

## ONCOLOGY

# Orbital exenteration in elderly patients: personal experience

## *Exenteratio orbitae in pazienti anziani: esperienza personale*

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## SUMMARY

Orbital exenteration is a disfiguring procedure which typically involves removal of the entire contents of the orbit including the periorbita, appendages, eyelids and, sometimes, a varying amount of surrounding skin. This operation is reserved for the treatment of potentially life-threatening malignancies arising from the orbit, paranasal sinuses or periocular skin. The marked increase in the average life span and resulting greater incidence of invasive malignant skin tumours of the face, typical of old age, is the reason for the increased rate of exenterations in elderly patients. The purpose of this report is to describe personal experience regarding 8 operations of orbital exenteration carried out on elderly patients, 6 males and 2 females, age range 66-85 years (mean 75), who came to our observation, from January 2002 to December 2007, on account of cancer (7 cases: 4 basal cell carcinomas; 1 squamous cell carcinoma; 1 fibrosarcoma; 1 melanoma) or infectious inflammatory disease (1 case of rhinocerebral mucormycosis) and were treated with type III orbital exenteration (2 cases) and type IV orbital exenteration (6 cases according to Meyer and Zaoli's classification). The methods used to reconstruct the eye-socket consisted of a full-thickness skin graft in 5 cases, pedicled myocutaneous flaps in 2 cases – a *latissimus dorsi* muscle flap alone, in one patient, and combined with a *pectoralis major* muscle flap in another – and a combined lateral-based frontal fasciocutaneous pedicled flap and full-thickness skin graft in the oldest patient. Regarding survival and the local clinical situation, 3 of the 4 patients with basal cell carcinomas are alive and disease-free after 6 years, 2 years and 20 months, respectively, while the oldest patient died of the disease after 10 months. The subject who underwent surgery for squamous cell carcinoma is alive and disease-free after 2 years. The patients with melanoma, fibrosarcoma and mucormycosis died. Although there are various options available for reconstruction, full-thickness skin graft or a pedicled musculocutaneous flap provide the simplest solution in the elderly population with significant co-morbidities. The final outcome is, in our experience, comparable to that of more complex flap reconstruction, obtaining very good final results with minimal donor site morbidity and a reduced operation time.

KEY WORDS: Orbit • Paranasal sinuses • Skin malignant tumours • Surgical treatment • Orbital exenteration • Skin graft

## RIASSUNTO

*Nell'ambito della chirurgia oncologica cervico-facciale gli interventi chirurgici di exenteratio orbitae, più o meno allargati agli annessi, palpebre e cute circostante, rivestono un ruolo ed una importanza del tutto particolare per l'alta demolitività della metodica, per le ripercussioni funzionali visivo-spaziali e psicologiche sul paziente, per le problematiche ricostruttive immediate e per quelle più tardive di tipo protesico. Scopo del presente contributo è riportare la nostra esperienza in tema di exenteratio orbitae relativa al periodo compreso tra gennaio 2002 e dicembre 2007 specificando che la classificazione adottata per i diversi tipi di exenteratio è quella di Meyer e Zaoli, del 1971, che distingue quattro interventi di exenteratio orbitae in relazione alla maggiore o minore estensione della demolizione. La casistica comprende 8 pazienti, 6 maschi e 2 femmine (età media 75 anni – range 66-85), sottoposti in 6 casi ad exenteratio orbitae di IV tipo ed in 2 casi ad exenteratio orbitae di III tipo per patologie tumorali e non (7 neoplasie maligne ed un processo infettivo). Le lesioni tumorali erano rappresentate da 6 plurirecidue locali. Istologicamente si è trattato di 4 carcinomi basocellulari a partenza dai canti o/e dalle palpebre, di un fibrosarcoma della cute fronto-nasale, di un carcinoma spinocellulare a partenza dalle vie lacrimali e di un melanoma maligno acromico esteso a tutto l'emivolto sinistro, mentre la patologia infettiva era rappresentata da una mucormicosi rinocerebrale. La riparazione è stata effettuata utilizzando in 5 casi lembi liberi alla Thiersch, in 2 lembi pedunculati mio-cutanei e in un caso l'associazione di un lembo pedunculato fascio-cutaneo frontale a base laterale e di un lembo alla Thiersch. Per quanto riguarda la sopravvivenza e la situazione clinica locale 3 dei 4 pazienti affetti da carcinomi basocellulari sono viventi e NED rispettivamente a 6 anni, 2 anni e 20 mesi mentre la paziente più anziana, comunque affetta da basalioma, è deceduta per malattia a 10 mesi; risulta vivente e NED a 2 anni il malato operato per carcinoma spinocellulare delle vie lacrimali; sono deceduti invece gli altri 3 malati, di cui uno per causa non neoplastica (rottura traumatica della milza per il paziente con mucormicosi). Al momento nessuno dei 5 pazienti viventi e NED ha accettato l'impianto di una protesi oculare.*

