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**ORIGINAL ARTICLE**

# PATTERNS OF TUMOURS INVOLVING THE ORBIT IN PATIENTS SEEN AT MENELLIK II REFERRAL HOSPITAL, ADDIS ABABA, ETHIOPIA.

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## ABSTRACT

***Background:*** *The incidence and relative frequency of orbital tumours varies widely and has been reported to be dependent on differences in geographical areas, race and age of the patient. Geographical differences may suggest environmental factors involving particular disease development and socioeconomic status.*
***Purpose:*** *To determine the pattern of tumors involving the orbit among patients seen at Menellik II referral hospital.*
***Methods:*** *Hospital based, prospective cross sectional study was conducted at Menellik II referral hospital, Addis Ababa, Ethiopia from April 2015 to July 2016 on 144 patients with orbital tumors who underwent surgical intervention and biopsy. Parameters included age, sex, patient complaint, duration and location of the lesion, description of the lesion, preoperative clinical diagnosis, and histopathological diagnosis.*
***Results:*** *From a total of 144 patients, 48.7% were male and 51.3% were female with mean age of 23.8 ± 21.2 years. The most prevalent histopathological diagnoses for patients under 15 years of age were retinoblastoma (31.1%) followed by dermoid cyst (28.4%), and rhabdomyosarcoma (12.2%). For patients above 15 years of age, the most frequently seen tumors were squamous cell carcinoma (23%), dermoid cyst (13.1%) and malignant lacrimal gland tumor (13.1%).*
***Conclusion:*** *Orbital tumors demonstrate a broad spectrum of biological behavior. The most common malignant tumors involving the orbit were retinoblastoma and squamous cell carcinoma for patients under and above 15 years of age respectively and the most common benign tumor was dermoid cyst in both age categories. The majority of the tumors in this study were secondary orbital tumors which implies late presentation of patients.*
***Key Words:*** *Pattern, Tumors, Orbit, Menellik II Hospital, Addis Ababa, Ethiopia.*

## INTRODUCTION

The orbit is composed of many different structures such as bone, extra-ocular muscle, and peripheral nervous system, optic nerve of the central nervous system, vessels, adipose tissue, exocrine gland and lymphoid tissue. Because of the presence of these various tissues, there are multiple types of tumors that originate from this region (1-4).

The incidence and relative frequency of orbital tumours varies widely and has been reported to be dependent on differences in geographical areas, race and age of the patient (2,5,6). Geographical differences may suggest environmental factors involving particular disease development and socioeconomic status (7). For instance, orbital lymphoma is more prevalent in Asia and Europe than in US (7,8) whereas retinoblastoma is more common in Africa with orbital extension (9-11).

Most of the tumours that occur in the orbit are benign, predominantly vasculogenic capillary hemangioma (12-14) in children and cavernous hemangioma in adults (5,10). Malignant tumours account for 20-25% and most malignant tumours occur at 7$^{th}$ decade of life and afterwards (12). The common malignant lesion in adults is lymphoma (10,11,15), whereas in children retinoblastoma with orbital extension is the most common (13,14). But in studies done in developed nations like the USA, rhabdomyosarcoma is the most common, followed by secondary malignant tumours (15,16).
Orbital tumours can be divided into three major subgroups: primary, secondary and metastatic. Further, the primary neoplasms are categorized based on the cell of origin (vascular, neural, etc.), and secondary and metastatic neoplasms are grouped based on the organ of origin (e.g., skin melanoma, breast carcinoma, etc.) (17,18). Another classification is based on the biologic behaviour of neoplasms: benign versus malignant (1,17,19).

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Although thought to be rare, orbital tumours contribute to significant morbidity and mortality (1,20) and may result in several complications such as cosmetic problems, severe ophthalmological deficit with loss of vision or eye movement deficit, and even loss of life. It is important to obtain maximum information about an orbital tumour prior to determining the appropriate treatment strategy in order to prevent postoperative visual impairment.

