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CLINICAL INVESTIGATION

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Orbital exenteration: surgical and reconstructive strategies

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Abstract ● Background: Radical exenteration procedures, which include the removal of orbital content and eyelids, result in serious functional limitations, especially with respect to eating and speaking. Therefore we have recently changed our surgical concept. ● Methods: Seventy-seven patients underwent orbital exenteration during the 20-year period from 1974 to 1995 at the Department of Maxillofacial Surgery, Essen University. The simultaneous removal of periorbital bone was performed in 45 of these cases. ● Results: The 1-year survival rate was 89%, the 5-year rate was 63% and the 10-year rate was 48%. The surgical approach, the amount of resected orbital tissue and the reconstructive procedure have been adapted to the individual needs, depending on the location and extent of the tumor. Subsequently, the surgical morbidity has decreased. ● Discussion: Detailed consideration of all clinical and histological findings is essential before surgery, in order to prevent a higher rate of recurrence following these modified operations.

Introduction

Orbital exenteration (originally described by von Arlt [49]) is defined as radical removal of the orbital contents, including the eyelids, the periorbit and the orbital septum [20, 25]. Without subsequent reconstructive surgery, this results in serious mutilation and inability to wear an artificial eye.

Younger patients, in particular, desire preservation of their eyelids, allowing them to wear an artificial eye instead of an external prosthesis. Thus, numerous modifications of the surgical technique have been introduced: preservation of the eyelids [44], the conjunctiva [14, 19, 43] or the periorbit [45].

Healing by spontaneous granulation takes several months [13, 36, 44], whilst the lining of the orbital bone with split skin graft [50] speeds up the healing process, but has the disadvantage of leaving a deep orbit. Naquin [30] and Reese [38] transposed the anterior part of the temporal muscle through the lateral orbital wall into the orbit to make it possible for the patient to wear an eye prosthesis.

The introduction of free tissue transplantation [11, 48] allows the covering of extensive combined defects and the implementation of early postoperative irradiation. On the other hand, some authors argue that any filling of the orbit could mask local recurrence of the tumor [5, 37, 42].

Orbital exenteration is a rare occurrence. Nevertheless, a large number of patients are needed to establish the results of any surgical and reconstructive modification. For this reason we have analyzed all our cases since 1974, to establish an up-to-date and appropriate surgical and reconstructive concept.

Patients and methods

Between 1974 and 1994, 77 patients underwent orbital exenteration in the Department of Maxillofacial Surgery, Essen University. All patients were followed up continuously after surgery, and we were able to examine all living patients for the purpose of this investigation. Those patients who had died in the meantime were evaluated by photographic documentation and by hospital reports. The cause of death was analyzed for all these patients. At the final examination of the living patients the esthetic outcome was documented photographically. Survival time and life table analysis were documented for all patients with tumor-related exenterations.

In all cases the following were analyzed: tumor site and extension, tumor histology, surgical technique, postoperative treatment and esthetic outcome (eye prosthesis, episthesis, opaque patch). Possible relations between the risk of a local recurrence or death on the one hand and the technique of resection and reconstruction on the other hand were investigated.

Results

The average age of the patients at the time of treatment was 53.2 years (range 4–85 years). In 74 of the 77 cases, the orbital content was removed because of a tumor (Table 1). The origin of the tumor varied: 38 originated primarily in the orbit, 34 invaded the orbit from neighboring anatomic structures and 2 were metastases. Three of the 77 orbital exenterations were performed following severe orbital trauma or bullet wound.

Extent of surgery and type of reconstruction

In 32 cases, removal of the orbital content without the surrounding bone was sufficient. In 45 cases, removal of bone in varying quantities was necessary (Table 2). Carcinomas of the maxillary sinus with secondary invasion of the orbital content required a vast block resection of the maxilla and the orbit. In 13 cases the anterior skull base had to be partially removed, in cooperation with a neurosurgeon.