PAROLE CHIAVE: Orbita • Seni paranasali • Tumori maligni cutanei • Trattamento chirurgico • Exenteratio orbitae • Innesto cutaneo

## Introduction

Orbital exenteration (OE) is a disfiguring procedure which typically involves removal of the entire contents of the orbit including the periorbita, appendages, eyelids and sometimes a varying amount of surrounding skin.

This operation is reserved for the treatment of potentially life threatening malignancies arising from the orbit, paranasal sinuses or periocular skin<sup>1-5</sup>.

OE results in devastating functional, aesthetic and psychological losses, presenting a reconstructive challenge, especially in elderly patients with significant co-morbidities.

An estimated 40-50% of exenterations are performed for tumours in the eyelid or peri-ocular skin; most of which are basal cell carcinomas and squamous cell carcinomas (SCC), followed by sebaceous-gland carcinomas and melanomas<sup>6-9</sup>. OE is rarely performed for non-neoplastic disease such as trauma or infections<sup>10-12</sup>.

The marked increase in the average life span and resulting greater incidence of invasive malignant skin tumours of the face, typical of old age, is the reason for the increased rate of exenterations in elderly patients.

The purpose of this report is to describe our experience regarding 8 operations of OE carried out on elderly patients for malignant tumours in 7 cases and in one for an infectious inflammatory lesion (1 case of rhinocerebral mucormycosis), in the period from January 2002 to December 2007.

## Patients and methods

The study included 8 patients who underwent different types of OE according to Meyer and Zaoli's classification dating from 1971<sup>13,14</sup>, which classifies four types of OE for tumours in relation to the extent of destruction involved in the surgery:

- Type I: palpebral skin and *conjunctiva* are spared;
- Type II: only the palpebral skin is spared and the eyeball and its appendages are removed with the *conjunctiva*;
- Type III: both eyelids are removed with orbital contents;
- Type IV: the eyeball, eyelids and appendages of the eye are removed with the involved bone structures.

The patients, 6 males and 2 females, aged 66-85 years (mean 75), who came to our observation, in this Division, from January 2002 to December 2007, on account of cancer (7 cases) or infectious inflammatory disease (1 case) were treated with type III OE (2 cases) and type IV OE (6 cases) (Table I).

The pathological conditions in all but one of the patients with tumours consisted of multiple local relapse and comprised: 4 basal cell carcinomas, involving the eyelids in two patients, the right medial canthus in one patient and the skin of the right temporal region and lateral canthus in one very elderly female patient; 1 fibrosarcoma of the fronto-nasal skin; 1 SCC spreading from the tear-ducts; 1 malignant achromic melanoma extending all over the left lateral facial area.

The infectious inflammatory lesion consisted of a rhinocerebral mucormycosis with concomitant involvement of the lung, spreading, as often occurs – and constantly confirmed in the literature<sup>10-12</sup> – to the *endocranium* via the optic nerve and ophthalmic artery.