Therefore, this study was conducted to determine the pattern of tumours involving the orbits among patients attending Menellik II referral hospital based on common clinical presentation and histopathologic diagnosis. The result of the study will serve as base line reference as there is no published data from our setting.

## SUBJECTS AND METHODS

This is a hospital based prospective descriptive cross sectional study conducted between April 2015 and July 2016 at Menellik II referral hospital, Addis Ababa, Ethiopia.

From all patients visiting Menellik II referral hospital during the study period, patients who were suspected to have orbital tumor clinically and who underwent excisional biopsy as part of surgical management were included in the analysis. No selective sampling was applied as all consecutive patients who fulfill the inclusion criteria were included into the study during the study period.

For the purpose of this study, tumor involving the orbit is defined as any tumor that arises primarily within the orbit and from adjacent structures such as the eyelids, intraocular structures, or paranasal sinuses or from distant areas that invade the orbit posterior to the orbital septum. Patients with presumed diagnosis of tumor involving the orbit were included in the study. Those patients for whom there was no biopsy from surgical intervention, those with tumours directly arising from within the globe, the eyelid skin, the conjunctiva, and the accessory lacrimal glands or the paranasal sinuses were excluded unless they had orbital extension.

Patients were evaluated based on the questionnaire developed by the principal investigator and all the necessary information was gathered based on the checklist and completed by a resident assigned to the specific clinic during the study period. The principal

The questionnaire included information on card number of the patient, age, sex, patient complaint, systemic illness, and duration and location of the lesion, and description of the lesion, clinical diagnosis and histopathologic diagnosis. The name of the patient was not recorded, and identity was kept confidential. All information was then coded, entered, and analyzed using SPSS version 20.

### *ETHICAL CONSIDERATIONS*

Ethical clearance was obtained from the Research and Publication committee of the Department of Ophthalmology, School of Medicine, College of Health Science, Addis Ababa University. Informed written consent was obtained from each adult study participant and from parents or guardians in the case of children.

## RESULTS

A total of 144 patients were included in the study. Among them, 68(47.2%) were male and 76(52.8%) were female. Mean age of patients included in this study was 23.3±21.3 years (range, 4 months to 80 years) and the median age was 16 years (95% CI, 20.43-27.17) **Figure 1**.

Common symptoms and signs encountered were swelling (63.9%), proptosis (43.1%), visual impairment (54.1%), ocular pain (13.2%), strabismus (7 %) and diplopia (2.8%). The most common clinical diagnosis was dermoid cyst 26(17.1%), followed by retinoblastoma (RB) 24(15.8%), and lacrimal gland tumor 15(9.9%) regardless of age and sex **Table 1**. Of 144 biopsy results, the most frequent histological types encountered were dermoid cyst, 29(21.5%), retinoblastoma 23(17%), and lacrimal gland tumor 16(11.9%) with a mean age of 12.28(8.76), 3.91 (1.80), and 24.19(14.59) years respectively **Table 2**. Histologic pattern was assessed by age category. From a total of 144 specimens, 74 belonged to patients of under 15 years of age and 70 to those above 15 years of age. Retinoblastoma (31.1%) was the most common histological diagnosis for age category below 15 years of age followed by dermoid cyst (28.4%) (**Figure 2)**. Squamous cell carcinoma (SCC) (23%) was the commonest for age group above 15 years of age followed by lacrimal gland tumor (LGT), 18% (**Figure 3)**. The number of benign tumors was slightly higher (52.1%) than malignant and no metastatic lesions were identified in both age categories. For patients aged 15 years or younger, 24.2% were primary and 75.8% were secondary tumours whereas for patients above 15 years of age, 22.2% were primary and 77.8% were secon-

