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Causes of enucleation in Northern India (1995-2005)

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PURPOSE. *There are limited data concerning the reasons for surgical removal of eyeball. The present retrospective study was undertaken to evaluate the frequency of diseases requiring surgical removal of eyeball and to study the histopathologic changes in these specimens.*
METHODS. *Forty-eight surgical eyeball specimens received between January 1999 and June 2005 were included in the study. Age, sex, and clinical diagnoses were recorded in each case. Specimens were classified on the basis of surgical procedure. Gross and microscopic findings were noted for all the specimens.*
RESULTS. *Between January 1995 and June 2005, there were 139,092 outpatients, 6,574 hospital admissions, 12,044 ophthalmic operations, and a total of 48 enucleations in 47 patients. Of these 47 patients, 24 were male and 23 female with almost equal male:female ratio. Right eye was involved in 29 cases while left was involved in 17 cases. Bilateral eye involvement was seen in one case. Surgical specimens included enucleation (28 cases, 58.3%), exenteration (12 cases, 25%), and evisceration (8 cases, 16.6%). On histopathologic examination, the lesions were categorized into two broad groups: neoplastic (8 cases, 16.6%) and non-neoplastic (40 cases, 83.4%). Both groups were further subcategorized and correlated with clinical diagnosis.*
CONCLUSIONS. *In our setting, non-neoplastic lesions are the main cause of eyeball surgery, as compared to the West, where trauma followed by neoplasms constitute important causes. Pathologic examination of eyeballs is a must for proper postoperative management of the patient. (Eur J Ophthalmol 2007; 17: 638-41)*

KEY WORDS. *Eyeball, Enucleation, Evisceration, Exenteration, Neoplastic, Non-neoplastic*

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INTRODUCTION

Ocular disorders are responsible for a great deal of morbidity. There are limited data concerning the reasons for surgical removal of eyeball (1). Western literature reports cite trauma as the most common condition leading to removal of eyeball, followed by ocular neoplasia (2, 3). The present retrospective study was undertaken to evaluate the frequency of diseases requiring surgical removal of eyeball and to study the histopathologic changes in these specimens.

MATERIALS AND METHODS

All surgical eyeball specimens received in the Department of Pathology, Government Medical College, Chandigarh, between January 1999 and June 2005 were included in the study. Age, sex, and clinical diagnoses were recorded in each case. Specimens were classified on the basis of surgical procedure: enucleation, evisceration, or exenteration. Histopathologic diagnosis was recorded for each case. Underlying disease process which eventually resulted in surgical removal was retrospectively determined in

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each case using all available clinical and pathologic information.

RESULTS

During the period between January 1995 and June 2005, there were 139,092 outpatients, 6,574 hospital admissions, and 12,044 ophthalmic operations (including minor and major surgeries). There were a total of 48 surgical eyeball specimens from 47 patients during this 6-year study period. Age of the patients ranged from 5 months to 80 years. There were 24 male and 23 female patients with almost equal male:female ratio. Right eye was involved in 29 cases while left was involved in 17 cases. Bilateral eye involvement was seen in one case. Surgical specimens included enucleation (28 cases, 58.3%), exenteration (12 cases, 25%), and evisceration (8 cases, 16.6%) (Fig. 1).

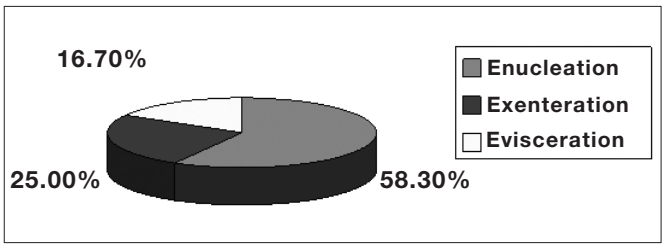

Fig. 1 - *Types of surgical specimens.*

On histopathologic examination, the lesions were categorized into two broad groups: neoplastic (8 cases, 16.6%) and non-neoplastic (40 cases, 83.4%). Both the groups were further subcategorized and correlated with clinical diagnosis (Tab. I, Fig. 2).

There was 100% clinicopathologic correlation in the neo-

TABLE I - CLINICO-PATHOLOGIC CATEGORIZATION OF NEOPLASTIC AND NON-NEOPLASTIC EYEBALL LESIONS

Subject no.	Clinical diagnosis (no. of cases)	Pathologic diagnosis	No. of cases with final pathologic diagnosis
	Neoplastic		**(n=8/48, 16.7%)**
1	Retinoblastoma (5)	Retinoblastoma	5
2	Squamous cell carcinoma (3)	Squamous cell carcinoma of conjunctiva	3
	Non-neoplastic		**(n=40/48, 83.4%)**
1	**Inflammatory/infectious**	**Inflammatory/infectious**	**19 (39.58%)**
	Endophthalmitis (2) Retinoblastoma (1) Painful blind eye (1) Trauma (1)	Endophthalmitis	5
	Panophthalmitis (3) Malignancy (1) Trauma (1)	Panophthalmitis	5
	Rhino-orbito zygomycetes (9)	Zygomycosis	8
		Zygomycosis + aspergillus	1
2	Anterior staphyloma (7)	**Anterior staphyloma**	**7 (14.58%)**
3		**Developmental**	**2 (4.17%)**
	Retinoblastoma (1)	Retinal dysplasia	1
	Retinal dysplasia (1)	Retinal dysplasia with PHPV	1
4	Phthisis bulbi (3)	**Phthisis bulbi**	**3 (6.25%)**
5	Traumatic injury (2)	**Traumatic injury**	**2 (4.17%)**
6	Spontaneous globe rupture (2) Perforated corneal ulcer (1)	**Descriptive**	**3 (6.25%)**
7	Normal donor eyeballs (4)	**Normal eyeballs**	**4 (8.33%)**