**Table I.**

Patients	Tumour		Previous treatment	Surgical treatment	Subsequent treatment	Complications	Follow-up
	Hystotype	Site					
1) DCN 76 yrs M	Rhinocerebral mucormycosis	Ethmoid bone, orbital fat, optic nerve, inf. rectus muscle and cavernous sinus	Atypical resection of right inferior basal segment + left superior lobectomy (presence of bilateral "mycetoma")	Type IV left OE + left subtotal maxillectomy extended to nasal fossa and ethmoid bone + repair with abdomen FTSG	Fluconazole (300 mg/die) + Liposomal Amphotericin B (150 mg/die)		DWD after 12 months of traumatic rupture of spleen
2) PG 80 yrs M	Basal cell carcinoma	Eyelids and left medial canthus	Removal of basal cell carcinoma of left superior eyelid; 4 years before; removal of relapsed basal cell carcinoma of left superior eyelid and repair with local flap. 3 years before	Type III left OE + submammary FTSG to cover eye-socket + repair of left periorbital region using sliding flap (from parotid and left cheek)	A) Removal in local anaesthesia of second basal cell carcinoma of right cheek skin + FTSG B) Right parotidectomy extended to Bichat's bulla and skin + right subtotal maxillectomy extended to the zygomatic bone + repair with right GPMF	FTSG moist and hyperemic after 5 years	NED after 6 years
3) FC 75 yrs F	Fibrosarcoma	Skin of right fronto-nasal region invading eye-socket and ipsilateral tear-ducts		Type IV right OE + dorsal nasal skin, right forehead skin, eyelids and right medial canthus, right anterior ethmoidectomy + upper half of tear-ducts + total right parotidectomy + repair using abdomen FTSG	Chemotherapy		DOD after 7 months

(continua)

(Table 1 cont.)

4) OA 73 yrs M	Basal cell carcinoma	Left lower eyelid, ipsilateral lower oblique rectus muscle	Removal of basal cell carcinoma of lower eyelid, 10 and 4 years before	Type III left OE + left lateral canthus skin and left cheekbones skin area + repair with abdomen FTSG	NED after 2 years
5) TD 77 yrs M	Squamous cell carcinoma	Left lacrimal sac	Removal of spinous cell carcinoma of left lacrimal sac, 1 month before	Type IV right OE + left canthus skin, lacrimal sac and ducts + right total ethmoidectomy + repair abdominal FTSG	Liquorrhea NED after 2 years
6) DLE 70 yrs M	Basal cell carcinoma	Right medial canthus, lateral wall of nose, ethmoid bone, orbital floor, ipsilateral maxillary sinus	Multiple removal	Type IV right OE + right total maxillectomy + repair with right LDMF	Liquorrhea NED after 20 months
7) D'OL 66 yrs M	Malignant achromic melanoma	Left parotid region, spreading to left hemimandibular bone, outer ear, eye-socket, maxillary sinus and ipsilateral infratemporal fossa	7 years before, left total parotidectomy with facial nerve preservation and ipsilateral neck dissection (level I-III) (poorly differentiated myoepithelial malignant tumour) 6 years before, RT on left parotid space and ipsilateral neck 4 years before, chemotherapy with cisplatin and 5F-uracile for left parotid relapse 3 years before, dissection of left parotid recurrence - reconstruction with sliding flap (melanoma) 2 years before, left anterior maxillectomy for melanoma relapse	Type IV left OE + parotid gland, hemimandibular bone, external and middle ear and left infratemporal fossa + repair with myocutaneous pedicled flaps of left LDMF and PMMF	Liquorrhea DOD after 4 months for cachexy
8) PMR 85 yrs F	Basal cell carcinoma	Skin of temporal region and right lateral canthus	Multiple removal + RT	Type IV right OE with dissection of right frontal maxillary bone and cheekbones + repair right lateral forehead fasciocutaneous pedicled flap + abdominal FTSG	DOD after 10 months

M: male; F: female; RT: radiotherapy; OE: orbital exenteration; FTSG: full-thickness skin graft; LDMF: *latissimus dorsi* muscle flap; PMMF: *pectoralis major* muscle flap; NED: non-evident disease; DWD: died without disease; DOD: died of disease

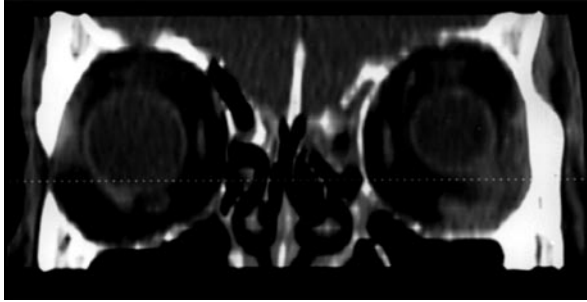


Fig. 1. CT scan of orbits – Patient 4.



Fig. 2. MRI scan of orbits – Patient 3.

