## Original article

# Clinical characteristics of orbital tumours as seen in a tertiary eye center

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**Abstract**: Clinical files of 93 patients with orbital tumours were examined. Primary orbital tumours accounted for 55% of cases and secondary tumours for 45%. The most frequently diagnosed primary orbital tumours were mucoceles (15%), lacrimal gland tumours (7.5%), and rhabdomyosarcoma (6.5%). The most common secondary orbital tumours were retinoblastoma (23.6%), and squamous cell carcinoma (10%). Commonly found physical signs, radiographic and ultrasonographic findings are reported. Most frequently employed surgical techniques for removal of the tumours was exenteration (64%). Orbitotomy through various approaches was employed in 36% of the patients. Agreement was obtained between the clinical and histopathologic diagnoses for the commonly encountered orbital tumours. Poor outcome is expected, because of late presentation in most patients. Discussion and recommendations on further management of cases is given. [*Ethiop. J. Health Dev.* 1997;11(3):201-206]

**Introduction**

The orbit can harbour primary tumours arising from orbital structures, it can be secondarily involved from eyelid and ocular tumours, or it can also be a seat for distant metastasis. Generally cystic, vasculognic and inflammatory tumours predominate the picture of primary orbital tumours in all age groups (1, 2, 7, 11). Retinoblastoma in children and eyelid tumours and uveal melanomas in adults are the most common causes of secondary orbital tumours elsewhere. In African studies the picture is similar with the exception of rarity of uveal melanoma as a cause of secondary orbital tumours.

The source of pathology specimens for orbital tumours is generally from exenteration of the orbit or by total or subtotal excision of the tumour mass through various surgical approaches (1, 2, 7).

Various orbital tumours the patterns of which tend to vary with the age of the patient and geographical location of the study area have been described by numerous investigators (1, 10, 12, 13).

The sources of materials of most of the reports on incidence of orbital tumours are from pathology units and few studies integrated the clinical characteristics with the histo-pathologically verified diagnosis (2, 3, 4, 5).

Although the pathologic patten of orbital tumours has been described in this tertiary eye center 14 years ago, the clinical characteristics of the different orbital pathologies were not studied (13). The purposes of this retrospective study are to:

1. describe the pattern of orbital tumours by age, sex, clinical impression and histopathologic diagnosis,
2. study the clinical characteristics of the different orbital tumours and compare with histopathologic diagnosis,
3. study the malignancy rate of orbital tumours.

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**Methods**

The study was conducted at Menilik II Hospital, Department of Ophthalmology which is a teaching and national referral center under the Medical Faculty of Addis Ababa University, Addis

Ababa, Ethiopia.

We looked for the clinical records of 143 patients with orbital tumours on whom surgery was performed and histopathologic verification of their tumours was made. Pathology files of the study population found at Tikur Anbessa Hospital, Addis Ababa University, Medical Faculty, were identified and studied in detail.

Patients who underwent orbital surgery from 1st July 1990 up to 30 th June 1994, the surgical specimens of whom were examined histo-logically and patients with both primary orbital tumours and lesions originating from eyelids and eyeball with secondary orbital involvement were included in the study. The study was limited to analyze four years records as data were incomplete prior to this period.

Patients with lesions confined to eyeball and eye lids without orbital involvement, dermoids and orbital inflammatory conditions not subjected to surgery were excluded from the study.

All available clinical information in each patient is recorded. The variables that were looked for are: age and sex of the patients, duration of illness, mode of proptosis (position of globe), visual acuity, general physical examination, ophthalmoscopic and extraocular motility findings, amount of proptosis (Hertel exophthalmometry), x-ray characteristics, ultrasonographic picture, type of surgery and histopathologic diagnosis. The classification system of orbital tumours given by Jerry A. Shields is used in the present study to fill and analyze the data available (2, 3).

**Results**

During the period 1st July 1990 to 30 th June 1994 a total of 143 patients were operated for orbital tumours with their specimens subjected to histological examination. Clinical data and histopathologic reports of 93 patients were available for study.

The frequency distribution and demographic data of patients with orbital tumours are shown in Table 1. Primary orbital tumours accounted for 55% of cases and secondary orbital tumours for 45% of all cases. The most frequently diagnosed primary orbital tumours were mucoceles 14 (15%), lacrimal gland tumours 7 (7.5%) and rhabdomyosarcomas 6(6.5%). The most common secondary orbital tumours were retinoblastoma 22(23.6%) and squamous cell carcinoma 15(16.1%). Forty eight percent of cases occurred in the right orbit and 52% in the left. The male to female ratio was 1:2.