The eyelids were preserved in 36 cases and had to be resected in 41 cases. In many of the former cases we succeeded in preserving a large amount of the tarsal and even bulbar conjunctiva.

The orbit was allowed to granulate spontaneously in 25 patients. The average healing time was 14 weeks, but for some patients the orbit took up to 6 months to heal. Seventeen patients developed a permanent perforation between the orbit and the nasal cavity or the sinuses. Only in seven cases did the procedure result in an impervious epithelial lining of the orbit. In none of these cases had any bone been removed. On the other hand, following bone removal all patients developed a permanent orbito-nasal fistula, even if the nasal mucosa had been preserved during the resection.

Postoperative radiotherapy resulted in partial loss of the secondary epithelial lining and subsequently in infected osteoradionecrosis (three cases). This could only be treated by removal of the complete orbital bone, parts of the frontal bone and the anterior skull base. The resulting vast defect required covering with microvascular tissue transplant.

Table 1 Histopathologic diagnosis in 74 patients who underwent orbital exenteration for neoplasia

Diagnosis	No. of patients
Carcinomas (n = 35)	
Squamous cell carcinoma	13
Adenoidcystic carcinoma	7
Basal cell carcinoma	6
Others	9
Sarcomas (n = 12)	
Liposarcoma	3
Osteosarcoma	1
Rhabdomyosarcoma	6
Leiomyosarcoma	1
Undifferentiated sarcoma	1
Melanomas (n = 13)	
Extraocular choroidal melanoma	9
Conjunctival melanoma	3
Secondary orbital melanoma	1
Miscellanea (n = 14)	
Neuroblastoma	3
Retinoblastoma	3
Malignant lymphoma	2
Malignant ameloblastoma	1
Meningeoma	1
Neurofibroma	1
Hemangiopericytoma	1
Plasmocytoma	1
Aggressive fibromatosis	1

Table 2 Extent of bone resection

Orbital exenteration without bone resection	32
Orbital exenteration with bone resection (n = 45)	
Resection of the orbital roof	13
Resection of other orbital walls	26
Complete orbitectomy	2
Partial maxillectomy	26
Partial resection of zygoma	11

Table 3 Reconstructive procedures

Free epithelialization	35
Surgical closure of orbit (n = 52)	
adaptation of the preserved conjunctiva	12
temporalis muscle transplantat (eyelids preserved)	13
pedicled flaps	14
free microvascular transplants	14[a]

[a] In one case secondary to temporal muscle transplant

Before the technique of microvascular tissue transplant was introduced in our clinic, defects following block resection of the orbit and maxilla remained uncovered. Therefore those patients had to wear a combination of obturator denture and episthesis. Severe functional limitations with respect to eating and speaking were incurred.

