Orbital Exenteration for Secondary Orbital Tumors: A Series of Seven Cases

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Background: Exenteration is indicated in patients with malignant neoplasms of orbital contents. It entails the removal of the eyeball together with its extraocular muscles and other soft tissues. Exenterations can be classified into (1) total, (2) subtotal, and (3) supertotal exenteration. Retrospectively study, we reviewed 7 patients that had received exenteration/subtotal exenteration with spontaneous granulation/myocutaneous flap implantation or eyelid-sparing exenteration with myocutaneous flap. Primary lesions, histopathological examination results, treatments, and recurrences are discussed.

Methods: A retrospective study of the years 1987 through 2000 disclosed 7 patients that underwent exenteration/subtotal exenteration. The patients ranged in age from 41 to 68 years. Two patients underwent total exenteration without socket augmentation; 4 patients underwent exenteration/ subtotal exenteration with immediate facial reconstruction, and 1 with delayed facial reconstruction.

Results: Classification of the 7 patients showed that 2 had basal cell carcinoma of the skin, 2 had squamous cell carcinoma of the conjunctiva, 1 had squamous cell carcinoma of the paranasal sinus, 1 had rhabdomyosarcoma of the paranasal sinus, and 1 had intracranial meningioma. Radiotherapy was performed in 6 of the patients and chemotherapy in 2. Central nerve system invasion was noted in 2 patients, and 1 died due to it.

Conclusion: Secondary orbital tumors involved the orbit from adjacent tissues: paranasal sinuses, nasopharynx, lacrimal sac, conjunctiva, eyelid, intraocular tissue, and intracranial tissues. Combined surgeries are necessary for complete tumor removal. And the imaging studies should include the field of the orbit, sinus, and brain to search for the primary lesions.
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Key words: exenteration, secondary orbital tumors, split - thickness skin graft, basal cell carcinoma, squamous cell carcinoma, rhabdomyosarcoma.

Secondary orbital tumors represent one third of all orbital tumors.(1) They arise in adjacent tissues including sinuses, nasopharynx, meninges, brain, intraocular, conjunctiva, lid, and lacrimal sac but most secondary orbital tumors arise from the paranasal sinus, especially the maxillary sinus. The signs are as follows: proptosis, global displacement, conjunctival congestion and chemosis, squinting,

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decreased acuity, and nasal obstruction. We present seven patients with secondary orbital tumors, who underwent total or eyelid-sparing exenteration. We emphasize the survey for the adjacent orbital tissue to make early diagnosis and combined therapy for the highly malignant tumors.

METHODS

A retrospective search of the years from1987 through 2000 identified 7 patients who underwent orbital exenteration. The patients ranged in age from 41 to 68 years. An ophthalmologist, otolaryngologist, or plastic surgeon performed the surgeries. Two patients underwent total exenteration with spontaneous granulation; 4 patients underwent exenteration/subtotal exenteration with myocutaneous flap or split - thickness skin graft (STSG), and 1 underwent exenteration with delayed myocutaneous flap.

RESULTS

In our retrospective study, the 7 patients ranged in age from 41 to 68 years. They all underwent tumor excision with total exenteration with spontaneous granulation (2 patients), total exenteration with delayed myocutaneous flap (1 patient), total/subtotal exenteration with myocutaneous flap (4 patients). Histopathological examination results revealed that 2 patients had basal cell carcinoma (BCC) (skin), 2 had squamous cell carcinoma (SCC) (conjunctiva), 1 had SCC (maxillary and ethmoid sinuses), 1 had rhabdomyosarcoma (maxillary and ethmoid sinuses), and 1 had meningioma (brain). Two patients showed central nervous system (CNS) invasion and one died due to it. One showed optic nerve invasion. Radiotherapy was performed on 5 patients and chemotherapy on 2 patients. Four patients showed recurrence before exenteration and 1 recurrence happened after exenteration. Two showed brain metastasis; one showed liver metastasis.

Case 1

A 62-year-old man had BCC over his right forehead, which was diagnosed at a different hospital. Twelve months later, he was presented at our hospital due to a recurrent mass over his forehead on the right side, size. Computed tomography (CT) revealed a subcutaneous soft tissue mass over his forehead on the right side. After wide excision of the tumor mass by plastic surgeon, BCC was diagnosed according to pathologic examination results. Three months after excision, recurrent extracranial mass over the temporal area was noted. Further surgery involving wide excision and temporal adipofacial flap with STSG was performed. Five years after excision, at regular follow-up, CT revealed orbital and outer dura invasion. Total exenteration accompanied by myocutaneous flap was performed. The patient underwent fractioned radiotherapy (6660 cGy/37) and no recurrence was noted after exenteration and radiotherapy, during 11 months follow-up.

