Case Report

Transitional Cell Carcinoma of the Lacrimal Sac

Passorn Preechawai MD*,
Robert C Della Rocca MD, FRCO**, David Della Rocca MD***,
Steven Schaefer MD****, Steven McCormack MD*****

** Department of Ophthalmology, Faculty of Medicine, Prince of Songkla University, Had Yai, Songkhla*
*** Department of Ophthalmic Plastic and Reconstructive Surgery, St. Luke-Roosevelt Hospital, New York, and The Department of Ophthalmic Plastic, Orbital and Reconstructive Surgery, The New York Eye and Ear Infirmary, New York*
**** The New York Eye and Ear Infirmary, New York*
***** Department of Otolaryngology, Head and Neck Surgery, The New York Eye and Ear Infirmary, New York*
****** Department of Pathology, The New York Eye and Ear Infirmary, New York*

Lacrimal sac tumor is a rare tumor, for which we do not know the exact incidence. Malignant tumors account for 70% of all cases. Epithelial tumors are the most common pathological type and squamous cell carcinoma is the most common of these, a transitional cell carcinoma is rare and has a poor prognosis. The authors report a rare case of transitional cell carcinoma of the lacrimal sac. A 57-year-old woman had tearing on her right eye. The pre-operative diagnosis was possible nasolacrimal duct obstruction with signs of dacryocele. Intraoperatively the authors found a lacrimal mass and a frozen section showed squamous papilloma with dysplasia. The tumor was completely removed and DCR was done. The subsequent pathological report of the right lacrimal sac was papillary transitional cell carcinoma, so medial maxillectomy and resection of the medial inferior orbit with ethmoidectomy were performed. She received radiation and has been tumor-free for 2 years.

Keywords: *Lacrimal sac tumor, Transitional cell carcinoma, Malignant lacrimal sac tumor*

J Med Assoc Thai 2005; 88 (Suppl 9): S138-42
Full text. e-Journal: *http://www.medassocthai.org/journal*

Lacrimal sac tumor is a rare tumor but is potentially life-threatening, and it can devastate the visual function, especially in cases of delayed diagnosis and treatment. There have been some 400 case reports in the literatures since 1937-2003[1-12]. The first large review series was undertaken by Duke-Elder in 1952[2]. He found 91 cases of true neoplasm, with 73 malignant cases (80%). A second large series by Radnot and Gall[3] in 1966 reviewed 138 neoplasms with 110 malignancies (80%). Schenck[4] et al. and Ryan[5] et al reviewed 25 and 27 cases respectively in 1973, finding 20 (80%), and 16 (60%) malignancies, respectively. Flanagan[6] 1978 and Hornblass[7] 1980 reported 5 cases, each with malignancy in half of their cases. The next large series in 1982 was from China, in which Ni[8] reported 82 cases, with the highest percentage of malignancies, 74 (90%). Stefanyszyn[9] reported 115 cases in 1994 with 59 malignancies (51%). The most recent known series was done by Parmer[10] in 2003, 15 cases with 87% (13 cases) malignant.

The epithelial lining in the lacrimal sac is similar to the lining in the upper respiratory tract, a pseudostratified columnar epithelium. Therefore, neoplasms arise as papilloma, squamous cell carcinoma, transitional cell carcinoma or adenocarcinoma. However, other non-epithelial tumors may arise from the lacrimal sac, such as malignant melanoma, lymphoma, leukemia, hemangiopericytoma, fibrous histiocytoma, oncocytic adenocarcinoma, etc. Primary epithelial neoplasm is the most common tumor of the lacrimal sac and accounts for 73% of all reported cases. Transitional cell carcinoma is a rare epithelial tumor, unlike squamous cell carcinoma. Herein the authors report a

Correspondence to : Preechawai P, Department of Ophthalmology, Faculty of Medicine, Prince of Songkla University, Had Yai, Songkhla 90110, Thailand.

case of transitional cell carcinoma of the lacrimal sac and treatment option for the patient.

