

# The Epidemiology of Orbital Exenteration in Southern Iran

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## Abstract

**Background:** Exenteration is performed as an infrequent procedure for the treatment of locally invasive or potentially life threatening orbital neoplasm, when less invasive management are inadequate. This study was performed to report the epidemiological features of diseases treated by orbital exenteration as well as surgical techniques used in south of Iran.

**Methods:** In a retrospective study, records of all patients' undergone exenteration of the orbit between years 1986 and 2006, at the Khalili Teaching Eye Hospital were reviewed. Correspondingly, records of pathology were also searched for the same cases to confirm the pre-operation diagnosis in the charts.

**Results:** There were 28 men and 20 women with a mean age of  $50 \pm 23.9$  years. Basal cell carcinoma (31%), squamous cell carcinoma (18%) and malignant melanoma (14%) were found to be the frequent ones. Consequently, eyelid was reported as the anatomic site for origin of tumors in 41.6% of patients. The exenterated cavity was left to heal by granulation tissue and epithelialization in 32 cases (66%), skin graft was placed in 13 cases (27%) and tissue flap were prepared for reconstruction in the remaining three cases.

**Conclusion:** Basal cell carcinoma with secondary orbital involvement was the most common cause of orbital exenteration and healing by the granulation tissue was the main rout of reconstruction in our cases.

**Keywords:** Epidemiology; Orbital exenteration; Iran

## Introduction

Tumors or diseases of orbit may be originated primarily from the orbit or involve it from adjacent tissues or metastatic sources.<sup>1</sup> Exenteration is performed as an infrequent procedure worldwide. It is a complex disfiguring procedure reserved for the treatment of locally invasive or potentially life threatening orbital neoplasms, when less invasive management are inadequate: however, this sophisticated procedure is not considered as a curative one in most cases without adjunctive therapy.<sup>2</sup> There are several techniques for reconstructive surgery after exenteration.<sup>3,4</sup> These include local, re-

gional and distant techniques. Local reconstructive techniques such as spontaneous granulation and split-thickness skin graft are more commonly used. Local reconstructive techniques are easier to perform, and recurrences are more simply detected compared with other techniques.<sup>5</sup>

Regional reconstructive technique is the temporal muscle transfer procedure, forehead and cheek flaps while distant reconstructive techniques are transfer procedure of latissimus dorsi or pectoralis flaps.<sup>6-10</sup> On the other hand, leaving the orbital cavity to heal with the granulation tissue has proved to be a relatively safe technique. Since, there were no particular complications related to granulation tissue reported till now, regional and distant reconstructive techniques were applied only when large defects were created and primary closure was necessary for the prevention of complications or for cosmetic goals.<sup>11</sup> In comparison to split-thickness skin graft, healing

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by spontaneous granulation leads to a shallower orbital cavity. Unfortunately, skin grafting on bone in the orbital cavity results in heavy desquamation and chronic discharge which is not seen with spontaneous granulation.<sup>12</sup> The single disadvantage of spontaneous granulation is that healing takes longer and dressing changes are required more frequently.<sup>12</sup> According to these various techniques of reconstructions, it is reasonable to search about outcome of these procedures in different centers. Our attempt was to report a center experience about the epidemiological features of diseases treated by this procedure as well as surgical techniques used in this relation.

### Materials and Methods

In a retrospective study, records of all patients undergone exenteration of the orbit between years 1986 and 2006, at the Khalili Teaching Eye Hospital (A major eye referral center in south of Iran) were reviewed. We found about 71 cases of exenterations according to the operation index book in the Khalili Hospital; however, after triple checking, just records of 48 patients were available. Correspondingly, pathology records were also searched for the same cases to confirm the pre-operation diagnosis in the documents.

### Results

There were 28 men and 20 women with a mean age of  $50 \pm 23.9$  years (range: 5-80 years). Exenterations were performed on the right and the left sides in 27 and 21 patients respectively. While in 36 patients, only simple orbital exenteration was done, 12 patients had extended exenterations due to more invasive pathologies such as basal cell and squamous cell carcinomas with extensive involvement of preorbital tissues. Subsequently, the exenterated cavity was left to heal by granulation tissue and epithelialization in 32 patients, skin graft was placed in 13 patients and tissue flap was prepared for reconstruction in the remaining three patients. Although different neoplasms were responsible for exenterations, basal cell carcinoma which was reported in 15 out of 48 cases (31%), squamous cell carcinoma in 9 out of 48 (19%) and malignant melanoma in 7 out of 48 (14%), were found to be the frequent ones (Table 1). Consequently, eyelid was reported as the anatomic site for origin of tumors in 41.6% of patients (20 out of 48). So, it was considered as the most common site of origin followed by the globe in 9 out of 48 (18%), para nasal sinuses in 7 out of 48 (14%), orbit in 4 out of 48 (8%), conjunctiva in 3 out of 48 (6%) and intracranial space in 2 cases (4%). Moreover, hard palate and the optic nerve as well as the lacrimal glands were found to be responsible for the origin of tumors in three patients

**Table 1:** Diseases found to be as the main causes of exenteration in 48 cases according to the reports of pathology

Site of origin	Tumors or Disease	Numbers
Eyelid	BCC	14
	SCC	6
Globe	Retinoblastoma	5
	Malignant melanoma	4
Para nasal sinuses	Malignant melanoma	2
	SCC	1
	Rhabdomyosarcoma	1
	Wegner's	1
	Small cell malignant tumor	1
Orbit	Ameloblastoma	1
	Chronic inflammation	2
	Hemangioendothelioma	1
	Optic nerve meningioma	1
Conjunctiva	SCC	2
	Malignant melanoma	1
Intra cranium	Meningioma	2
Palate	SCC	1
Scalp	BCC	1
Lacrimal gland	Mucoepidermoid carcinoma	1

separately (Table 2).