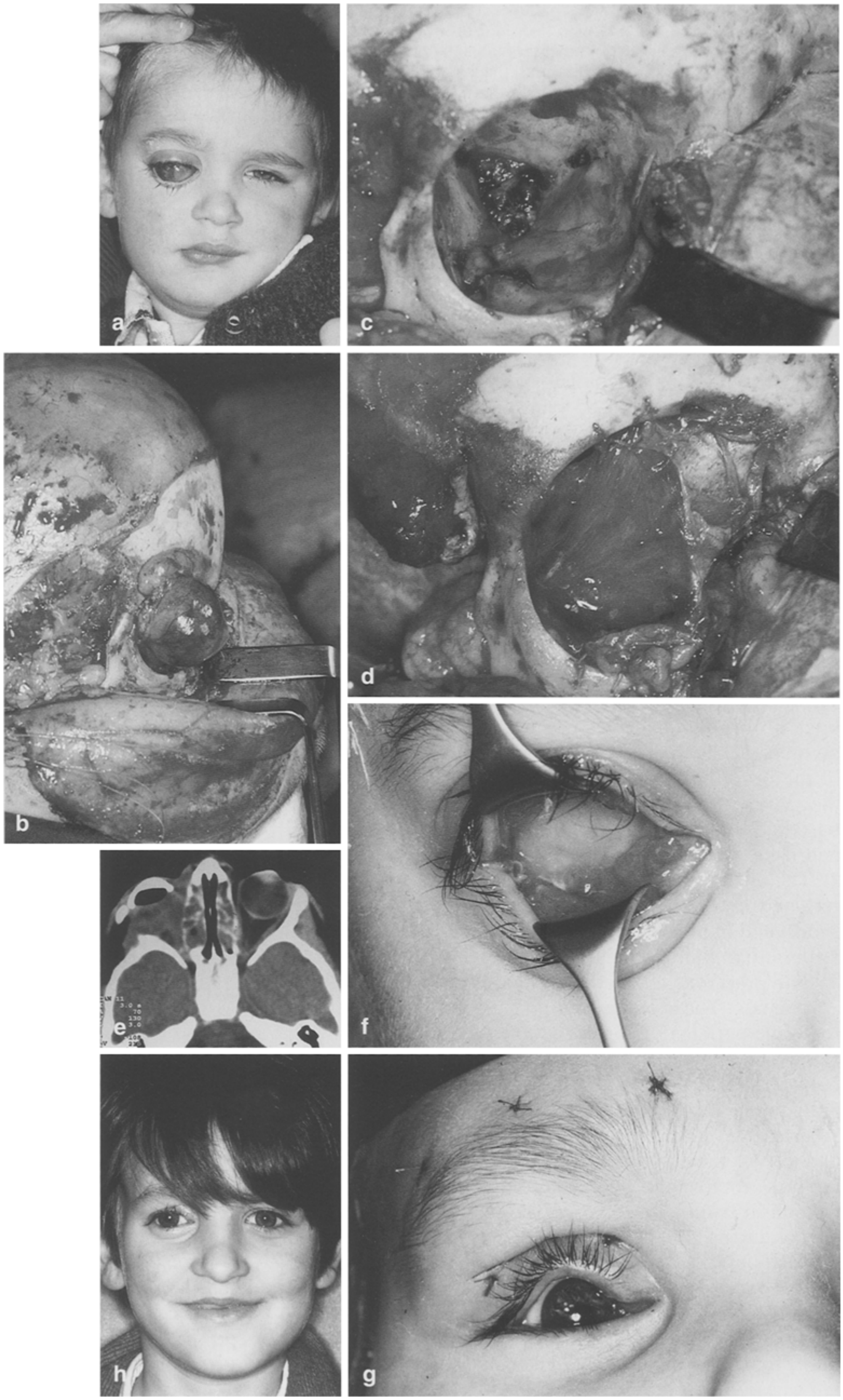

Fig. 1 A 4-year-old patient with a vast, untreated unilateral retinoblastoma (**a**). A bicoronal incision was chosen to provide exenteration without compressing the voluminous tumor mass (**b**). After the complete exenteration of the orbital content (**c**) the empty orbit was filled up with the anterior part of the temporalis muscle through the lateral orbital wall (**d, e**). The resected parts of the bulbar conjunctiva were replaced with a buccal mucosa graft (**f**). The preservation of the eyelids allowed suspension of the upper lid from the frontal muscle with two strips of autologous fascia lata (**g**). After fitting of an artificial eye the final outcome was pleasing. At 45 months after surgery there has been no evidence of local recurrence or distant metastasis (**h**)

Fig. 2 An 81-year old patient with choroidal melanoma and massive orbital involvement (**a**, **b**). The lids had to be removed completely and a forehead flap was designed (**c**) to be shifted into the orbit (**d**). The primarily inserted intraosseous implants were laid open 4 months later (**e**). The covering of the orbit with the completely integrated flap and the implants in the orbital frame provided optimal conditions for the fixation of the episthesis. The follow-up period of 18 months has been uneventful (**f**)