Case Report

A 57-year-old woman had first noticed tearing in her right eye in September 2001. She received anti-biotics and steroids at that time, but the symptom did not improve. She was referred to the authors in July 2002. She had no other medical problems. Her vision was 20/20 in both her eyes without correction. She had swelling on her right lacrimal sac and obstruction of the lacrimal passage. She was given a CT scan to rule out mucocele. The CT scan revealed dilatation of the right lacrimal sac with a focal mass effect and slight remodeling of the junction of the posterior crest of the right lacrimal bone with adjacent ethmoid labyrinth. There was no evidence of mass lesion, calculus or osseous remodeling or destruction along the remaining course of the right nasolacrimal duct. No masses were apparent in the right inferior meatus (Fig. 1). She was diagnosed dacryocystitis with nasolacrimal duct obstruction OD. She had an appointment for external dacryocystorhinostomy with silastic intubations but she was lost to follow up after she was diagnosed as having colon cancer. She came back one and a half years later with persistent tearing and a mass at the medial canthal area. The pre-operative diagnosis was possible nasolacrimal duct obstruction with possible dacryocele. At the time of operation, the authors found an orbital mass somewhat infiltrative in character. Frozen section specimens were sent and the preliminary diagnosis was squamous papilloma with dysplasia. Further dissection and removal of the tumor was completed. The nasolacrimal tissue was taken to biopsy and frozen section showed that it was mucosa. The lacrimal bone was intact following the resection. Intubation of the lacrimal system was completed. The pathological report of the right lacrimal sac was papillary transitional cell carcinoma with frequent mitoses and focal invasion into the soft tissues (Fig. 2, 3). The right nasal mucosa was fibrotic with chronic inflammation. She was referred to an otolaryngologist for joint management. Upon examination the otolaryngologist found slight edema of the uncinate process on the right suggesting involvement of the ethmoid sinus with the tumor and slight induration immediately medial to the punctum of the right eye, possibly reflecting postoperative change or tumor infiltration. She was re-operated on by the otolaryngologist and a medial maxillectomy and resection of the medial inferior orbit with ethmoidectomy were performed. She received radiation which was completed in June 2004.

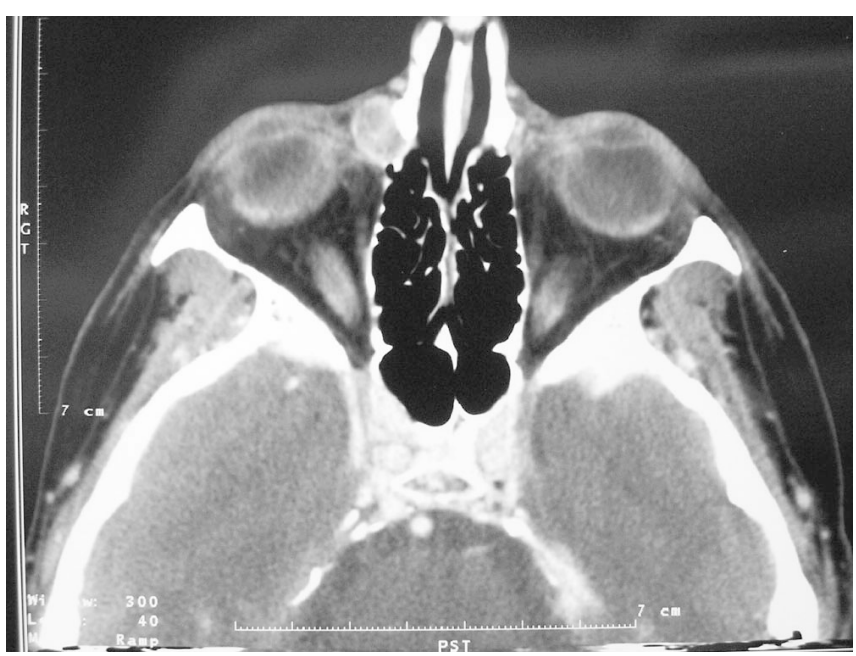

Fig. 1 Contrast enhanced axial CT scan demonstrating dilatation of the right lacrimal sac, 9x9 mm with rim enhancement in the right lacrimal sac region with focal mass effect and slight remodeling to the junction of the posterior crest of the right lacrimal bone with adjacent ethmoid labyrinth

