Lacrimal Gland Tumors in a Medical Center

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Abstract

Between 1971-2001, there were 9 cases of lacrimal gland tumors in the hospital medical records, 3 adenocystic carcinomas, 1 adenosquamous, 3 benign mixed tumors and 2 malignant mixed tumors. Only 4 had complete clinical histories: 1 benign mixed tumor and 3 adenocystic carcinomas. Among the 4 patients, 3 of them had the tumors removed by transeptal techniques and only 1 had complete removal of the tumor in a single mass. The histopathology proved to be a benign mixed tumor and was completely cured. The other 2 were adenocystic carcinomas which could not be completely removed, resulting in recurrences of the tumors and distant metastases. Both of them died from intracranial extensions. In 1 case of adenocystic carcinoma, the tumor was removed easier by the lateral orbitotomy technique. The nature of this kind of tumor must be known for accurate diagnosis, proper plan of the management and prognosis of the patient.

Key word : Lacrimal Gland Tumor, Benign and Malignant

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A lacrimal gland tumor is not often found. Among the orbital tumors, the malignant epithelial of the lacrimal gland is the most common, (32%) the second most common is the carvernous haemangioma and dermoid cysts and benign epithelial tumors of the lacrimal gland are the third (13%)[1]. The patient usually presents with a mass at the upper outer quadrant of the eye lid and may or may not be associated with eye pain or headache, depending on the rate of growth, the size and type of the tumor. A malignant tumor is mostly associated with symptoms of pain. The diagnosis of this kind of tumor can not rely only on the clinical symptoms. Many special investigative tools are necessary and helpful for the diagnosis,

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such as skull X-rays, skull computed tomography (CT) scan, and magnetic resonance (MRI)[2-4]. A benign epithelial tumor is a benign mixed tumor, the malignant ones are adenocystic carcinoma, malignant mixed tumors, and adenocarcinomas. The percentages of each kind of tumor vary from different reports[5]. The lacrimal gland tumor is located under the upper lid and in the anterior part of the orbit and can extend into the orbital cavity. If the incision is made through the upper lid or by the transeptal technique, sometimes the tumor can not be removed in a single mass, as the tumor may extend into the posterior part of the orbit. In the benign mixed tumor, it is necessary to remove this kind of tumor in a single mass as it could transform into a malignant mixed tumor if some of the tumor tissues remain in the orbit. Therefore, it is necessary to understand the nature of this kind of tumor for the proper diagnosis, treatment and prognosis of the patient.

MATERIAL AND METHOD

The medical records of patients with the diagnosis of lacrimal gland tumors between 1971-2001 were reviewed. 4 of the 9 cases had complete clinical histories and are reported as follows.

CASE REPORTS

Patient 1 (H.N. 1068327)

A 43 year old Thai male had a history of nonpainful exophthalmos, with a palpable mass under the upper lid of his right eye for 15 years. The mass became rapidly enlarged with diminished vision during the previous year. On examination, a 2 cm in diameter, non cystic, smooth surface, no tender mass was found on the upper outer part of the right upper lid. The right eye protruded and was slightly lower than the left eye (Fig. 1). The ocular motility was normal. Visual acuities were 2/60, 6/6. Fundus examination revealed a slightly blurred disc margin at the upper nasal region. The blood vessels were tortuous. The upper temporal retina was lifted up by the pressure effect of the extra ocular mass. The skull X-rays revealed an enlarged right orbit and there was haziness of the right orbit and pressure effect at the upper outer part of the right orbit. (Fig. 2) A benign mixed tumor was suspected and was completely removed by the transeptal technique. (Fig. 3-4) The round, smooth surface, whitish in color, encapsulated tumor mass was easily detached from the surrounding tissues by blunt dissection. (Fig. 5) Histopathologically, an HE stain revealed a benign mixed tumor. (Fig. 6) One week post-operatively, the visual acuity was slightly improved to 3/60. (Fig. 7) There was no recurrence of the tumor during the 11 year follow-up period.

