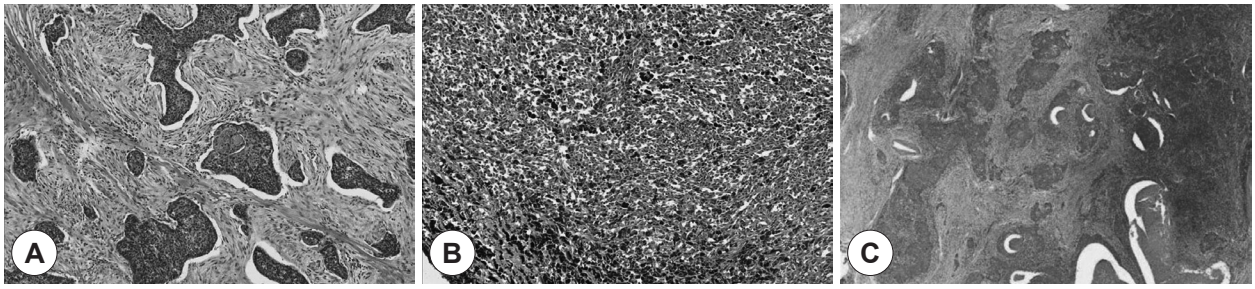

Fig. 1. Histopathologic findings of lacrimal sac tumors. (A) Squamous cell carcinoma (Case 10). The tumor shows a squamous nest containing keratin pearls and hyperchromatic cells with nuclear atypia (×100, H&E stain). (B) Malignant melanoma (Case 8). The tumor shows diffuse sheets of melanoma cells, with melanin pigments that show hyperchromatic nuclei and prominent nucleoli (×100, H&E stain). (C) Undifferentiated carcinoma (Case 9). The tumor shows a nested pattern with comedonecrosis without squamous or glandular differentiation (×40, H&E stain).

and supraomohyoid neck dissection was performed on one patient with a malignant melanoma. Post-operative radiotherapy was performed in four cases of malignant tumors.

In the eight cases of malignant tumors, the mean follow-up period was 65 months (median, 18 months; range 2−217 months). Five patients were alive and three patients with malignant melanomas and one patient with a squamous cell carcinoma died of metastatic disease.

DISCUSSION

Until now, approximately 700 lacrimal sac tumors have been reported worldwidely; herein we add ten more cases of lacrimal sac tumors and review the literature. This study is the first report of clinical characteristics of lacrimal sac tumors, and includes the largest number of cases in South Korea. The most common presenting symptoms for lacrimal sac tumors in our series were epiphora and a medial canthal area mass, as in previous studies.[7] Because of the rarity or similarity of lacrimal sac tumor symptoms with dacryocystitis, diagnosis is frequently delayed. In our series, six patients were diagnosed with dacryocystitis or dacryostenosis, all of whom underwent dacryocystorhinostomy at other institutions. The average duration of symptoms preoperatively was 20.8 years, with a range of a few months to five years. Therefore, a history of repeated dacryocystorhi-