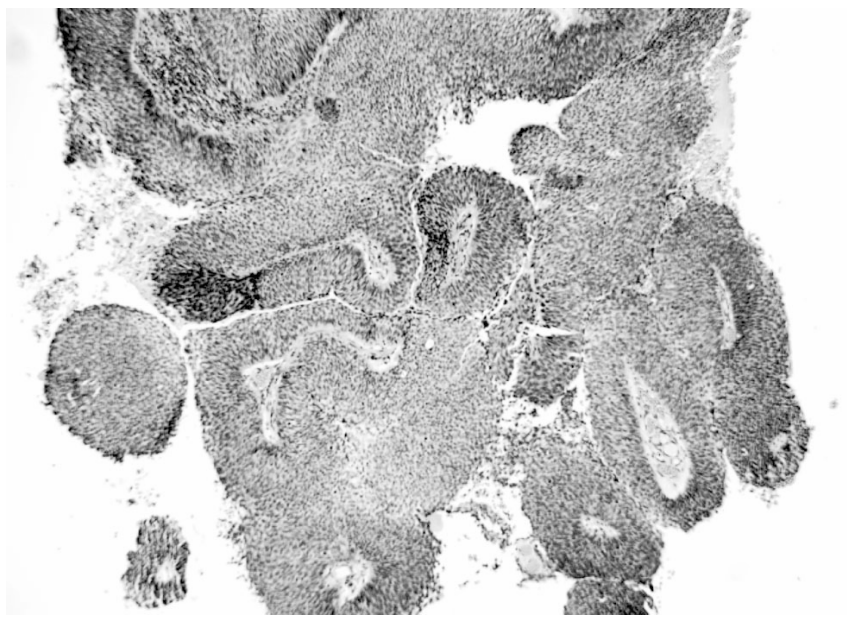

Fig. 2 (H&E stain, x 10) the tumor shows papillary configuration with fibrovascular core

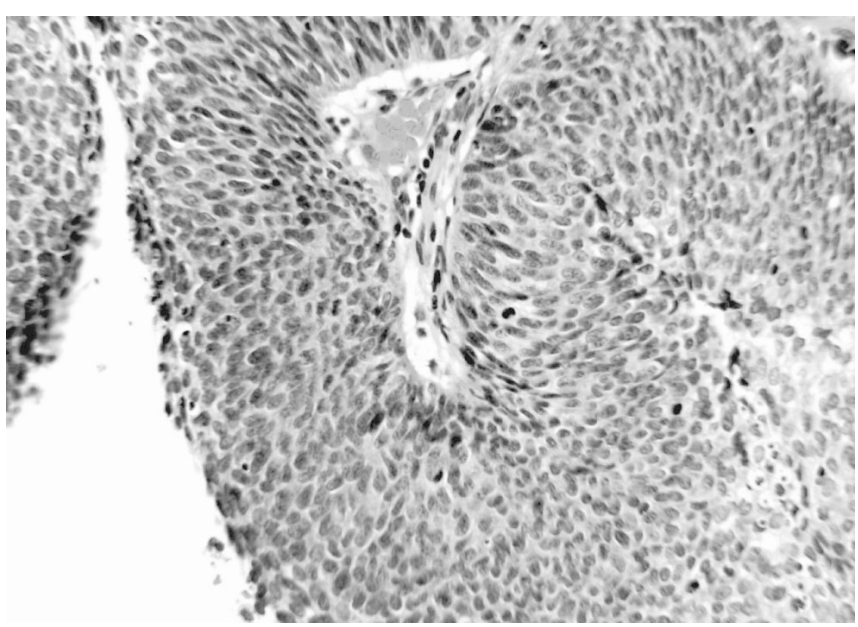

Fig. 3 (H&E stain, x 40) this tumor composes of full thickness dysplastic transitional epithelium with frequent mitoses

On the last follow up date (August 5th, 2004) she had occasional epiphora in the right eye with mild ptosis and no complications from the radiation.

Discussion

The authors reviewed 28 cases of transitional cell carcinoma of the lacrimal sac[4-6,8-12], a rare epithelial tumor. The mean age of those afflicted with transitional cell carcinoma was 50 years, similar to other lacrimal sac tumors which mainly occur in the fifth decade of life, although they have also been found in the young and middle aged (20 years[8] and 36 years old[12]). There is no significant sexual predilection. The most frequent symptom is epiphora, in 67% of cases, and

epiphora with a mass accounted for 53% of all cases. The authors found that a mass without epiphora was the main presenting symptom in one-third of the cases; there was one case of a mass with diplopia. Epiphora with mucocele was present in one case. No cases presented with bloody discharge from the punctum but there was one case of blood stained mucoid material following irrigation. Some mistaken diagnoses were dacryocystitis or mucocele which may have resulted in delayed treatment[5,10], as in the presented case in which the original diagnosis was mucocele. In approximately 40% of lacrimal sac tumors in the series of Stefanyszyn[9] the tumor was not suspected and was only discovered at the time of dacryocystorhinostomy for presumed dacryostenosis.