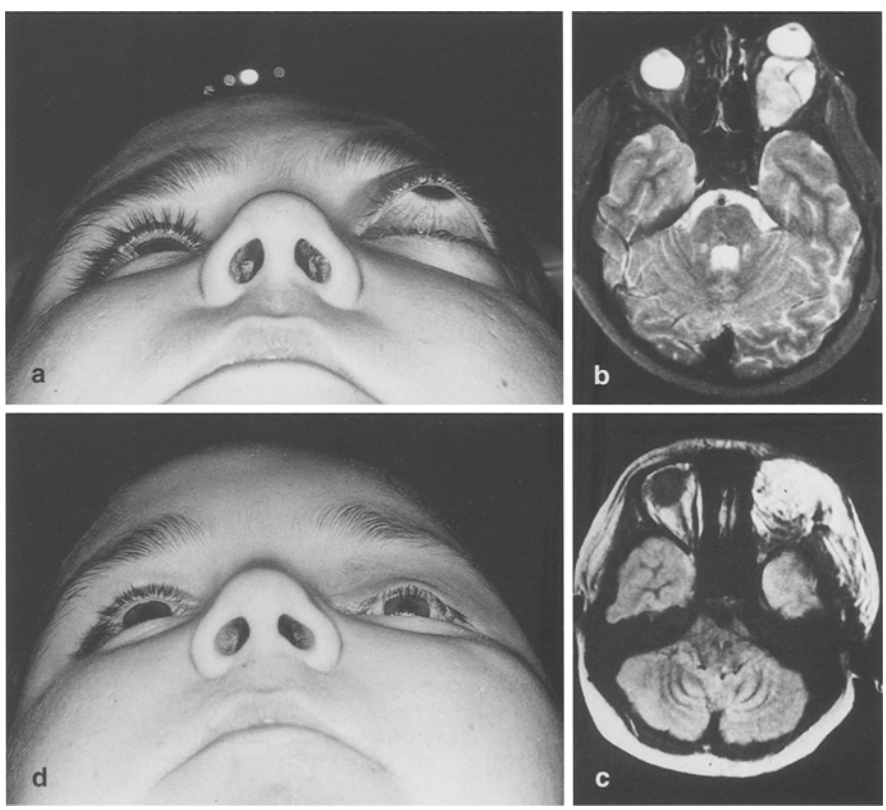

Fig. 3 A 10-year-old girl with massive exophthalmos caused by a huge recurrent embryonal rhabdomyosarcoma (**a**, **b**). After radical removal of the orbital content a free microvascular tissue transplant from the latissimus dorsi muscle was inserted into the enlarged orbital cavity (**c**). The well-perfused tissue of the flap was resistant to shrinking due to the postoperative radiotherapy and allowed fitting of a prosthesis. The girl has remained free of disease for 52 months (**d**)

The orbit was surgically closed in 52 patients. The related reconstructive techniques are listed in Table 3. The orbit was reconstructed with parts of the temporal muscle and closure of the preserved eyelids in 13 patients (Fig. 1). In 14 patients, after full resection of the lids, the empty orbit was filled with cheek or forehead flaps (Fig. 2). Microvascular tissue transplants were used in 14 patients (latissimus dorsi 11, axillary flap 2, parascapular flap 1) (Fig. 3).

Long-term follow-up and survival rate

The prognosis depended on the individual histopathological diagnosis and on the site and extent of the respective tumor, as expected. There were major differences between carcinomas, sarcomas and malignant melanomas as well as between the histopathological subgroups (Table 4). Because of the limited number of patients within the different histopathological groups we refrained from calculating a survival curve for each group of diagnoses.