Table 1: **Frequency distribution, age range, means of age and duration of illness of patients with orbital tumour**

| Pathologic diagnosis | Cases | | Age | Mean Age | mean duration |
|---|---|---|---|---|---|
| | No | % | Range (yrs) | (Yrs ±SD) | (Month) |
| Retinoblastoma | 22 | 23.6 | 0.7-7 | 3.3±1.3 | 10 |
| Squamous cell ca. | 15 | 16.1 | 22-75 | 47±15.9 | 18 |
| Mucoceles | 14 | 15.0 | 17-70 | 40±19.1 | 36 |
| Rhabdomyosarcoma | 6 | 6.5 | 4-20 | 9± 6.1 | 2 |
| Lacrimal gland tumours | | | | | |
| Pleomorphic adenoma | 3 | 3.2 | 16-50 | 32±17.0 | 76 |
| Carinoma | 4 | 4.3 | 22-60 | 45±16.3 | 11 |
| Malignant melanoma | 5 | 5.4 | 28-70 | 54±16.2 | 9 |
| Neurofibroma | 4 | 4.3 | 4-63 | 32±24.1 | 16 |
| Hemangioma | 4 | 4.3 | 18-50 | 35±13.4 | 96 |

| | | | | | |
|---|---|---|---|---|---|
| Meningioma | 4 | 4.3 | 11-45 | 26±15.9 | 55 |
| Lymphoma | 2 | 2.2 | 35-50 | 42±10.6 | 21 |
| Adipose tumours | 2 | 2.2 | 50-60 | 55± 7.0 | 106 |
| Others* | 8 | 8.6 | | | |
| Total | 93 | 100.0 | | | |

* One each case of: Pseudotumour, Fibrohistocytosis, Varix, Angiofibroma, Basal cell ca., Granuloma, Neuroblastoma, Hydatid cyst.

The age range and mean ages at presentation for orbital tumours are also shown in Table 1. The mean age at presentation for retinoblastoma was 3.3±1.3 SD yrs, for mucoceles 40±19.1 SD yrs and squamous cell carcinomas 47±15 SD yrs. Patients with pleomorphic adenoma of lacrimal gland had lower mean ages (32±17 SD yrs) as compared to those of lacrimal gland carcinoma (45.5±16.3 SD yrs). The mean duration of clinical history was least in patients with rhabdomysacrcoma (2 months) and retino-blastoma (10 months).

The clinical data on patients with orbital tumours is shown in Table 2. Eighty nine percent of patients presented with proptosis. Positive family history was found only in one patient who had squamous cell carcinoma. None of the patients with retinoblastoma had positive family history of the disease.

Table 2: **Clinical data on patients with orbital tumours**

| Signs | Cases | |
|---|---|---|
| | No | % |
| **Proptosis** | | |
| Present | 83 | 89 |
| Absent | 10 | 11 |
| Total | 93 | 100 |
| **Globe status*** | | |
| Intact | 49 | 53 |
| Destroyed | 44 | 47 |
| Total | 93 | 100 |
| **Eyelid involvement** | | |
| Present | 43 | 46 |
| Absent | 50 | 54 |
| Total | 93 | 100 |
| **Ocular motility** | | |
| Restricted | 33 | 79 |
| Normal | 9 | 21 |
| Total | 42 | 100 |

* Patients with intact globes only.

The globe was destroyed at presentation in 47% of patients. All of the 22 patients with retinoblastoma and 13 of the 15 patients with squamous cell carcinoma (86.7%) had ruptured globes at the time of presentation.

Eyelids were infiltrated by tumours in 46% of patients. Thirteen out of 22 patients with retinoblastoma (59%) and 13 of the 15 patients with squamous cell carcinoma (86.7%) had lid involvement at presentation. Out of 42 patients who had records of ocular motility examination 33(79%) had restricted ocular motility in one or more directions.

Details on position of globe at presentation and visual acuity levels are as shown in Table 3. Out of the 49 patients with intact globes, record on globe position was obtained in 46 patients. Four (9%) patients had normal globe position and the remaining 42 patients (91%) had globe displacement in one or more directions. Together with the proptosis, abnormal globe displacement observed was, down and out (28%), vertically down (15%), horizontally out (13%) and axial displacement (11%). Seventy four percent of patients were blind in the involved eye.