Irrigation in cases with a history of epiphora and mass may or may not reveal the obstruction, depending on the advancement of the mass. In an early stage, there is usually no or only partial obstruction reversing from the advanced stage. Other investigations should include dacryocystography and / or a CT scan which will reveal any filling defects in conjunction with delayed draining of the contrast material or a distended sac in dacryocystography. This may not be obvious in inverted type tumors which grow inward toward the underlying stroma, especially in the early stages of tumor growth. A CT scan reveals the size of the tumor, any extension to the adjacent tissues such as the sinus, and bony erosions which can affect decision making when planning for surgery.

The differential diagnosis may include dacryocystitis which may result in delayed diagnosis and proper management. Some cases[5] performed dacryocystorhinostomy as initial treatment and had recurrence of transitional cell carcinoma. Mucocele is another differential diagnosis. Clinicopathological findings from lacrimal sac biopsy specimens obtained during dacryocystorhinostomy[13] with unsuspected tumor before biopsy have also found neoplasms in 2.1% of cases, and of these 1 case was transitional cell carcinoma (0.25%). However, lacrimal sac biopsy specimens are not routinely submitted for pathologic examination during dacryocystorhinostomy surgery[14] unless a tumor is found or suspected at the time of operation.

Treatment of transitional cell carcinoma varies from simple daryocystectomy to more extensive surgical excision of the canaliculi, the sac, the nasolacrimal duct, the sinus excision and orbital exenteration. Extension of tumors down the nasolacrimal duct has accounted for many recurrences and failures in therapy[9]. There-fore, a lateral rhinostomy should be performed in cases that show a tendency for intraepithelial spread. Ni[8] compared treatment and results in cases of malignant lacrimal sac tumor, and found that local excision, lateral rhinostomy and irradiation had a much lower mortality rate than cases that did not do lateral rhinostomy (12.5%, and 43.7% respectively).

Despite aggressive treatment in some cases, both recurrence, and metastasis from transitional cell carcinoma are high. One third of the patients had recurrent diseases and 22% had distant metastasis to the lung and esophagus, of which pulmonary metastasis was the most common site, usually causing death. The overall mortality rate was 44% in all of the reviewed literature, but Ni[8] found that 100% of transitional cell carcinomas resulted in death. In considering the treatment and prognosis, pathological types and growth pattern are extremely important factors. Epithelial carcinoma of the lacrimal sac is said to have a recurrence rate of 50%[5], and transitional cell carcinoma give the worst prognosis in this group. An inverted growth pattern is associated with the guarded prognosis because it invades into the underlying stroma of the lacrimal sac wall, causing an invasive acanthosis[5,9].

Some cases of transitional cell carcinoma arose from inverted papilloma[5,12] which is a benign tumor but has a tendency to recur or result in malignant transformation, especially in cases with inadequate initial excision. Nakamura and Mashima[15,16] found that human papilloma virus may be related to inverted squamous papilloma of the lacrimal sac. Therefore, physicians should be alert to the possibility of human papilloma virus infection, especially in young adults with lacrimal sac papilloma, because it may have oncogenic potential.

Conclusion

Lacrimal sac tumor is a rare tumor, and the authors do not know the exact incidence of this disease. Seventy percent of all cases are malignant, with epithelial tumor the most common pathological type and squamous cell carcinoma is the most common in this group. The transitional cell carcinoma is rare and has a poor prognosis. Early diagnosis and prompt treatment is essential to improve this guarded prognosis. Wide excisions with the document of extension from examination and imaging studies with pre or postoperative radiation is recommended to discourage recurrence and metastasis of this disease. Currently, the authors are unable to give an accurate recurrency or mortality rate because of limited follow up studies

(3 months to 6 years)[4-6,8-12]. There is a possibility of a relationship between squamous papilloma of the lacrimal sac and human papilloma virus infection[15,16], similar to HPV infection affecting the female reproductive system via dysplastic change or carcinoma.