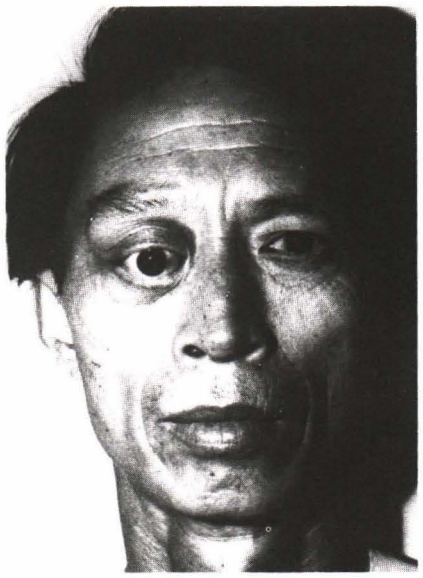

Fig. 1. Patient 1 Showing upper outer quadrant mass with downward displacement of the right globe.

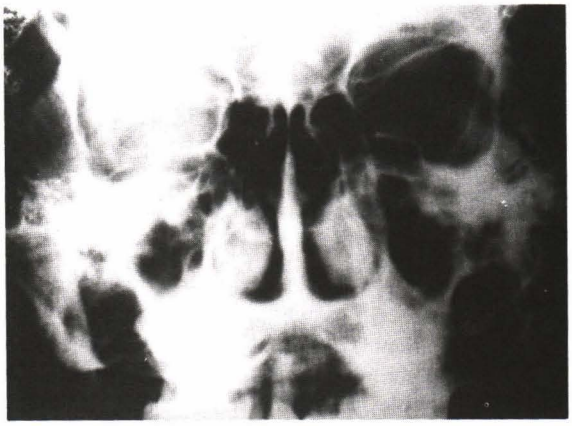

Fig. 2. Patient 1 Skull X-rays, showing haziness, enlargement of the right lacrimal gland fossa & orbit as compare to the left.

Patient 2 (H.N. 1877059)

A 16 year old Thai female had a painless, gradually enlarged mass at the upper outer part of the

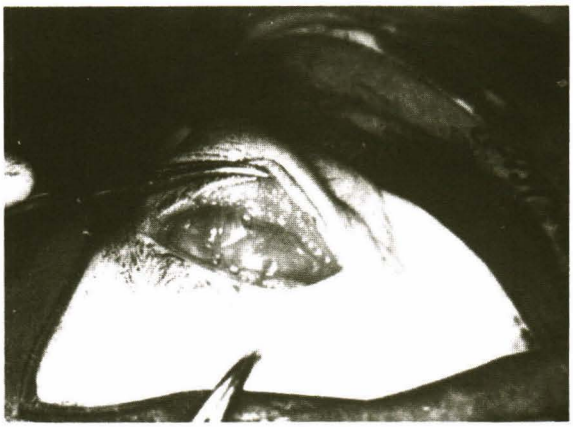

Fig. 3. Patient 1 The well encapsulated tumor mass appeared through the transeptal incision.

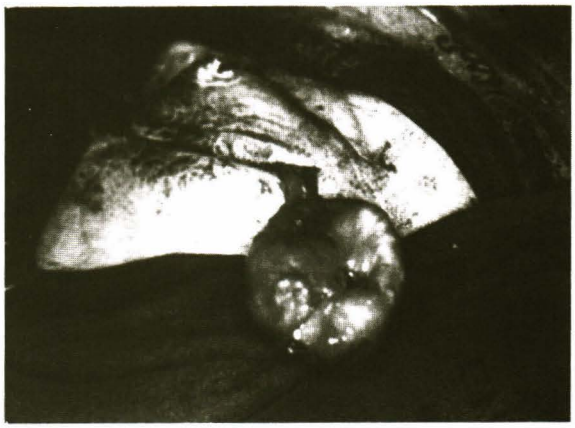

Fig. 4. Patient 1 The tumor mass was removed out by blunt dissection.

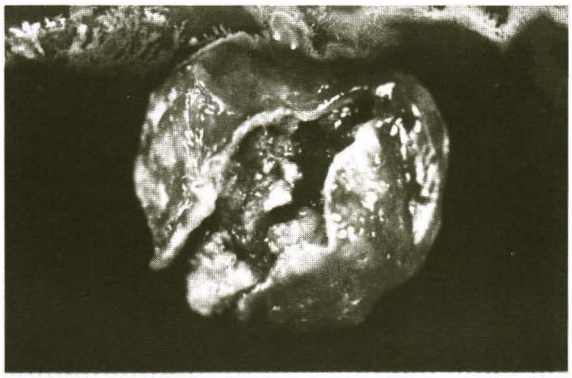

Fig. 5. Patient 1 The 3.5 cm in diameter of the tumor mass & its cut surface showing the friable greasy material inside the tumor.