Table 3: **Details on position of globe and visual acuity levels**

| Globe Displacement | No. | % |
|---|---|---|
| Down and Out | 13 | 28 |
| Down | 7 | 15 |
| Out-horizontally | 6 | 13 |
| Axial | 5 | 11 |
| Normal | 4 | 9 |
| Others | 11 | 24 |
| Total | 46 | 00 |
| **Visual Acuity (n=93)** | | |
| NLP* - ≤3/60 | 70 | 74 |
| >6/36 = ≤6/24 | 12 | 13 |
| ≥6/18 | 12 | 13 |
| Total | 93 | 100 |

The amount of proptosis was quantified by means of Hertel's exophthalmometry in 29 of the 49 patients with intact globes. The mean difference in exophthalmometric readings between the involved eye and fellow eye of the entire group was 6.3 mm, being 7.5 mm for mucoceles and 7.6 mm in meningiomas.

Table 4: **Mode of surgery in 93 patients with orbital tumours**

| Type of Surgery | No. | % |
|---|---|---|
| Exenteration | | |
| Total Exenteration | 37 | 40 |
| Lid Saving | 22 | 24 |
| Orbitotomy | | |
| Brow Incision 24 | 26 | |
| Trans Conjunctival | 4 | 4 |
| Coronal Flap | 3 | 3 |

| | | |
|---|---|---|
| Kronlein | 3 | 3 |
| Total | 93 | 100 |

Ophthalmoscopic examination was possible in 32 patients. In the rest of the patients fundus examination was not possible either because of destroyed globes (44 patients), opaque media (10 cases) or unknown reasons (7 cases). The fundus appeared normal in 20 patients (63%). The most common abnormal fundus finding was choroidal striae (16%). Other abnormal fundus findings include optic nerve pallor (3 cases or 9%), choroidal striae and optic nerve pallor (2 cases or 6%) and papilloedema (1 cases 3%). Orbital x-ray was obtained in 58 (62%) of the patients. The commonest x-ray abnormality found was increased soft tissue density which, in isolation or in combination with other abnormalities, was seen in 45% of the patients. Orbital widening was seen in 36% of cases either as an isolated finding (12%) or in combination with other x-ray findings (24%). Bone destruction with sinus involvement was seen in 16% of patients. Two patients had enlarged optic canals.

Table 5: **Mode of surgery for common orbital tumours**

| Diagnosis | | |
|---|---|---|
| Retinoblastoma | Exenteration | Orbitotomy |
| Squanous Cell Ca | 22 | |
| Mucocele | 15 | |
| Lacrimal Gland Tumours | - | 14 |
| Rhabdomyosarcoma | 3 | 4 |
| Malignant Melanoma | 6 | 4 |
| Neurofibroma | 5 | |
| Hemangioma | 2 | |
| Meningioma | - | 4 |
| Total | 2 | 2 |

Solid tissue pattern was the most frequently noticed ultrasonographic finding (63%). Cystic picture was noticed in 11 (34%) patients. Infiltrative pattern in one patient (3%). The surgical procedures performed on patients with orbital tumours are given in Tables 4 and 5. The most common surgical procedure performed was exenteration (37 or 40%) or partial, lid saving exenteration (22 or 24%). Orbitotomy through different approach accounted for 34 (36%) of the operations. The specific type of surgery performed for common orbital tumours is shown in Table 5. Agreement between clinical and histopathological diagnosis of the commonly diagnosed orbital tumours is depicted in Table 6. The agreement is higher for retinoblastoma, squamous cell carcinoma, mucocele and rhabdomyosarcoma.

Table 6: **Agreement between clinical and histopathological diagnosis of the common orbital tumours**

| Disease Condition | Clinical Diagnosis | Histologically Confirmed | Missed No. |
|---|---|---|---|
| Retinoblastoma | 22 | 21 | 1 |
| Squamous cell Ca | 17 | 15 | 2 |
| Mucocele | 18 | 14 | 4 |
| Rhabdomyosarcoma | 8 | 6 | 2 |
| Lacrimal Gland Tumours | 6 | 7 | 4 |

| | | | |
|---|---|---|---|
| Malignant Melanoma | 4 | 5 | 2 |
| Hemangioma | 8 | 4 | 1 |
| Retinoblastoma | 22 | 21 | 1 |

**Discussion**

The comprehensive classification system of orbital tumours and pseudotumours was given by Shields et al (2). This classification system was utilized in the present series. In the review of orbital tumour by Shield's et al, cystic, inflammatory, lacrimal and secondary orbital lesions (in that order) dominated the picture.