References

1. Spratt CN. Primary carcinoma of the lacrimal sac. Arch Ophthalmol 1937; 18: 267-73.
2. Duke-Elder S. Tumors of the lacrimal passages. In: Textbook of Ophthalmology; vol 5. St. Louis: CV Mosby, 1952: 5346-58.
3. Radnot M, Gall J. Tumoren des Traenensacks. Ophthalmologica 1966; 151: 1-22.
4. Schenck NL, Ogura JH, Pratt LL. Cancer of the lacrimal sac. Ann Otol Rhino Laryngol 1973; 82: 153-61.
5. Ryan SJ, Font RL. Primary epithelial neoplasms of the lacrimal sac. Am J Ophthalmol 1973; 76: 73-88.
6. Flanagan JC, Stokes DP. Lacrimal sac tumors. Ophthalmology 1978; 85: 1282-7.
7. Hornblass A, Jakobiec FA, Bosniak S, Flanagan J. The diagnosis and management of epithelial tumors of the lacrimal sac. Ophthalmology 1980; 87: 476-90.
8. Ni C, D'Amico DJ, Fan CQ, Kuo PK. Tumors of the lacrimal sac. Int Ophthalmol Clin 1982; 22: 121-40.
9. Stefanyszyn MA, Hidayat AA, Pe'er JJ, Flanagan JC. Lacrimal sac tumors. Ophthal Plast Reconstr Surg 1994; 10: 169-84.
10. Parmar DN, Rose GE. Management of lacrimal sac tumors. Eye 2003; 17: 599-606.
11. Jones HM, Thornhill CW. Transitional celled carcinoma of the lacrimal sac. J Laryngol Oto 1969; 83: 397-401.
12. Anderson KK, Lessner AM, Hood I. Invasive transitional cell carcinoma of the lacrimal sac arising in an inverted papilloma. Arch Ophthalmol 1994; 112: 306-7.
13. Anderson NG, Wojno TH, Grossniklaus HE. Clinicopathologic findings from lacrimal sac biopsy specimens obtained during dacryocystorhinostomy. Ophthal Plast Reconstr Surg 2003; 19: 173-6.
14. Lee-Wing MW, Asenhurst ME. Cinicopathologic analysis of 166 patients with primary acquired nasolacrimal duct obstruction. Ophthalmology 2001; 108: 2038-40.
15. Nakamura Y, Mashima Y, Kameyama K. Human papilloma virus DNA detected in case of inverted squamous papilloma of the lacrimal sac. Br J Ophthalmol 1995; 79: 392-3.
16. Nakamura Y, Mashima Y, Kameyama K, Mukai M, Oguchi Y. Detection of human papilloma virus infection in squamous tumors of the conjunctiva and lacrimal sac by immunohistochemistry, in situ hybridization, and polymerase chain reaction. Br J Ophthalmol 1997; 4: 308-13.

รายงานผู้ป่วยมะเร็งถุงน้ำตาชนิด Transitional cell carcinoma

ภัสสร ปรีชาไว, โรเบิร์ต ซี เดลลา ร็อคคา, เดวิด เดลลา ร็อคคา, สตีเฟน เซฟเฟอร์, สตีเฟน แมคคอร์แมค

เนื้องอกถุงน้ำตาเป็นโรคที่พบได้น้อยและไม่ทราบอุบัติการณ์ของโรคที่แท้จริง พบว่ามะเร็งเป็นสาเหตุได้ถึงร้อยละ 70 และกลุ่ม epithelial tumor เป็นสาเหตุที่พบได้มากที่สุด squamous cell carcinoma เป็นสาเหตุที่พบได้บ่อยที่สุดในกลุ่มนี้ ในขณะที่ transitional cell carcinoma พบได้น้อยมาก และการพยากรณ์โรคไม่ดี ได้รายงานผู้ป่วยหญิง อายุ 57 ปี ซึ่งเป็น transitional cell carcinoma ของถุงน้ำตา ผู้ป่วยมีน้ำตาไหลจากตาข้างขวาได้รับการวินิจฉัย ก่อนผ่าตัดว่าเป็นท่อน้ำตาอุดตันร่วมกับ dacryocele ขณะทำผ่าตัดพบเนื้องอกที่ถุงน้ำตาได้ตัดชิ้นเนื้อตรวจ frozen section พบ squamous papilloma with dysplasia ได้ผ่าตัดเอาก้อนออกทั้งหมดและทำ dacryocystorhinostomy หลังจากรายงานผลชิ้นเนื้อครั้งที่สองพบ papillary transitional cell carcinoma ที่ถุงน้ำตา จึงทำผ่าตัดอีกครั้งด้วยการทำ medial maxillectomy และตัด medial inferior orbit ร่วมกับทำ ethmoidectomy หลังจากนั้นผู้ป่วยได้รับการฉายรังสีรักษาจนครบและปราศจากการกลับเป็นซ้ำที่ระยะเวลา 2 ปี