Rather, we preferred to summarize the survival observations of all 74 patients following exenteration because of an orbital tumor in order to elucidate the curative efficacy of the exenteration procedure (notwithstanding the obvious limitations when throwing together tumors with different histopathology). The average duration of follow-up was 57.8 months (min. 1 month, max. 18 years). Of the tumor cases, 89% survived more than 1 year, 63% more than 5 years and 48% more than 10 years after surgery (Fig. 4). Thirty-three patients died during follow-up (primary tumor-related 23, malignant secondary tumor 3, unrelated to tumor 7).

Twenty-one patients developed local or regional recurrences, occurring in only five cases inside the orbital cavity. In the other cases, the recurrence occurred in the periorbital skin, the maxilla, the sinuses, the neck nodes or intracranially. Nineteen patients suffered from distant metastases.

Most of the cases of local or regional recurrences (14/21) were originally localized outside the orbit. In only 7/21 cases was the primary tumor situated inside the orbit. In 12/21 cases the eyelids had been removed, in 9 cases the lids had been preserved.

In spite of all aggressive surgical approaches the highest rate of recurrence was observed in maxillary carcinomas (13/20). 15 of the patients with maxillary carcinoma died, 13 as a result of the tumor.

The five intraorbitally localized recurrences had the following histology: embryonal rhabdomyosarcoma of an adult, basal cell carcinoma (multiple recurrences), ethmoidal cell carcinoma, extensive maxillary carcinoma, malignant melanoma. The eyelids had been removed in three of these patients (Table 5).

A similar distribution was found with respect to primary tumor-related fatalities: eyelids removed 13, preserved 13. There was no significant difference between the survival curves with and without lid resection. It became clear that the extent and behavior of the tumor determines the outcome of the patient more than the extent of surgical resection.

Table 4 Correlation of histological diagnoses and survival observations for carcinomas, sarcomas and malignant melanomas

Histological diagnosis	Alive	Dead (by tumor)	Mean follow up of surviving patients (months)	Mean survival of deceased patients
Carcinoma (n = 35)				
Adeno carcinoma	0	3	–	47
Adenoid cystic	3	4	69	38
Basal cell	2	4	51	80
Squamous cell carcinoma	5	8 (6/8)	81	16
Others	3	3	10	15
Total	13	22	57	38
Sarcomas (n = 12)				
Rhabdomyo sarcoma	5	1	106	18
Liposarcoma	2	1	58	12
Others	2	1	78	12
Total	9	3	78	14
Malignant melanomas (n=13)				
Conjunctival melanoma	2	1	7	13
Secondary orbital	0	1	–	11
Choroidal melanoma with orbital extension	7	2	21	13
Total	9	4	18	12

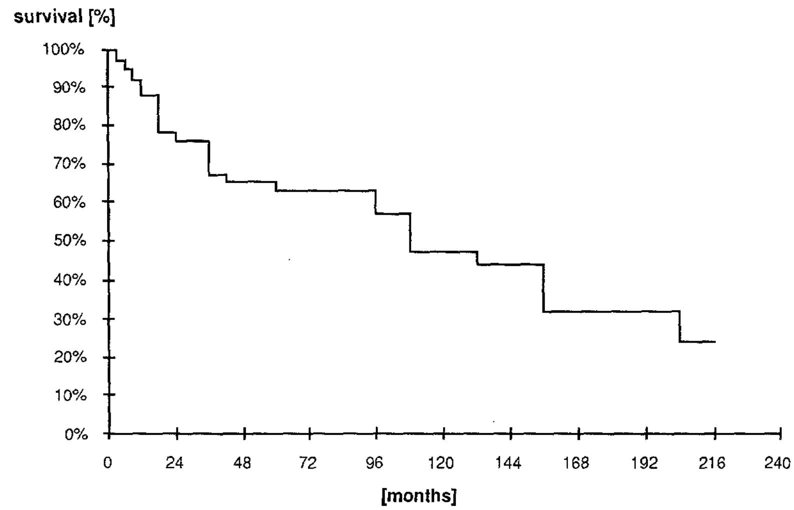

Fig. 4 Survival rate (Kaplan-Meier) of 74 patients after orbital exenteration due to an orbital tumor. Some 63% of patients survived more than 5 years, 48% more than 10 years