Case 2

A 67-year-old woman with a huge mass over the left medial canthus and nose for more than 10 years and came to visit our clinic. CT revealed a 6.5 cm irregular fungating mass in the left periorbital, glabellar, and nasal regions, which impressed skin cancer. Palliative tumor excision was performed by otolaryngologist doctor and the histopathologic examination results were basal cell carcinoma. Three months after excision, tumor regression was noted. But due to the possible residual tumor, electron beam therapy was performed during follow-up period of 18 months. Later, the patient had corneal perforation. Her visual acuity showed light sense negative and recurrence was noted on CT and bone scans. The patient underwent complete tumor excision and total exenteration with implantation of myocutaneous flap. After exenteration, no recurrence was noted during 6 months follow-up.

Case 3

A 68-year-old man had a conjunctival mass over the left eye for 1 month and decreased visual acuity for 1 year. Conjunctival masses were noted over the nasal and temporal conjunctiva and the superior and nasal cornea. A B scan revealed an intraocular mass. CT revealed an inferior left orbital mass. Histopathlogic examination results revealed well-differentiated SCC after total exenteration was performed. One and a half months after exenteration, the patient underwent augmentation of socket with temporal myofascial flap and STSG for socket reconstruction. After exenteration, no recurrence was noted during 20 months follow-up.

Case 4

A 36-year-old woman had previously had intracranial meningioma after surgery at a different hospital. Six years later, she visited our hospital due to prominent proptosis for 6 months. CT revealed left orbital tumor secondary to intracranial meningioma; however, she refused treatment. When she was 49-year-old, she was admitted due to weakness in her four limbs and prominent proptosis. CT revealed a recurrent meningioma involving the juxtasellar, suprasellar, and frontal areas. The patient underwent combined surgery of right orbito-frontal craniotomy, tumor excision, and eyelid- sparing exenteration, followed by right rectus abdominis muscle flap by a neurosurgeon, plastic surgeon, and ophthalmologist. Radiotherapy was performed, with the total doses of 2880 cGy/16fr. Major depression was noted during the radiotherapy; thus, the radiotherapy was suspended. The patient was lost to follow up 1 year postoperatively.

Case 5

A 45-year-old man had complained of nasal obstruction and purulent rhinorrhea for 10 years. During a visit at an otolaryngeal clinic, a nasal mass was noted and CT revealed right ethmoid, maxillary and nasal mass with posterior orbital wall defect. The patient underwent right pansinusectomy. Histopathologic examination results revealed the tumor as SCC. Radiotherapy (6200 cGy/31fx) was performed. After 14 months, a right orbital tumor with lateral rectus muscle and lateral wall destruction was noted on magnetic resonance image (MRI) study. The patient underwent right maxillectomy, total exenteration followed by myocutaneous muscle flap and STSG. Chemotherapy was also performed 6 months after exenteration. However, right orbit, apex, and temporal base invasion were noted on a follow-up image study 2 years after total exenteration. Fever, vomiting, and poor appetite were noticed and patient died due to bacterial meningitis.

Case 6

A 62-year-old woman complained of epistaxis for 1 week. Functional endoscopic sinus surgery was performed in a different hospital and malignancy was told, so she was referred to our hospital for further treatment. At our otolaryngeal clinic, MRI was arranged and revealed a mass in the maxillary, ethmoid sinus, and optic nerve involvement , with brain spared. Thus, she was referred to the ophthalmologic clinic for the combined surgery. Her visual acuity was counting fingers 50-60 cm and ocular ductions were completely limited in all directions. An otolaryngologist and ophthalmologist performed the tumor removal and total exenteration with spontaneous granulation. Histopathological examination results of specimens included rhabdomyosarcoma (alveolar type). Chemotherapy with oncovin, endoxan, and adriamycin was given. Radiotherapy with the dose of 6840 cGy/38fr was performed. Six months after beginning chemotherapy, the patient came to the emergency room due to abdominal distention and died due to suspected liver metastasis.

Case7

A 47-year-old man complained of blurred vision, eye pain, and frequent discharge from the left eye. Tumor infiltrate was noted near the superior and nasal conjunctiva and limbus. CT revealed left

Table 1. Clinical Data of the Seven Cases

	Y/O	Gender	cell type, origin	ex/sub	flap	R/T	C/T	CNS	live/dead
Case 1	62	M	BCC, lid	ex	(+)	(+)	(-)	(-)	live
Case 2	67	F	BCC, lid	ex	(+)	(+)	(-)	(-)	live
Case 3	68	M	SCC, conj.	ex	(+)*	(-)	(-)	(-)	live
Case 4	41	F	Meningioma, brain	sub	(+)	(+)	(-)	CNS	live
Case 5	45	M	SCC, e & m	ex	(+)	(+)	(+)	CNS	dead
Case 6	62	F	Rhabdomyosarcoma, e & m	ex	(-)	(+)	(+)	ON	dead
Case 7	47	M	SCC, conj	ex	(-)	(+)	(-)	(-)	dead