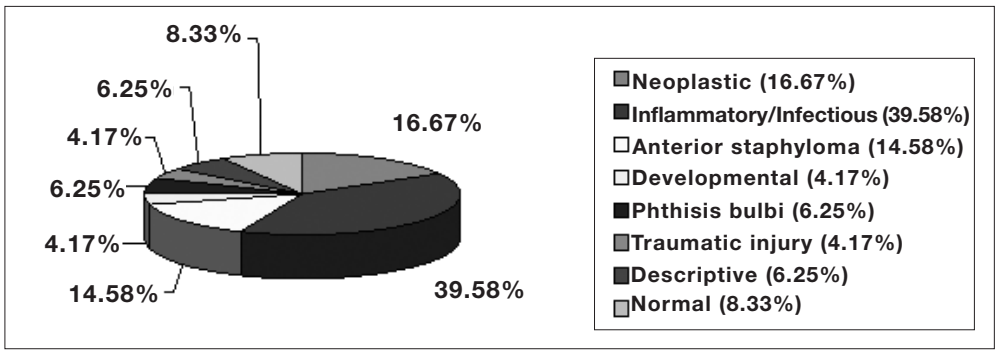

Fig. 2 - *Clinico-pathological categorisation of neoplastic and non-neoplastic lesions.*

plastic group with a clinical diagnosis of retinoblastoma and squamous cell carcinoma. Out of five cases of retinoblastoma, three were well-differentiated based on rosette formation and two were poorly differentiated. Three cases showed tumor infiltration into optic nerve. Ocular surface tumor included three cases of squamous cell carcinoma arising from conjunctiva and invading into ocular coats.

Non-neoplastic lesions constituted the major group (40/48 cases, 83.4%). Nineteen out of 48 cases (39.58%) were of inflammatory/infectious etiology. There were five cases each of suppurative endophthalmitis and panophthalmitis. History of trauma in two cases, ulcerated corneal ulcer in six cases, and diabetes mellitus in one case was present. One case was clinically suspicious of retinoblastoma. Orbital zygomycosis was seen in nine cases, one case showed both aspergillus and zygomycosis. Out of nine cases, five showed infiltration of fungal hyphae into the layers of eyeball. Fungal inflammation was one of the major causes of exenteration.

The second commonest lesion in the non-neoplastic group was staphyloma, constituting 14.48% (7/48) cases. History of trauma and perforated corneal ulcer was present in one case each. Phthisis bulbi (3/48 cases, 6.25%) was characterized by shrunken hard eyeball and ossification was noted in two cases. There were two cases of retinal dysplasia, out of which one was clinically suspicious of malignancy. In addition, there was primary hyperplastic vitreous in one case. Traumatic injury was cause of enucleation in two cases. In three cases no definite diagnosis could be given on pathologic examination. In four cases normal donor eyeballs without cornea were submitted, which did not reveal any significant pathologic change.

DISCUSSION

Surgical removal of the eyeball is a major procedure and indications vary across different parts of the world. Most of the studies from Western literature have reported traumatic injury as a leading cause of surgical eyeball removal followed by tumors (3, 4). There are limited Indian data on causes of enucleation/evisceration/exenteration of eyeballs.

In the present study, tumors constituted only 16.6% of cases of surgical resection of eyeballs. A study by Vemuganti et al (1) reported tumor as a cause of enucleation in 49% of cases while other studies have reported incidence of 20–34% (5). Out of eight malignant cases, five were retinoblastoma, which other studies have also reported as one of the important causes of enucleation. Also, there is a reported rise in the incidence of retinoblastoma in India (6). Invasive squamous cell carcinoma of conjunctiva is rare and intraocular extension is even rarer (7). Local excision with wide surgical margin is the treatment of choice; however, for intraocular invasion, enucleation is required and for intraorbital extension exenteration is mandatory.

In the non-neoplastic group, inflammation/infectious etiology was a prominent cause of surgical eyeball excision, constituting 39.58% of cases (19/48). Similar higher incidence of inflammation/infections as cause of eyeball resection was reported by Kitzmann et al (2). Trauma, ulcerative keratitis, and complications of diabetes mellitus leading to endophthalmitis have been reported. All nine cases of fungal endophthalmitis were secondary to rhino-orbito-cerebral mucormycosis, which is known to exist in two forms, the acute form and the less well-recognized chronic form.

Anterior staphyloma was the next most common indication in the non-neoplastic group for enucleation, consti-

tuting 14.58% of all cases. Possible factors implicated in pathogenesis include old trauma, postoperative state, malnutrition, corneal ulceration, or perforation. In the present study, the main pathogenetic factors were corneal inflammation and old trauma (1, 8). Trauma has been a leading cause of enucleation and evisceration, as reported in the literature. However, in the present study there were only two cases (4.17%) of traumatic injury requiring enucleation. Recent advances in the management of ocular injuries have resulted in increased salvage rate of severely injured eyes, and thus decreased rate of enucleation and evisceration. Intraocular ossification in cases of phthisis bulbi is as a result of osseous metaplasia in subretinal region accompanied by fibrovascular proliferation and inflammation (9).

Unilateral retinal dysplasia is a rare entity resulting from disturbed retinal growth and differentiation. The cause of such unilateral eye abnormality is unknown (10). Clinically this lesion mimics retinoblastoma and diagnosis rests on microscopic examination. In the present study one case was clinically diagnosed as retinoblastoma but the pathologic examination changed the postoperative management.

To conclude, in our setting, non-neoplastic lesions are the main cause of eyeball surgery as compared to the West, where trauma followed by neoplasms constitute important causes. Pathologic examination of eyeballs is a must for proper postoperative management of the patient.

Proprietary interest: None.

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