The single most common cystic lesion in the series conducted by Shields was dermoid cyst accounting for 80% of the cystic lesions. Because of lack of definition of exact tumour site of dermoid tumours, this tumour was excluded from our study. Excluding dermoid cyst the second most common cystic tumour was mucocele. This condition was the most common benign lesion observed in the present series.

The relative frequency of orbital tumours obtained in the present study agrees with other studies conducted on the subject in developing countries (4, 6, 13). Exception to this is the fact that significant proportions of the patients with orbital tumours observed in Nigeria had extension of Burkitt's lymphoma from maxilla. Rapid regression of Burkitt's lymphoma with cytostatic drugs can be achieved and, as treatment of this condition is in pediatric hospitals, the patients could have escaped histopathological examination of their orbital tumours. In our series retinoblastoma and squamous cell carcinoma were observed to be the most common secondary orbital tumours. This is in agreement with other studies in developing countries (1, 4, 6). In contrast to this in a series conducted by Shield et al, basal cell carcinoma and Uveal melanomas were most common origins for secondary orbital tumours. Both of these conditions are said to be rare in non-white populations.

A recent study on the pattern of orbital tumours in children over the past 60 years has shown that the number of secondary tumours has decreased significantly. This reflects the improved early diagnosis of diseases with potential secondary orbital involvement. This study also signifies the necessity for periodic evaluation of orbital tumours for changing pattern of diseases in response to improved early detection and management of the tumours (5).

Previous studies conducted in developing countries indicated that patients with retinoblastoma usually present in advanced stages. The mortality rate of patients with orbital involvement has been shown to be 100% (6). The present study reconfirmed this fact. Early detection and management of the disease is of paramount importance in the effort to improve vision and survival of the patient (8, 9, 11). A report by Abramson et al indicated that early detection of retinoblastoma with implementation of less destructive modes of treatment such as cryo and laser photocoagulation has led to a decrease in the frequency of enucleation and prolonged the survival of patients with the disease (8).

In the present study, none of the patients with retinoblastoma had a positive family history. These patients probably had the non- hereditary form of the tumour. This form of the tumour is further evidenced by the unilateral nature of the tumours and presentation at a higher mean age than the usual presentation for the hereditary forms (9, 14).

All of the patients with secondary orbital tumours and most of those with primary tumours that were reported in this series came to the hospital with an advanced stage of the disease. Fifty percent of the patients had destroyed globes with eyelid involvement and three quarters with visual acuity of counting fingers at three meters or less.

In the analysis of 20 patients with exophthalmos Frieberg and Associates have shown that the choroidal folds they observed occurred with anteriorly located orbital tumours and in patients with greater amount of exophthalmos (14). In the present study, although only a small number of patients were noticed to have choroidal folds, it was the most common Ophthalmoscopic abnormality detected and the patients had either anterior location or large proptosis.

In this study plain x-ray films and B-scan Ultrasonography have been observed to be important ancillary tools in the evaluation of orbital tumours. In areas where computerized tomography or MRI is not available these diagnostic tools are important in the management and follow-up of patients with orbital tumours.

Unlike the studies from areas where early detection and less drastic modes of treatment are employed, our study shows that the great majority of the surgeries were done for palliation of advanced orbital tumours. Orbitotomy with resection of the tumour or drainage of cyst was performed in only 1/3 of the patients. Late presentation of the tumours in far advanced disease state made them less amenable for non-destructive forms of treatment.

This study showed an agreement between the clinical and histopathological diagnosis for the commonly seen orbital tumours. A proportionally small number of cases were missed clinically. A higher index of awareness saves time and unnecessary cost spent during the preoperative evaluation of the patients and narrows the areas where the pathologist should be looking for.

**Conclusion**

Delay in presentation of patients with malignant orbital tumours is observed in this series. These conditions should be diagnosed early and prompt referral and management of cases should be made. Complete evaluation of proptosis should be performed, including clinical history, physical and radiological examination to facilitate early diagnosis and management.

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