Table 5 Correlation of histological diagnosis, surgical technique and survival of five intraorbitally localized recurrences subsequent to orbital exenteration

Histological diagnosis	Intraorbital recurrence	Eyelid resection	Dead from tumor
Rhabdomyosarcoma	1/6	–	+
Basal cell Carcinoma	1/6	+	+
Squamous cell Ca.	2/13	++	++
Choroidal melanoma with orbital extension	1/13	–	+

Discussion

Orbital exenteration is generally reserved for malignancy. The procedure may, however, be required in benign orbital disease or severe orbital trauma [41]. However, 100 of 102 exenteration published by Bartley et al. [5] and 93 of 99 published by Levin et al. [23] had been carried out for malignant orbital tumors. In our study, we found a similar preponderance (74/77) of tumor cases.

The chance of cure by orbital exenteration has been called into question, especially for adenoid cystic carcinomas of the lacrimal gland [18, 35] and for malignant melanoblastomas of the orbit [22, 39]. These conclusions have to be interpreted with caution, however, because of insufficient numbers of patients and a negative bias of the surgically treated patients. Furthermore, no randomized trial has compared the outcomes of radical and conservative treatment for these entities.

Radical exenteration for squamous cell carcinomas of the maxilla with orbital invasion has also been a subject of heated debate. Graamans and Slootweg [16] demonstrated a close correlation between ophthalmological symptoms and pathohistological findings. On the other hand, if the ophthalmological findings were normal, tomographic findings could be misleading, as a comparison with pathohistological and intraoperative findings revealed. In similar cases no negative influence of preservation of orbital contents on survival and local recurrence rates was reported [26, 34, 46].

These statements are of limited value: Perry et al. [34] did not show a life table analysis and explored a selected patient collective (twice as many esthesioneuroblastomas as squamous cell carcinomas). Following globe-preserving surgery, Mann et al. [26] found 7 cases of intraorbital recurrence among 40 patients in spite of postoperative radiotherapy. Secondary enucleation could not be avoided in 25% of their patients. Stern et al. [46] reported severe functional limitations following removal of the orbital floor and recommended orbital exenteration, especially if postoperative radiotherapy is proposed.

Considering the poor prognosis for patients with advanced maxillary cancer (13 of 26 tumor-related deaths in our study involved patients with maxillary carcinomas with orbital invasion), orbital exenteration is only justified if orbital infiltration has been confirmed beyond all doubt. With respect to the quality of life, primary reconstructive surgery is strongly recommended for these patients.

Orbital exenteration has been proved curative for orbital malignancies, and this is endorsed by the high survival rate in our patients. The survival rates in our study are nearly identical with the results of Bartley et al. [5], who reported 88% survival at 1 year and 57% survival at 5 years after surgery. This concordance is especially remarkable in that the eyelids had been preserved in 44% of our patients, the proportion of basal cell carcinomas (which have a comparatively good prognosis) was low, and extended, interdisciplinary frontobasal resections were more frequent in our study. In contrast to Bartley et al. [5], who lined the orbital cavities with split skin grafts or left them to spontaneous granulation in about 75% of their patients, we preferred a primary surgical closure of the cavities in 64% of our patients, with no negative influence on the prognosis.

If orbital exenteration is necessary, the eradication of the tumor has priority over any esthetic considerations. Accompanying resection of orbital bones or parts of the anterior skull base is often unavoidable. In some cases the adjacent dura also has to be resected [8, 9, 33]. The resulting defects are not only a cosmetic problem; the mode of reconstruction determines the functional outcome.