Abbreviations: Conj: conjunctiva; e: ethmoid sinus; m: maxillary sinus; mn:month; y:year; ex: exenteration; sub: subtotal exenteration; R/T: radiotherapy; C/T: chemotherapy; CNS: central nerve system involvement; ON: optic nerve involvement; *: delayed flap, pre-recurrence: recurrent tumor before exenteration.

lower orbital tumor. Aspiration cytology from the infiltrate showed conjunctival SCC. Total exenteration with spontaneous granulation was performed and the histopathological examination results were well-differentiated SCC. Radiotherapy was performed (4000 cGy/20fx). Postoperatively, socket skin necrosis was noticed but no intracranial invasion occurred. The patient died due to liver cirrhosis 10 years after total exenteration (Table 1).

DISCUSSION

Exenteration is indicated in patients with malignant neoplasm of the orbital contents, either primary or secendary extension from adnexal tissue that cannot be controlled by simple excision or radiotherapy.[2] It entails the removal of the eye together with its extraocular muscles and the soft tissue of the orbit.[3] According to the lesion excised, exenteration can be classified into (1) total exenteration, (2) subtotal exenteration, and (3) superexenteration.[4] Levin et al showed the following indications for orbital exenteration: (1) eradication of presumed life-threatening malignancy (89%), (2) eradication of life-threatening infection (6%), and (3) alleviation of intractable pain or deformity (5%).[3] Our study also revealed that orbital exenteration was most frequently performed for the treatment of life-threatening malignancies.

During the procedure of total exenteration, the incision site was carried from the skin down to the level of periosteum. The orbital contents with the periosteum were excised after the periosteum was elevated from orbital bones. Subtotal exenteration involved the removal of soft tissue with eyelid preserving. Superexenteration required frontotemporal sphenoidal craniotomy and en bloc orbitectomy. This procedure involved a neurosurgeon, ophthalmologist, and plastic surgeon. In a study by Shields et al, 16 exenterations were performed using an eyelid-sparing technique and six were left to heal by spontaneous granulation. They concluded that the eyelid-sparing technique was more rapid healing and enabled earlier fitting of a prosthesis. In performing an eyelid-sparing procedure, it is necessary to leave some of the orbicularis muscle with the skin flaps, which permits better vascularization.

The 0.3 mm skin graft was harvested from non-hair-bearing anterior or inner sides of the upper thigh by the dermatome. The exenterated orbit was well epithelialized within 3 to 4 months. During the surgery, the orbital content was cut off using a snare. The snare itself was sometimes broken without the contents removal if the instrument was not deeply inserted. It was suggested that the snare should be tightened first and then the whole instrument be rotated. Secondary orbital tumor could arise from paranasal sinuses, nasopharynx, lacrimal sac, CNS, skin (eyelid), and conjunctiva . The common presenting symptoms included chronic sinusitis, epistaxis, nasal obstruction, painful eye, nonaxial displacement of the globe, lid swelling, diplopia, and blurred vision.[5]

Paranasal sinus tumors (SCC type) are the most common sinus malignancies to invade the orbit and they most often involve the maxillary sinus and then ethmoid sinus and frontal sinus. Paranasal sinus tumors (Rhabdomyosarcoma type) involve the ethmoid sinus the most. In addition, they are the most common malignancies in head and neck of the children. They may originate from the ethmoid sinus or nasal cavity and invade into the orbit. They tend to involve the superior part of the orbit. Rhabdomyosarcoma is a highly malignant myogenic tumor and is classified into four subtypes: pleomorphic (adult), embryonal, alveolar, and botryoid. The alveolar type is the least common of the subtypes.[6,7] The initial symptoms may include nasal obstruction or epistaxis. Distant metastasis to the lung and cervical lymph nodes has been reported. Surgical intervention, chemotherapy, or radiotherapy should be combined for the treatment. Secondary orbital tumors from rhabdomyosarcoma are rare and patients usually have poor prognosis for long-term survival. In a report of four patients by Walton et al, the most common manifestations included proptosis, reduced visual acuity, and motility disorders. Despite the combination of chemotherapy and radiotherapy, all patients died within 6 months of orbital metastasis.[7]

Conjunctival SCC arises from perilimbal conjunctiva. Clinically, it presents as dysplasia, solar keratosis, and carcinoma in situ. Because the tumors have low rates of recurrence, they have been treated successfully with local resection and cryotherapy.