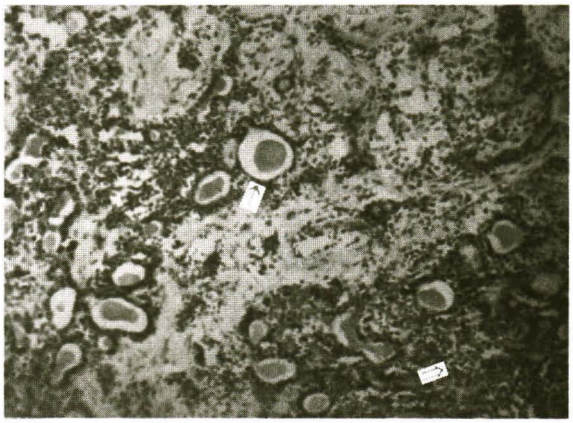

Fig. 6. Patient 1 Histopathologic section showed groups of acinar cells tending to arrange in duct like structure and retention microcysts (→) containing hyalinized fluid among fibromyxoid stroma (⇐) (An HE stain X10).

right eye for 6 months. On examination, there was a 2 cm in diameter mass, firm in consistency, slightly movable on the upper outer part of the right upper lid. (Fig. 8) The ocular motility was normal. Visual acuities were 6/9, 6/6. The upper temporal retina was lifted up from the outside mass. The skull X-rays and optic foramen appeared normal. She was diagnosed as having a lacrimal gland tumor and the mass was removed piecemeal by the transeptal technique. The gross specimens appeared fishy. Histopathologically, an HE stain revealed an adenocystic carcinoma of the lacrimal gland. Radiotherapy at the dosage of 5,000 Rad was given to her for 6 weeks. (Fig. 9) At the 4 month follow-up period there was a recurrent mass at the lower lid region of the left eye. (Fig. 10) The X-rays skull film showed a 1.3 cm nonosteolytic lesion at the apex of the orbit. Exenteration of the right eye was delayed for 4 months as she was 6 months pregnant. The mass was more enlarged after her parturition (Fig. 11). The repeated skull X-rays

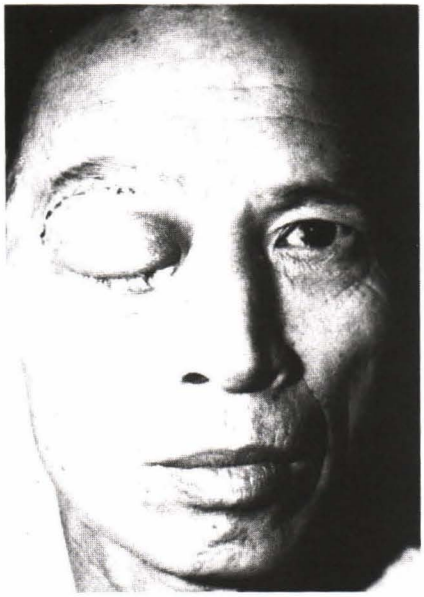

Fig. 7. Patient 1 Showing Ptosis of the right eye at 1 week post-operative.

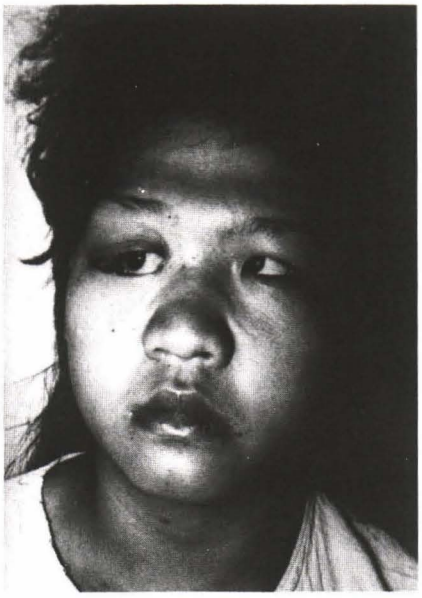

Fig. 8. Patient 2 Showing a 2 cm mass on the right upper lid on the first admission.