The high rate of sinuorbital fistulas following spontaneous granulation of exenterated orbital cavities in our series (17/25 cases) contrasts with the recommendation that orbital exenteration should generally be performed without surgical closure of the orbit [36]. Few authors give information on this issue: Bartley et al. [5] also found permanent fistulas between the empty orbit and the sinuses (17/49 cases) after spontaneous granulation, though among their cases from the Mayo Clinic bone resection was less frequent. A permanent fistula should be regarded as an unsatisfactory result: we did not find any completely covering epithelium layer in any case of exenteration with accompanying bone removal, and these persisting defects led to an inconvenient secretion of mucus into the orbit with repeated maceration of the epithelium (especially when having a cold) and to a nasal twang of speech. The improved fixation techniques of epistheses by intraosseous implants [1, 31] do not provide a satisfactory alternative; the acceptance of the epistheses rises, but an esthetically, hygienically and functionally adequate long-term result can only be achieved when the underlying orbital cavity has been covered surgically.

An argument widely advanced against primary reconstruction is that recurrent tumors cannot be detected adequately, but this argument does not seem to be valid in light of the low rate of intraorbital recurrences (5/74 cases). Moreover, if a tumor returns after extended surgery the prognosis is very poor. Thus primary closure of the orbit seems advisable to improve the quality of life for these patients by caulking the orbit to the sinuses, the nasal cavity and the oral cavity [15].

Covering the orbital bones with soft tissue is, furthermore, a condition for early postoperative irradiation and an effective protection against precocious lethal complications due to septic meningoencephalitis [5], as well as against late complications such as infectious osteoradionecrosis or cerebral abscess. Roosen [40] underlined that, when dealing with cranio-orbital tumors, covering by soft tissue is the most powerful prophylaxis against CSF fistula, frontobasal osteomyelitis and life-threatening meningoencephalitis. The three most important methods for covering an orbital cavity are the following:

1. The eyelids can be preserved during the exenteration procedure and cover the orbit with epithelial tissue. This technique requires a well-vascularized support for the conjunctiva by pedicled soft tissue (e.g. the temporal muscle [2, 30, 38], the frontal muscle [6], or a galeaperiostal flap [7, 8, 9, 21, 33]) or by free tissue transfer into the orbit [29].

2. Following removal of the eyelids, the orbital cavity can be filled with local or regional flaps. Median forehead flaps [12], frontotemporal flaps [24], retroauricular island flaps [17] and cervicofacial flaps [27] have been recommended for this purpose. Socket reconstruction and even the ability to wear an artificial eye can be achieved by a one-step combination of different local flaps [32] or by step-by-step reconstruction of the eyelids [28].

3. Microvascular free flaps [3, 10, 11, 47] have also been recommended to cover defects following full resection of the eyelids. Facial or temporal vessels may be suitable for microvascular anastomoses.

Our own investigation disclosed that suturing of the conjunctiva provided a tight anterior closure of the orbit if the eyelids could be preserved. Covering with the anterior part of the temporal muscle proved to be sufficient in case of small associated bone defects. Extensive combined defects of the orbit and the surrounding bones necessitate filling with fasciocutaneous or myocutaneous microvascular tissue transfer. Though a surgical repair of the periorbital structures [13] may be worse than an episthesis from the esthetic point of view, surgical covering of an orbital defect is recommended first and foremost to ameliorate the functional result.

The following conclusions may be drawn:

1. An individually tailored surgical procedure of orbital exenteration (if possible with preservation of eyelid skin, tarsal and bulbar conjunctiva) does not influence the risk of local recurrence or tumor-related death.

2. Healing by spontaneous granulation following orbital exenteration should be reserved for tumors of the periorbital skin with infiltration of the eyelids, but without involvement of the orbital bones.

3. Primary surgical closure of the orbital cavity is advantageous with respect to both function and esthetic outcome. It renders early postoperative radiotherapy possible. The risk of surgical covering of the orbital cavity masking a local tumor recurrence can be neglected.

4. Following orbital exenteration, local surgical reconstruction can be achieved by various surgical techniques which have to be adapted to the individual defect.

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