In all cases, an OE was programmed after performing a computerised tomography (CT) scan (Fig. 1) and magnetic resonance imaging (MRI) (Fig. 2) of the upper maxillofacial bones, an ophthalmological consultation with Hess's screen and/or echography of the eye-socket, biopsy sampling and exclusion, in the cases of malignant, non-basal cell tumours, of any remote metastases.

It should be pointed out that sight had been almost completely lost in 5 of the eyeballs removed (4 of which for tumours and 1 for mucormycosis; in these cases, imaging techniques gave clear confirmation of eye-socket invasion), while sight was maintained in 3 cases, which, however, also presented clinical and radiological evidence of invasion of the tear-ducts, extrinsic muscle and fat tissue of the eye-socket. All cases had normal sight contralaterally.

Surgery was extended to adjacent skin areas and ipsilateral organs on the basis of tumour spread (2 total parotidectomies; 1 hemimandibulectomy; 4 maxillectomies, 2 of which total and 2 subtotal; 3 anterior ethmoidectomies) (Table I), while the methods used to reconstruct the eye-socket consisted of a full-thickness skin graft (FTSG) in 5 cases, pedicled myocutaneous flaps in 2 cases – a *latissimus dorsi* muscle flap (LDMF) alone in one patient and combined with a *pectoralis major* muscle flap (PMMF) in another – and a combined lateral-based frontofasciocutaneous pedicled flap and FTSG in the oldest patient.

No patients underwent complementary radiotherapy (although 2 had previously received radiation).

## Results

All the patients recovered satisfactorily after surgery without serious complications in the immediate post-operative period.

Regarding survival and the local clinical situation, 3 of the 4 patients with basal cell carcinomas are alive and disease-free after 6 years, 2 years and 20 months, respectively, while the oldest patient died of the disease after 10 months. The patient who underwent surgery for SCC of the tear-ducts is alive and disease-free after 2 years. The other 3 patients died; the one with melanoma died of cachexy, the fibrosarcoma patient on account of widespread disease and the mucormycosis patient due to traumatic rupture of the spleen. Death occurred after 4, 7 and 12 months, respectively (Table I). As far as concerns complications, 3 cases presented slight liquorrhea during the operation because of a fracture of the sphenoid bone at the apex of the orbit. The fistula was immediately repaired by padding the fracture spot with reabsorbable material and the fluid loss ceased instantly or within a few days (although it persisted in the form of rhinoliquorrhea for 5-6 days in the 2 patients in whom it was associated with total maxillectomy).

The myocutaneous pedicled flaps and the frontal fasciocutaneous flap transposed to the eye-socket took without any problem. The period of normalisation for the FTSGs was, instead, relatively long, since they required repeated medication and cleaning to remove scabs and granulations. Only one diabetic patient presents a graft which is, in some parts, moist and hyperaemic after 5 years. Two patients with a basal cell lesion – one of the eyelid and the other of the medial canthus – underwent surgery again, one after nearly 4 years, first under local and then general anaesthesia, to remove another basal cell tumour which had deeply invaded the parotid gland, maxillary sinus and contralateral cheekbones, and the other patient to reshape the “extra” myocutaneous part of the dorsal flap transposed to the eye-socket.

In the patient with a basal cell tumour which developed contralaterally to the previous OE, a right PMMF had to be prepared during the repair stage, transposed to the ipsilateral region between the nose and cheek.

The surviving patients were offered a prosthesis, but to date none of them has accepted to undergo this procedure.

## Discussion

In the field of neck and face oncology, few operations are as destructive as OE, or as unpleasant to perform. Our patients were prevalently elderly males. Six out of 7 neoplasms were multi-relapsing tumours that eventually involved the eye-socket, so that complete removal of the eyeball, extended in varying degrees to surrounding structures, was the only radical and permanent solution.

In fact, the OEs were carried out for tumours not strictly of the eye but deriving from the eyelids, appendages and periorbital skin, and surgery often involved the removal of adjacent structures and organs (maxillary sinus and/or anterior ethmoid, parotid gland, lymph nodes, etc.), where the disease subsequently spread by direct invasion or metastasis.