According to the tumor types, there were 15 patients of basal cell carcinoma and 10 patients with squamous cell carcinoma originated from scalp and eyelids, para nasal sinuses, conjunctiva and hard plate. Moreover, there were seven cases of malignant melanoma originated from the globe, para nasal sinuses, and conjunctiva. Further, among three cases of meningioma reported, two were originated from the intracranium and the other one originated from the optic nerve. Chronic inflammations of orbit because of unknown cause were reported in two cases. Other rare causes of exenterations were rhabdomyosarcoma, Wegner's granulomatosis, hemangioendothelioma, ameloblastoma, mucoepidermoid carcinoma and small cell carcinoma (Table 2).

## Discussion

To our knowledge, our case series were the first unique review of orbital exenteration in Iran. orbital exenterations are mainly performed in this center as it is one of the important patients' referrals in south of Iran. As was mentioned, most of the cases (72 %), undergone exenteration, were those with secondary orbital tumors and only 29% were originated primarily from the orbit and globe. Eyelid tumors were the most frequent origin for secondary orbital tumors

(41.6%), followed by the globe (18.7%) and paranasal sinuses (14.5%). In the same manner, Bartley *et al.* introduced the secondary tumors invading the orbit as the main cause of exenterations; however, in their study, eyelid and paranasal sinuses/nasal cavity shared nearly equal percent as the causes of exenterations i.e. 28% and 26% respectively, while globe accounted for about 15% of the total orbital tumors.<sup>15</sup> Moreover, in the present study, basal cell carcinoma, originated from the eyelid and scalp, was the most frequent tumor type (31.25%) for which orbital exenteration has been carried out similarly. Rahman *et al.* found that the basal cell carcinoma was the most frequent tumor type leading to exenterations,<sup>13</sup> while it is in the opposite of other previous reports that squamous cell carcinoma were the most important leading causes of exenteration.<sup>2,14,15</sup> Squamous cell carcinoma invading the orbit was the second cause of exenteration in our series. There were 10 squamous cell carcinomas comprising 20.8% of all lesions. In our series, eyelid was found to be the most common site of origin of squamous cell carcinoma (12.5%) followed by conjunctiva (4.1%), paranasal sinuses (2.08%) and hard palate (2.08%). Therefore, the results were nearly correlated with Levin *et al.* study, who reported eyelid as the origin of tumors in 12.1% and paranasal sinuses in 13.1% followed by conjunctiva (6.06 %) and lacrimal sac (1.01%).<sup>14</sup> Malignant melanoma invading the orbit was the third

**Table 2:** Breakdown classification of the tumors and diseases as causes of exenterations in the overall 48 numbers of cases according to the site of origin

Tumors or diseases	Primary sites of origin	Numbers
BCC	Eyelid	14
	Scalp	1
SCC	Eyelids	6
	Conjunctiva	2
	Para nasal sinuses	1
	Hard palate	1
Malignant melanoma	Globe	4
	Para nasal sinuses	2
	Conjunctiva	1
Retinoblastoma	Globe	5
Meningioma	Intra cranium	2
	Optic nerve	1
Chronic inflammation	Orbit	2
Rhabdomyosarcoma	Para nasal sinuses	1
Wegner's granulomatosis	Para nasal sinuses	1
Hemangioendothelioma	Eyelids	1
Ameloblastoma	Para nasal sinuses	1
Mucoepidermoid carcinoma	Lacrimal gland	1
Small cell carcinoma	Para nasal sinuses	1

common cause of the exenteration in our series. The original site of melanoma invading the orbit was from the globe, followed by paranasal sinuses and conjunctiva. In the literature, malignant melanoma of the conjunctiva and choroids accounted for the most of the exenteration performed for malignant melanoma.<sup>14-16</sup>

In four major reviews, the proportion of patients with benign diseases undergoing exenteration was between 3% and 17%.<sup>1,14-16</sup> In our study, there were eight benign diseases (three meningioma, two pseudo tumor, one Wegner's granulomatosis, one hemangiopericytoma and one ameloblastoma) accounting for 16.6 % of all exenterated cases. It should be emphasized that exenterations is not a routine treatment in inflammatory pseudo tumor except the time that patient is severely incapacitated because of pain, progressive proptosis and visual loss.

As was mentioned, several techniques are currently used for reconstructive surgery after exenteration.<sup>3,4</sup> In our center, the exenterations cavity was

allowed to heal by granulation and epithelialization in 32 cases (66%), skin graft was placed in 13 cases (27%) and tissue flap was used only for the reconstruction in the three cases. In other words, selection of the granulation tissue as the main route of reconstruction in our cases that can be interpreted logically by the fact that our patients were old and did not seek for cosmetic results.

Basal cell carcinoma with secondary orbital involvement was the most common cause of orbital exenteration and healing by the granulation tissue was the main route of reconstruction in our cases.

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**Conflict of interest:** None declared.

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