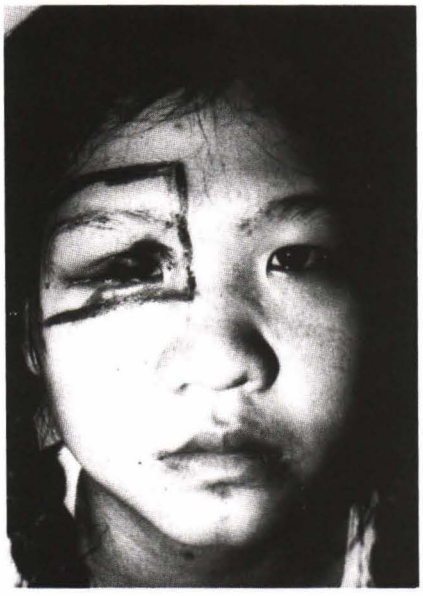

Fig. 9. Patient 2 Radiotherapy at the first week post-operative.

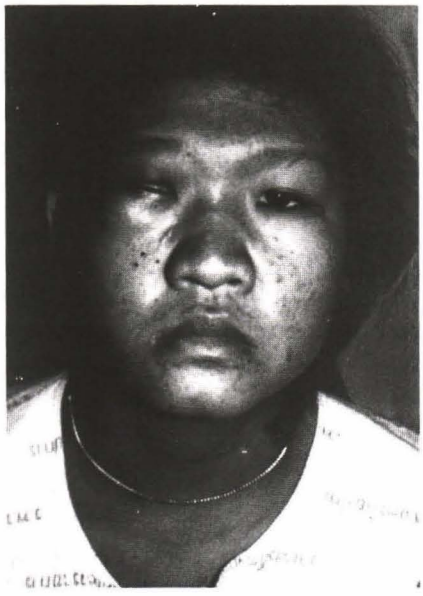

Fig. 10. Patient 2 Showing recurrence of the tumor at the upper & lower lids at 4 months follow-up.

showed an enlarged right orbit with expansion of the floor and superior orbital fissure and also erosions of the superior orbital rim and sphenoid bone. Another 5,000 Rad of repeated radiotherapy was given but 6 months later the tumor had extended to the maxillary, ethmoid, sphenoid bones and the nasopharynx and

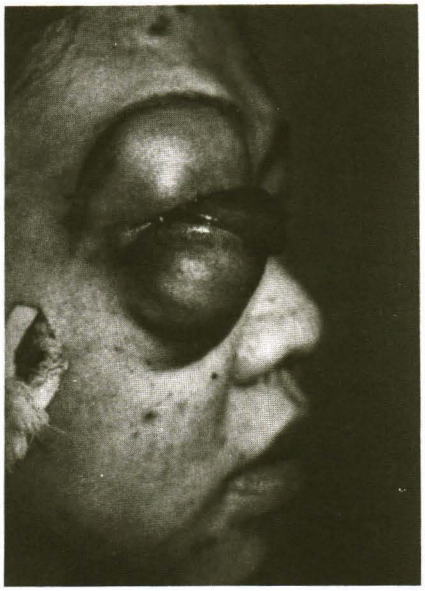

Fig. 11. Patient 2 Showing the more enlarged recurrent mass at 8 months follow-up.

Fig. 12. Patient 3 Showing the transeptal operative scar at 7 weeks post-operative.

into the intracranial cavity. She died from intracranial extension 3 years and 5 months post-operatively.

Patient 3 (H.N. 260476)

A 45 year old Thai female had a history of pain on the left side of the face off and on for 1 month. On examination, there was entropion at the left lower eye lid and was surgically corrected. Four months later, a 2 cm in diameter mass appeared at the upper outer quadrant of the left upper lid. Chronic dacryoadenitis was suspected and treatment with prednisolone 30 mg/day was started. She had more pain and the mass was larger after 5 months' treatment. The globe had been pushed downward and inward but without visual damage. The skull X-rays revealed a solid tumor in the left orbit without any bony destruction. The chest X-rays appeared normal. She was diagnosed as having a lacrimal gland tumor which was excised piecemeal by the transeptal technique. Histopathologically, an HE stain revealed an adenocystic carcinoma of the lacrimal gland. Radiotherapy at the total dose of 5,000 Rad was given over a 6 week period. One week later she developed sudden blindness of her left eye with a recurrent mass on the upper lid. (Fig. 12) The skull CT scan showed the recurrent mass lying on the left optic nerve and destruction of the lateral and medial walls of the orbit (Fig. 13). The chest X-rays showed the multiple nodular infiltrations. Exenteration of the left eye was done and 5 Fluouracil introduced intravenously, followed by triple regimens of chemotherapy, platinol, adriamycin, mitomycin C. (Fig. 14) She died from intracranial extension of the tumor after 4 years, 8 months.