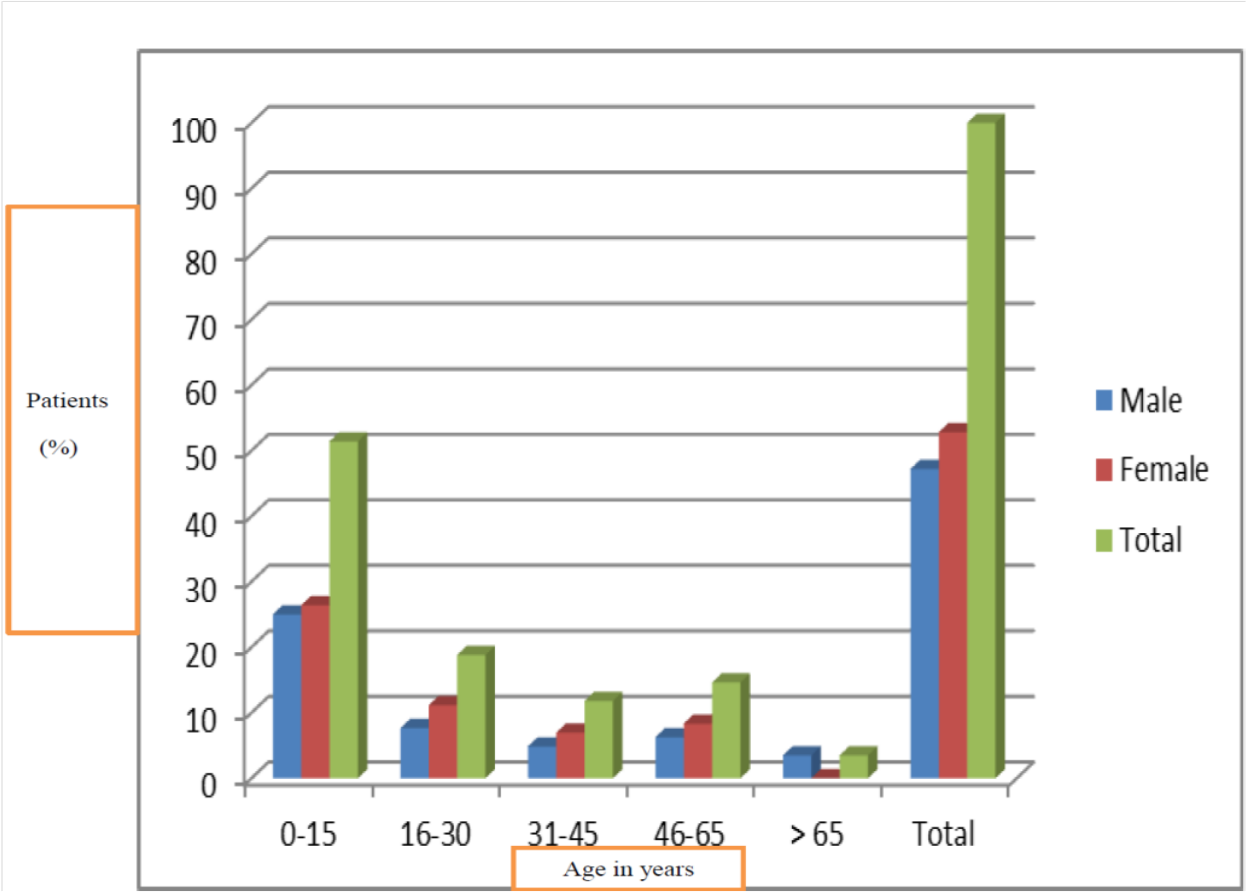


**Figure 1.** Age and sex distribution of study participants, Menellik II hospital, April 2015 to July 2016.

**Table 1**. Summary of Clinical Diagnosis of Orbital tumouurs in Menellik II Referral Hospital, April 2015 to July 2016.

| Clinical diagnosis | Frequency (N) | Percent (%) |
|---|---|---|
| Dermoid cyst | 28 | 19.4 |
| Retinoblastoma | 24 | 16.7 |
| Lacrimal gland tumor | 15 | 10.4 |
| Optic nerve glioma | 15 | 10.4 |
| SCC | 13 | 9.0 |
| BCC | 6 | 4.2 |
| Ethmoidal mucocele | 5 | 3.5 |
| Rhabdomyosarcoma | 5 | 3.5 |
| Osteoma | 4 | 2.8 |
| Lymphoma | 4 | 2.8 |
| Orbital mass | 4 | 2.8 |
| Capillary hemangioma | 4 | 2.8 |
| Cavernous hemangioma | 4 | 2.8 |
| Neurofibromatosis | 4 | 2.8 |
| Frontal mucocele | 4 | 2.8 |
| Melanoma | 3 | 2.1 |
| Orbital cellulitis | 1 | 0.7 |
| Optic nerve sheath meningioma | 1 | 0.7 |
| Total | 144 | 100 |