In this regard, it is important not to underestimate malignant facial skin tumours, even in elderly patients and even if they are basal cell cancers (4 cases in our experience), since they can become recurrent if treated ineffectively and may

spread, locally and deeply, towards the eyelids and medial or lateral canthus, outside the range of straightforward objective examination and imaging techniques<sup>15</sup>.

Meyer and Zaoli's classification was used as this appears complete and suitable, but some other classifications should also be mentioned; in 1980, Curioni<sup>16</sup> classified nasal-paranasal malignant tumours with orbital involvement into five groups of surgery related to the primary site of the neoplasm (medial wall, lateral wall, centre of the orbit, orbital floor or inferior part of the maxillary sinus; midfacial region between orbits). Cordeiro and Santamaria<sup>17</sup>, in their classification system for maxillectomy and midfacial defects, propose four basic types of resections of the maxilla for tumours including the OE in the treatment of cases of advanced cancers of the maxillary sinus invading the orbit. The maxillary and midfacial defects are classified as follows: type I, limited maxillectomy; type II, subtotal maxillectomy; type IIIa, total maxillectomy with preservation of the orbital contents; type IIIb, total maxillectomy with orbital exenteration; type IV, orbitomaxillectomy.

Goldberg<sup>18</sup> redefined OE as the extensive removal of orbital and peri-orbital tissues in which the primary goal of surgery is the complete extirpation of an orbital disease process, with the preservation of vision being a secondary consideration.

Typically, this procedure will include the removal of the periorbita and the establishment of a bony plane at least in one quadrant of the orbit. Ben Simon<sup>15</sup> reported a classification of OE as subtotal when partial removal of orbital tissue, with sacrifice of the eye, is performed, as total when all orbital contents including the globe and periorbita are removed and as extended exenteration when the excision includes the adjacent bones.

Yeatts<sup>19</sup> divided OE into two categories: total exenteration as removal of the entire orbital contents with or without sacrifice of eyelid skin, and subtotal exenteration as partial removal of orbital tissues with sacrifice of the eye, which can be considered as an extended enucleation.

According to Meyer and Zaoli's classification, the lesions we treated always required type III or type IV OE and only in the 2 cases undergoing type III did the operation not involve other structures in addition, obviously, to the eyelids. Among our patients, it is worthwhile mentioning the patient with mucormycosis who had no sight in the eye and

necrosis of the entire maxillary bone and part of the ethmoid bone. This made surgical intervention easier; it was, in fact, limited to removal of the soft orbital tissues with low blood perfusion and parts of the face bones that had already been sequestered for some time. The final histological examination showed thrombosis of the ophthalmic artery due to mycosis and atrophy of the optic nerve. In fact, in this particular type of infectious condition, the mycete is known to proliferate rapidly and become pathogenous, especially in immune-depressed subjects (our patient was a long-term diabetic in a critical condition), and pass through the ophthalmic artery and optic nerve, travelling from the nose and paranasal system via the eye-socket and reaching the encephalon, thus changing a "rhinogenous" infection into a dangerous "rhinocerebral" process. All this makes surgery essential and an OE almost mandatory, followed by long-term anti-mycotic therapy, peripherally by intravenous infusion and orally<sup>10-12</sup>. It is also worthwhile recalling that in many cases, as in our patient, rhinocerebral mucormycosis is associated with pulmonary mycetomas (the patient had previously undergone bilateral surgery for that condition).

Concerning the phase of repair work after OE, a large number of flaps are needed to cover the orbital bones: the temporalis muscle pedicled flap, the galea fascia or pericranial flap, the myocutaneous pedicled flap and the revascularized free flap<sup>20-30</sup>.

The temporalis muscle flap is one of the most frequently used flaps to obliterate the orbital cavity but only a small portion of the muscle can be used for this purpose because most of the muscle is used as the pedicle<sup>26</sup>. To solve this problem, it is necessary to create a large window in the lateral orbit through which to pass the pedicle, without resection of the lateral orbital rim<sup>27</sup>.

The galea fascia flap<sup>30</sup>, pericranial flap, skin graft and osseointegrated implants can also be used<sup>21</sup>, without forgetting the myocutaneous pedicled flaps, particularly the PMMF, which Ariyan<sup>20</sup> first described and used in 1979 to repair 2 orbital defects after OE.

The most used microvascular free flap, after OE, with or without total maxillectomy, is the *rectus abdominis* myocutaneous free flap with one or more skin islands<sup>31-33</sup>.