Patient 4 (H.N. 1218890)

A 29 year old Thai female had a right upper lid mass, occasional eye pain associated with blurred and double vision for 1 month. Visual acuities were 6/6, 6/9. The Hertel ophthalmometer was 17, 14 at 94 mm distance. A 1.5 cm in diameter mass with a rough surface, firm in consistency mass was seen at the upper outer lid region with downward displacement of the globe. There was limitation of the upper gaze eye movement. Fundus examination revealed engorgement of the central retinal artery and vein. The optic disc margin appeared slightly blurred. The skull X-rays showed an erosion of the lacrimal gland fossa. The skull CT scan revealed a superolateral mass of the right orbit with lateral rectus muscle involvement. A lacrimal gland tumor was expected and an incisional biopsy was done by the lateral orbitotomy technique. Histopathological report of the frozen section was adenocystic carcinoma. The tumor

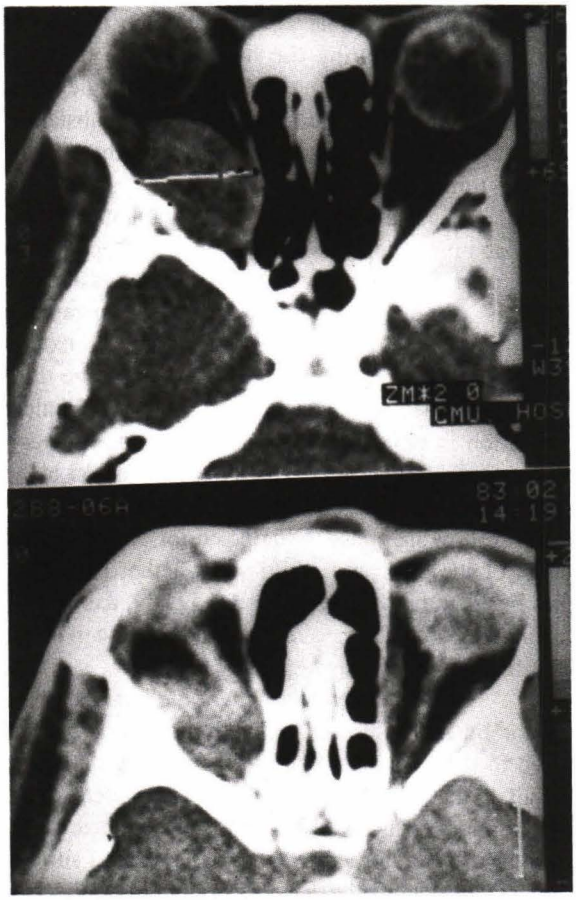

Fig. 13. Patient 3 The skull CT. scan showing the recurrent mass extending to the apex, medial, & lateral wall of the orbit with the optic nerve involvement.

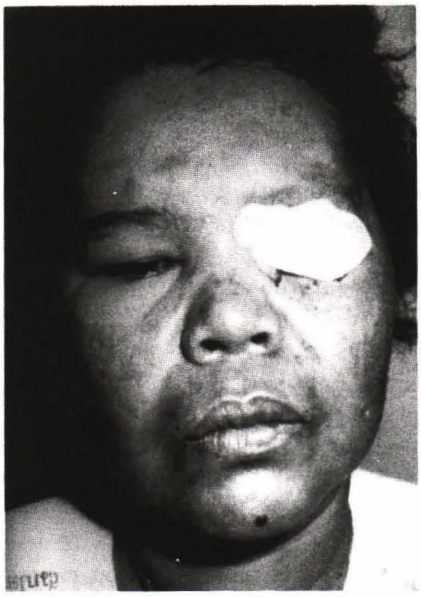

Fig. 14. Patient 3 At 1 week after the exenteration of the left eye.