**Table 2.** Distribution of tumours of the orbit by histological diagnosis, frequency, age, sex, and duration of symptoms, Menellik II Referral Hospital, April 2015 to July 2016.

| Histological diagnosis | Number (%) N=144 | Mean age in years | Sex ratio (M:F) | Mean duration of symptoms in months (SD) |
|---|---|---|---|---|
| Dermoid cyst | 29(20.1) | 12.3 | 1:1.9 | 57.59(46.1) |
| Retinoblastoma | 23(16.0) | 3.9 | 1:2.3 | 6.24(6.3) |
| Lacrimal gland tumor | 16(11.1) | 24.2 | 1.25:1 | 13.59(11.3) |
| Squamous cell carcinoma | 16(11.1) | 50.9 | 1.5:1 | 46.03(39.9) |
| Rhabdomyosarcoma mucocele | 9(6.3) | 6.3 | 8:1 | 11.28(22.9) |
| Optic nerve glioma | 7(4.9) | 11.8 | 4:3 | 9.71(7.8) |
| Cavernous hemangioma | 5(3.5) | 33.2 | 3:2 | 63.60(72.2) |
| Melanoma | 4(2.8) | 64.0 | 3:1 | 18.50(14.7) |
| Lymphoma | 4(2.8) | 29.0 | 4:0 | 11.75(8.7) |
| Basal cell carcinoma | 3(2.1) | 58.3 | 1:2 | 52(36.7) |
| Optic nerve meningeothelial meningioma | 3(2.1) | 30.0 | 2:1 | 24.50(41.1) |