In our experience, due to the advanced age of the patients and the poor general conditions (average age 75 years;

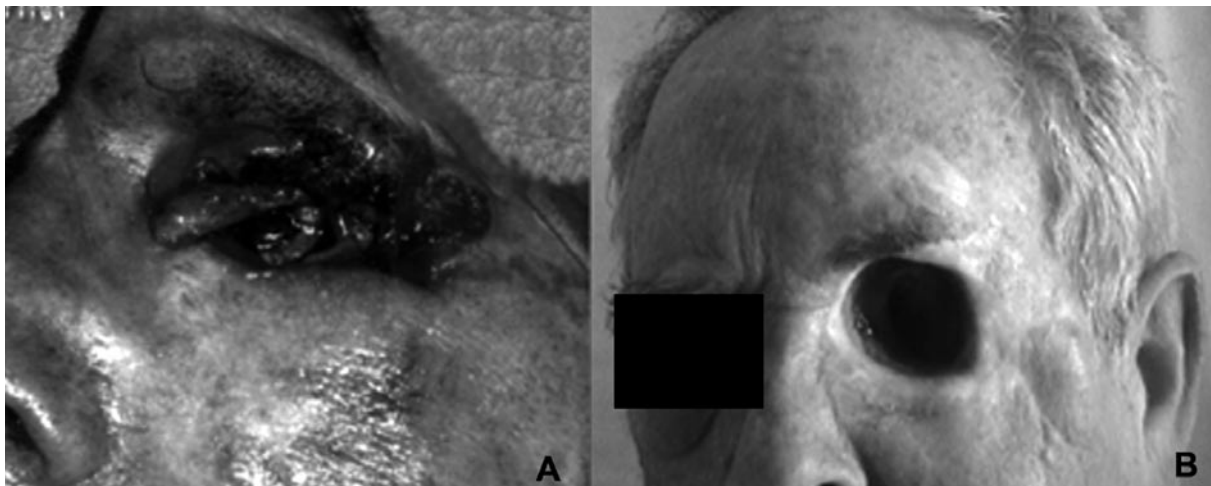


Fig. 3. A) Pre-operative view; B) post-operative view – Patient 2.

important metabolic diseases), we employed a FTSG in 5 cases, a myocutaneous pedicled flap in 2 cases and a combination of a fasciocutaneous pedicled flap (lateral-based frontal flap) with a FTSG in one case.

If not too much of the bone and periorbital cutaneous structures have been destroyed and there seems to be some “support” in the middle third of the face, the problem can be solved easily and excellently with a FTSG, although a certain amount of time is needed for it to take completely in the eye-socket and for scabs and granulations to be removed at the points of suture (Fig. 3).

Pedicled myocutaneous flaps were employed in 2 cases of total maxillectomy because the orbital floor, which supports the graft, was missing, and in the patient with achromic melanoma, it was necessary to compensate for particularly extensive loss of bone and subcutaneous and cutaneous substance.

Regarding the cases with a complication, the spinal fluid loss probably occurred because of micro-fractures created during ligation of the orbital pedicle or during the ethmoidectomy. However, as already mentioned, immediate repair of the fistula brought the situation back to normal within a few days.

## Conclusion

All the patients in whom OE was performed were over 65 years of age. They were mostly males and affected by cancer (basal cell carcinoma in 4 out of 7 cases).

Tumours were almost all multi-relapsing neoplasms and for these advanced periorbital malignant skin cancers, other treatment modalities, such as radiation or chemotherapy, were considered unlikely to lead to recovery.

For the reconstruction, obliteration of the orbital cavity and continuation of the epithelial lining are required. Although there are various options available for reconstruction, a full-thickness skin graft or a pedicled myocutaneous flap provide the simplest solution in the elderly population with significant comorbidities. The final outcome is, in our experience, comparable to that of more complex flap reconstruction, obtaining very good final results with minimal donor site morbidity and a limited operation time.

The only important surgical complication, in a few of our cases, was spinal fluid loss, but immediate repair of the fistula with reabsorbable material solved the problem instantly or within a few days.

In elderly patients, advanced periorbital malignant skin cancers, which are often relapsing, must be treated radically to avoid the risk of orbital invasion.

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