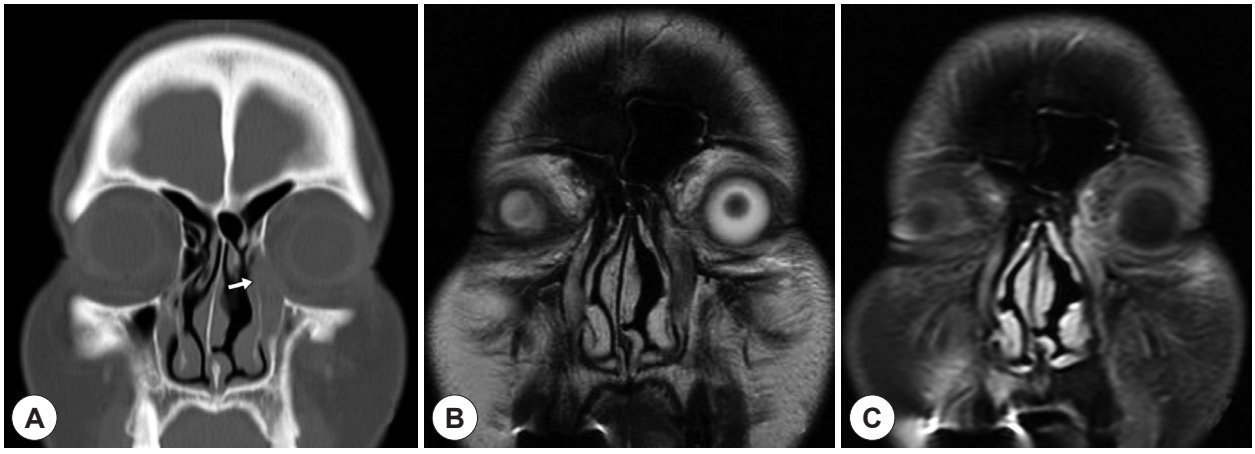

Fig. 2. Radiologic findings of the patient with undifferentiated carcinoma (Case 9). (A) Computed tomogram shows expansion of the lacrimal sac fossa with bony erosion (white arrow). (B) Magnetic resonance imaging shows a low signal intensity lesion on T2 weighted image, extending to upper nasolacrimal duct. (C) The mass shows heterogeneously enhancement on contrast enhanced T1 weighted image. These findings provide more accurate soft-tissue differentiation.

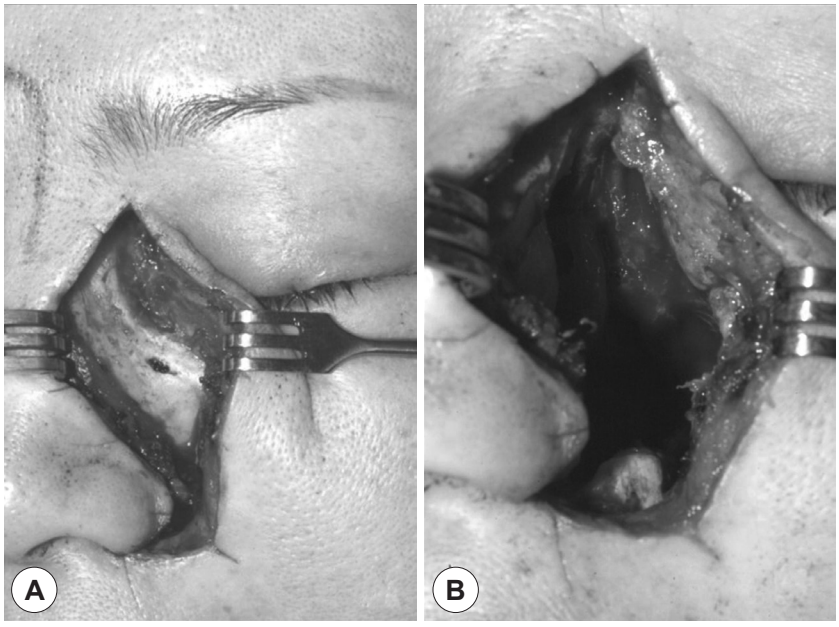

Fig. 3. Intra-operative findings (Case 9). (A) Exposure of the lesion was done with a lateral rhinotomy incision. (B) The tumor and surrounding structures were excise d with a medial maxillectomy.

nostomy and a mass in the medial canthal area could suggest a lacrimal sac tumor. Non-axial globe displacement occurred in one patient who was diagnosed with squamous cell carcinoma with orbital extension. Involvement of the regional (preauricular, submandibular, or cervical) lymph nodes occurs in the case of malignant melanomas.

The epithelium of the lacrimal drainage system changes from stratified squamous epithelium in the canaliculus to pseudostratified columnar epithelium in the lacrimal sac and nasolacrimal duct area. Primary epithelial neoplasms constitute 75% of all reported cases, with the remaining 25% of tumors non-epithelial.[5] In our cases, 70% of lacrimal duct tumors were epithelial neoplasms (one squamous papilloma, one undifferentiated carcinoma, and six squamous cell carcinomas). The non-epithelial tumors included two cases of malignant melanomas and one case solitary fibrous tumor. The ratio of malignant tumors amongst lacrimal sac tumors was reported to be 40−90% in a prior series.[5] In our series, the malignant ratio amongst lacrimal sac tumors was 80%.