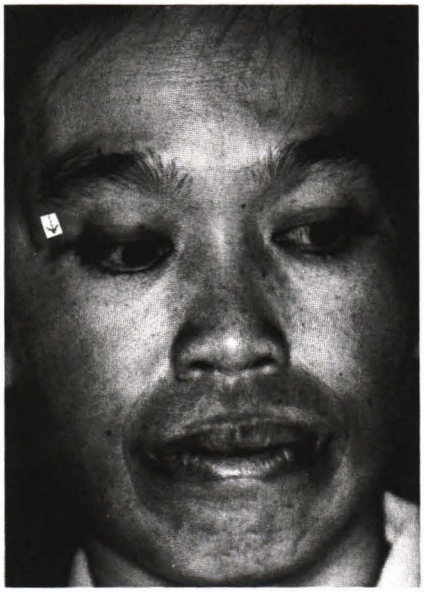

Fig. 15. Patient 4 Showing the lateral orbitotomy incisional scar at 1 week post-operative. (arrow head)

was removed piecemeal as the tumor had extended to the orbital apex. The bony involvement was also removed and replaced by an acrylic plate. Radiotherapy at the total dose of 6,000 Rad over a 6 week period was given. At 8 years' follow-up, there was still no recurrence of the tumor. (Fig. 15)

DISCUSSION

The lacrimal gland is divided into palpebral and orbital lobes. The orbital lobe is deeper into the roof of the orbit and has more chance of becoming malignant than the palpebral lobe[6]. Lacrimal gland tumors can be benign or malignant and both have different clinical pictures. The age range is between 4-70 years old and are found both in males and females. An adenocystic carcinoma is more common in females than in males as can be seen in 3 patients in this report[7]. The patient usually presents with a mass under the upper lid, some of them could palpate

the mass themselves. Some may or may not be associated with pain. In malignancy, the mass usually grows quickly and is often associated with pain as was seen in the third and fourth patients. The abnormal ocular motility and decrease in VA. usually reflect to ocular muscles or nerve supply involvement. These events usually occur in the malignancy cases as seen in the fourth patient. In a patient with a short history of mass, without pain and abnormal ocular motility, malignancy can not be ruled out as in the second patient in this report. Fewer patients present with Trichiasis or Entropion as seen in the third patient, and is probably caused by the traction of the tumor mass resulting in lid deformity.

A lacrimal gland tumor, both benign and malignant with a short clinical history less than 6 months, usually has no bony orbital wall destruction. However, if there is bony destruction and associated abnormal ocular motility, it usually reflects to malignancy as seen in the fourth patient. The configuration of the mass from the skull CT scan could be helpful in the diagnosis of this kind of tumor. The contour of the mass, internal structure, enhancement and change of the nearby bony structures are helpful in the diagnosis. A benign tumor usually has a round, smooth surface. The rough surface is more likely to be malignancy[8]. A skull CT a scan with a 1 mm section is better than a 2 mm section. A multiplanar cut skull CT scan constructed in a 3 dimensional image, is equally good compared with the MRI. The internal structure of the mass and integrity of the nearby bony structures and intracranial extension of the tumor mass are better seen by the MRI than a skull CT scan[9].

Misdiagnosis can still happen with the use of many clinical and investigational tools. Therefore, clinicians must know the nature of this kind of tumor, especially the benign mixed tumor which usually has a history of one year or more as was seen in the first patient in this report. If the ophthalmologist can not remove the tumor as a single mass, the remaining tissues, though the histopathological picture is benign, can invade the nearby structures or even intracranial extension. Therefore, one should separate a benign mixed tumor from the other tumors. When a benign mixed tumor is expected, the ophthalmologist must remove it in a single mass without prior biopsy[10]. On the contrary, if a carcinoma is expected, an incisional biopsy with a frozen section should be done and the ophthalmologist could perform a further operation when the results of the histopathology are available. To prevent the malignant cells dispersing during the biopsy and to help the ophthalmologist to remove the tumor in a single mass, Tse DT, is recommended as the chemical substance, butyl-2-cyanoacrylate which could coat the surface of the tumor[11].

The life expectancy of the patient with this kind of tumor depends on the choice of operative technique. The transeptal technique is easier but there is a higher risk of not being able to remove the tumor in a single mass as the tumor can extend to the posterior part of the orbit, as seen in the third and fourth patients. The lateral orbitotomy or Kronlein operation is more useful though it is more difficult to perform as the bony wall of the orbit has to be removed during the operation. The ophthalmologist could follow the mass to the posterior or even to the apex of the orbit and could see whether the tumor has invaded the bony walls or not. Lastly, there could be a better survival rate for the patient as seen in the fourth patient of this report.