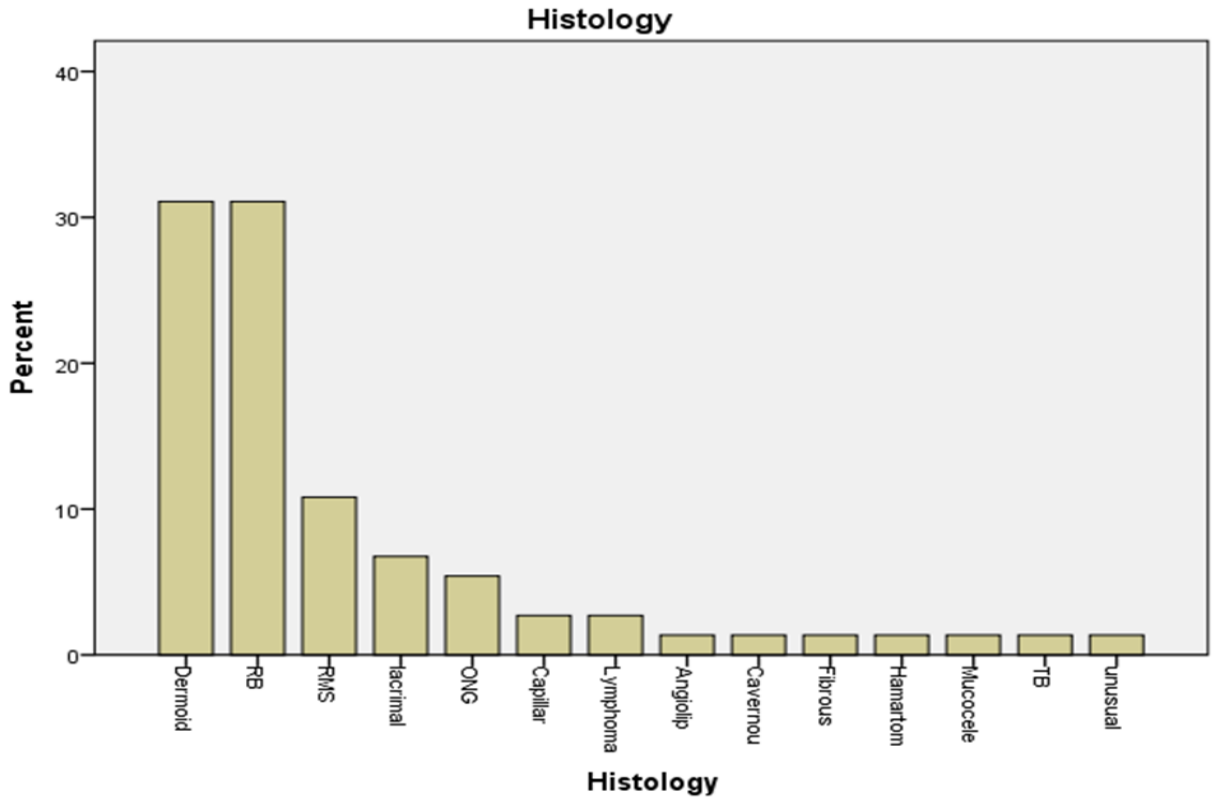


**Figure 2.** Histological diagnosis in tissue biopsies of study participants 15 years or younger, Menelik II Referral Hospital, April 2015 to July 2016.

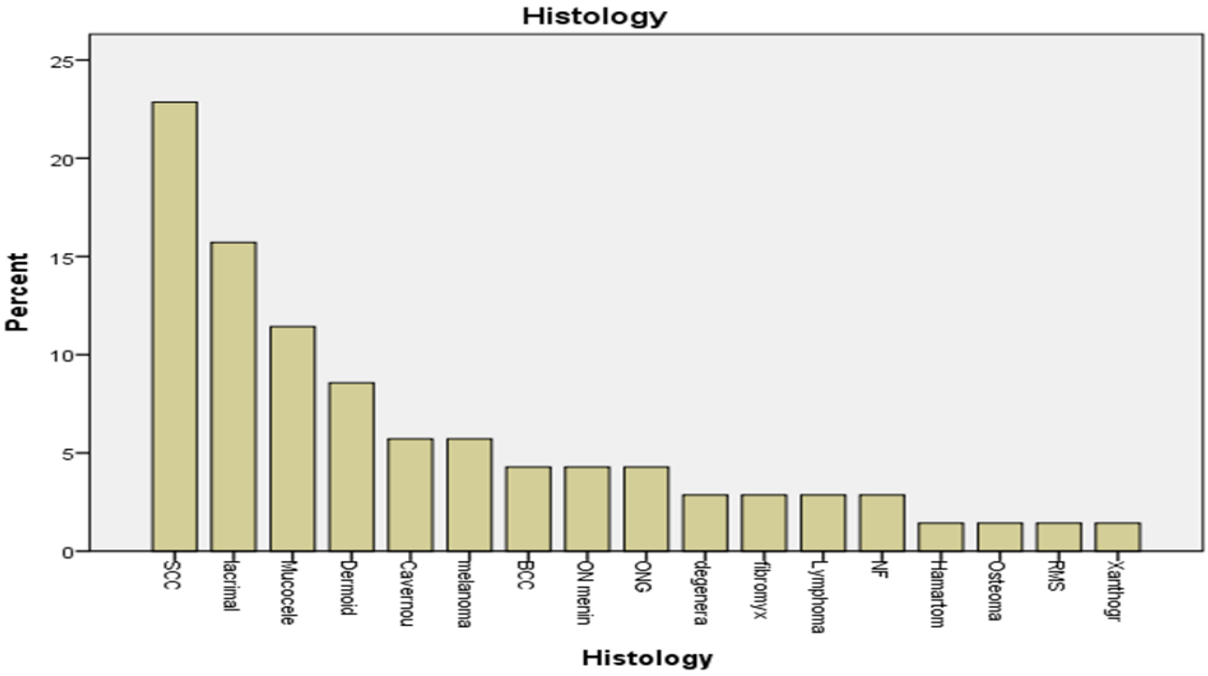


**Figure 3.** Histological diagnosis in tissue biopsies of study participants older than 15 years of age, Menellik II hospital, April 2015-July 2016.