An orbital or sinus CT is essential when the tumor is suspected and will provide evidence of expansion or erosion of the lacrimal sac fossa, or invasion into the surrounding structures.[4] When invasion into the surrounding structures is suspected, MRI can provide more accurate soft-tissue differentiation. CT and MRI could be considered complementary modalities in patients with suspected lacrimal duct tumors. Previous studies have shown that dacryocystography suggests the diagnosis by demonstrating irregular duct obstruction, however, it is not a specific finding and not helpful for staging.

For a definitive diagnosis, a tissue biopsy is mandatory. After diagnostic confirmation of the tumor is achieved, definitive treatment for lacrimal sac tumors consists of surgery, radiotherapy, and infrequently, chemotherapy.[7][8][11] For a benign tumor, dacryocystectomy is a good choice. However, even though a tumor is confined to the sac, dacryocystectomy may be not good choice of treatment for malignant tumors due to the possibility of extension.[8] Ni and colleagues showed that the results of excision and irradiation are inferior to excision and irradiation supplemented by a lateral rhinostomy.[3] Patients with a wide excision and lateral rhinostomy had a recurrence rate of 12.5%, in contrast to 43.7% of patients with localized lacrimal sac excision. One of our patients who was treated with dacryocystectomy for squamous cell carcinoma in the ophthalmologic department had a local recurrence within six months and metastasized to the lung in 15 months (Case 2). The primary resected specimen showed an involvement of resection margin. The recurred lesion with tenderness was developed on previous operation field. The patient refused further treatment and did not show up at the outpatient clinic. Among patients with malignant lacrimal sac tumors, all except the above-mentioned patients were treated by *en bloc* resection of the tumor by medial maxillectomy or total maxillectomy in our department. In advanced cases, it may be necessary to perform orbital exenteration, resection of paranasal sinuses, or lymph node dissection. For patients who has orbital invasion, orbital exenteration was performed with total maxillectomy, and for patients with malignant melanomas who presented with submandibular gland area masses, supraomohyoid neck dissection was performed with a medial maxillectomy. Adjuvant radiotherapy for malignant tumors in this lesion is often necessary to minimalize the risk of locoregional recurrence and to prolong the disease-free survival.[11] Post-operative radiotherapy was administered to five patients with malignant lacrimal sac tumors in our series. Pre- and post-operative radiation therapy in the range of 3,000−5,000 cGy is recommended for malignant epithelial tumors of the lacrimal sac.

Prognosis of patients with malignant lacrimal sac tumors was relatively poor. Ni and colleagues in a series of 71 epithelial malignant lacrimal sac tumors, reported an overall mortality of 37.5% in patients treated with a combination of wide surgical excision and radiation.[3] Lymphatic spread occurred in 27% and hematogenous metastases to the lung or the esophagus developed in 9.1% of patients. A general recurrence rate of 50% has been reported for carcinoma of the lacrimal sac, of which 50% are fatal. There is no standard treatment for recurrent lacrimal sac tumors; therefore, according to previous studies, local recurrences were treated in a case-by-case manner with surgical resection and/or radiotherapy.[8][9][11] Due to its rarity, a staging system for lacrimal sac malignancies, excluding lymphoma, has not been established. Only case reports or studies analyzing prognosis using the staging manual of lacrimal gland tumor from American Joint Committee of Cancer (7th edition) were reported.[12] However, the manual clearly states for the staging manual not to be applied to malignancies of the lacrimal sac and drainage apparatus. Therefore, a new reliable staging classification for lacrimal sac malignancies is necessary. In our series, three of eight malignant lacrimal sac tumor

patients died of the disease; two patients had malignant melanomas and one patient had a squamous cell carcinoma. On the other hand, four patients with squamous cell carcinomas who were treated with medial or total maxillectomy and adjuvant radiotherapy were all alive without disease.

CONCLUSION

Even though lacrimal sac tumors are rare, the early diagnosis and management of lacrimal sac tumors is important due to the high ratio of malignancy, and therefore, a high rate of mortality and morbidity. A vague suggestion for lacrimal sac tumors is repeated dacryocystitis and epiphora, and sometimes medial canthal area palpable masses. The high ratio of malignant tumors needs preparation for definite treatment before incisional biopsy. We concluded that wide surgical *en bloc* resection via medial or total maxillectomy and/or post-operative radiotherapy are appropriate for malignant lesions of the lacrimal sac to improve patient survival.

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