The adjuvant treatment of the lacrimal gland tumor by radiation or chemical substances as was used in the second and third patients, may not help at all if the tumor is not removed completely. On the contrary, if it could be removed completely and followed by radiotherapy, the patient could survive longer as seen in the fourth patient. Melerum ML reported 2 patients with advanced adenocystic carcinoma with intracranial involvement. He used the treatment technique called Neoadjuvant chemotherapy. Cisplatin was injected into the intracarotid artery followed by intravenous Doxyrubin hydrochloride before performing the exenteration. After which 50-60 Gy deep X-ray was done and followed by another 2 courses of the Neoadjuvant chemotherapy. The cancer size decreased in one patient and the clinical picture of the other one changed from intracranial to intraorbital involvement. Lastly, the tumor could be removed and both patients survived for 9 and 7 years respectively[12]. Naugle T Jr reported extensive local involvement without distance metastasis in a 70 year old female who had two local excisions of recurrent adenocystic carcinoma. Total exenteration was performed with aggressive resection of the local involvement followed by radiotherapy. She died from extension of the cancer to the scalp 13 years later.

A patient with a lacrimal gland tumor should have a proper plan for the diagnosis and treatment as soon as possible. If a malignant epithelial tumor is expected, the ophthalmologist must discuss the

possibility of an exenteration before the operation and must decide whether to make an incision biopsy or not. The choice of operative technique is also important. If the tumor mass can not be removed in a single mass, though followed by chemical therapy or radiotherapy or exenteration, the tumor usually recurs and death ensues. Heaps RS. recommended exenteration from the beginning for a patient with an adenocystic carcinoma. He found that 57 per cent of the cases had no recurrences[14]. Polito E reported 20 patients with lacrimal gland tumors comparing extensive surgery and conservative treatment preserving the eye. The 7 patients with orbital exenteration and or bony orbit removal had a median survival of 6.75 years. The 12 patients with preserved eye followed by radiotherapy had a median survival of 9 years. Therefore, surgery of the lacrimal gland should have a very good judgment, there is no need for exenteration from the beginning in every case[15]. As the tumor can recur even after 10-20 years, therefore, whole life follow-up is necessary for this kind of tumor[16].

In conclusion, the ophthalmologist must differentiate a benign mixed tumor from other tumors of the lacrimal gland and biopsy is prohibited for this kind of tumor. The tumor should be removed in a single mass. If malignancy of the lacrimal gland is expected, incisional biopsy should be done with frozen section stand by and the patient ready for further operation once the pathological report is known. A lateral orbitotomy or Kronlein operation is recommended for the possible complete removal of the mass.

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เนื้องอกของต่อมน้ำตาในโรงเรียนแพทย์แห่งหนึ่ง

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ในช่วงระหว่างปีพ.ศ. 2524–2544 มีผู้ป่วย lacrimal gland tumors 9 ราย 3 รายเป็น adenocystic carcinomas 1 รายเป็น adenosquamous 3 รายเป็น benign mixed tumors 2 รายเป็น malignant mixed tumors มีผู้ป่วยเพียง 4 ราย เท่านั้นที่มีรายงานที่สมบูรณ์และได้นำมาศึกษาอย่างละเอียดดังนี้ Benign mixed tumor 1 ราย 3 รายเป็น adenocystic carcinomas ผู้ป่วย 3 รายได้รับการผ่าตัดโดยใช้เทคนิคที่เรียกว่า Transeptal ปรากฏว่าสามารถผ่าเอาก้อนเนื้องอกออกได้ทั้งก้อนและผู้ป่วยหายดีเพียงรายเดียว คือรายที่เป็น benign mixed tumor อีก 2 รายเป็น adenocystic carcinoma ไม่สามารถผ่าเอาก้อนเนื้องอกออกได้เป็นก้อนเดียวทำให้เกิด recurrences และมีการแพร่ของมะเร็งไปยังสมองและส่วนอื่นของร่างกายและถึงแก่กรรมต่อมา ผู้ป่วย adenocystic carcinoma 1 ราย ได้รับการผ่าตัดโดยการผ่าเข้าทางด้านข้างของกระบอกตาที่เรียกว่า Lateral orbitotomy ทำให้ผ่าตัดเอามะเร็งออกได้ง่ายกว่า จักษุแพทย์จำเป็นต้องเข้าใจธรรมชาติของเนื้องอกชนิดนี้เพื่อให้ได้การวินิจฉัย การวางแผนการรักษา การพยากรณ์โรคที่ถูกต้อง

คำสำคัญ : เนื้องอกต่อมน้ำตา, เซลล์ชนิดธรรมดาและเนื้อร้าย

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