Eyelid SCC occurs commonly in fair-skinned elderly people with a history of chronic sun exposure and skin damage.[8] SCC, although less common than BCC, is more aggressive and invasive.

Complete surgical resection of the tumors has been advocated due to the potential for metastasis and its lethality.[9,10] The tumors must be treated using surgical excision and radiotherapy.

Eyelid BCC is the most frequent malignant lid lesion, representing approximately 80% of eyelid neoplasms.[11-15] The malignant cells arise from the so-called basal or germinal cells of epidermis. Most lesions occur in fair-skinned individuals. The lesions involve the lower lid (70%), medial canthal area, upper lid, and lateral canthal area. BCC lesions do not metastasis, and patients have shown 5-year tumor control rates of up to 95%. Cryotherapy for skin tumors has gained in popularity due to the easy application and low rate of complications.[1,5,6] The treatment involves a rapid freezing and a slow thawing and creates a tissue temperature of -40 ℃. Two to three cycles follow each other. But I will treat patient as a combined therapy of surgery and cryotherapy/irradiation, instead of cryotherapy only. Radiotherapy may be useful for BCC, but the results are not as satisfactory as those of cryotherapy. Rhadomyosarcoma is the most common malignancy in head and neck region of children.[16] Primary optic nerve meningioma may arise from the optic nerve sheath and meningeal cell structures near the optic nerve. Orbital involvement by intracranial meningioma is rare and arises from the sphenoid bone. Treatment of intraorbital meningioma is controversial. Visual improvement is not worth the risk of surgery.[17,18] Radiotherapy plays little role in the treatment.[19,20]

Conclusion

Secondary orbital tumors may arise from the sinus, pharynx, meninges, brain, eye, conjunctiva, eyelid, and lacrimal sac. Therefore, the imaging studies must encompass the fields of the orbit, sinus, and brain.

Total exenteration with spontaneous granulation is more effective, more acceptable for cosmesis, and easier for detection of recurrence than other therapies. However, it is time-consuming for complete granulation (3-4 months). Total exenteration with lining of orbital walls with temporalis muscle transplantation and STSG has been suggested by many surgeons.[21,22] After this procedure, the wound heals rapidly and it is easy to care for. However, it is cosmetically unacceptable due to the over-sized graft and it is difficult to detect the recurrence.

Subtotal exenteration is also effective in saving lives, more rapid healing (1-6 weeks) and enables earlier fitting of prosthesis than other treatment modalities. During the procedure, it is important to leave some orbicularis muscle with the skin flaps to permit continued vascularization, and to prevent them from becoming necrotic.[23,24]

Total/subtotal exenteration with spontaneous healing for the sake of early detection of recurrence and good cosmesis is advocated. Delayed socket reconstruction with temporalis muscle rotational flap and STSG is also indicated if poor healing of apex occurs.

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接受眼窩摘除術的續發性眼窩腫瘤：七例報告

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背 景： 眼窩摘除術是應用在眼窩組織的惡性腫瘤，包括源發性及續發性手術時，要將眼球及眼眶軟組織、骨膜完全摘除。眼窩摘除術可以分爲(1)完全的(2)近乎完全的(3)超乎完全的摘除。在回溯研究報告中我們發現七位病例接受眼窩摘除術或是加上顳肌移植，並分析其源發位置、組織學報告、治療方式、復發與否。

方 法： 從1987年至2001年3月，共有七位病例被納入在本研究中，病患年齡從41歲到68歲，這七例中有兩例是單傳純執行眼窩摘除術，有四例執行眼窩摘除術及由整形外科醫師執行顳肌移植及身體其他部位的植皮。一例執行眼窩摘除術後1.5個月執行顳肌移植。

結 果： 這七例的病理報告中，有兩例是基底細胞癌(皮膚)，兩例是鱗狀細胞癌(結膜)，一例是鱗狀細胞癌(鼻竇)，一例是橫紋肌肉瘤(鼻竇)，一例是顱內腦膜細胞瘤。有五病例接受放射線治療，兩個病例接受化學治療；有兩例有轉移到腦部且一例因此而死亡；有一例轉移到肝臟而死亡。

結 論： 續發性眼窩腫瘤有可能來自於眼窩附近組織，包括鼻竇咽喉、淚囊、結膜、眼瞼、眼球內組織、顱內組織。一旦外直肌或軟組織有影響到，眼窩摘除術是必須執行的。影像學檢查時除了眼球外，也要特別注意頭頸部、咽喉部及鼻竇部位。術前的評估及眼科醫師、神經外科醫師、整形外科醫師、耳鼻喉科醫師一起合作，才能將手術完成。

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關鍵字： 眼窩摘除術，續發性眼窩腫瘤，基底細胞癌，鱗狀細胞癌，橫紋肌肉瘤。

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