Capillary hemangioma, fibromixoid sarcoma, hamartoma, degenerated vascular lesion and neurofibromatosis each accounted for two cases. Angiolipoma, Fibrous dysplasia, Osteoma, TB, Unusual undifferentiated congenital tumor and Xanthogranulomatous inflammation each accounted for one case.

## DISCUSSION

One hundred forty four patents with orbit involving tumor were assessed based on clinical and histopathological examination of specimens. The mean age was 23.3 ± 21.3 years (range, 4 months to 80 years) and the majority were female (52.8%).

In this study, there is a long list of tumours involving the orbit due to the presence of a vast number of various tissues in the area. The proportion of benign tumors was slightly higher (52.1%) than those of malignant. In reports from the USA, Japan and the Philippines (5,21,22), most orbital lesions were reported as benign whereas in Nigeria (23) most lesions were reported as malignant (75.5%). This variation may be due to differences in genetics and geography, implying that genetic and environmental factors are involved in disease development.

Among malignant lesions, 24.2% were primary and 75.8% were secondary tumours in patients 15 years or younger whereas in patents above 15 years, 22.2% were primary and 77.8% were secondary. No metastatic lesions were identified in both age categories. A study done by Johansen S *et al* in Denmark (6) showed primary (65%), secondary (29%), metastatic (6%) tumours in children and primary (43%), secondary (48%), and metastatic (9%) tumours in adults. This could be due to delay in diagnosis and management of secondary tumours at their primary site in our series. The most malignant cases were retinoblastoma in the under 15 years of age in our study. This agrees with studies done in Nigeria, Uganda, India and Nepal (9,13,15) but differs from studies conducted in the USA, Japan and the Philippines where rhabdomyosarcoma was most common (15,16).

The average age of retinoblastoma was 3.25 years in our study. Studies done in both Pakistan and India reported a mean age of 3.8 years (11,12) whereas studies done in western countries showed earlier ages of presentation. This earlier age of presentation and primary tumour predominance in developed countries is probably due to better diagnostic facilities and increased awareness among the public, which is lacking in our country.

In this study, squamous cell carcinoma (25.4%) was the commonest malignant tumour in patients above 15 years of age, which is different from other studies where the most common malignant tumour in adults was lymphoma (2,5,11). Our finding is however similar to a study done in Cameroon (14). In children, different frequencies were reported regarding benign orbital tumors in the literature. Kodsi SR *et al* and Stefanyszyn MA *et al* (21,24) reported dermoid cyst (12.2%) as the commonest. Nath, *et al* reported pseudotumours (23.33%) as the most frequent (25). Hemangioma was top on the list in the study of Shield JA(26). Pleomorphic adenoma of lacrimal gland was commonest in the study by Mohan *et.al* (27). In our study, dermiod cyst was the most common benign tumour for all patients regardless of age.

This study also showed that the most common clinical presentation was that of a mainly fungating mass, which is similar to a study in Nigeria (11). This differs from a study done in Denmark (12) where proptosis was the most common (13,24). This could be due to the fact that the study was done at a tertiary hospital and most of our patients are referred at advanced stage and several benign entities were underrepresented.

In this study, the prevalence of seropositivity for HIV in patients with SCC was 60 % which is in line with studies made on conjunctival SCC in Nigeria (75%), Uganda (71%) and Malawi (86%) (20,28).

Clinically, a significantly large number of tumours invades the orbit secondarily from adjacent structures such as the globe and eyelids. This made up a large proportion of all orbital malignancies in this study. This proves the fact that in developing countries like Ethiopia, late diagnosis and treatment of malignancies is still prevalent and many patients seek medical attention at late advanced stages of the disease.

## CONCLUSION

In conclusion, a total 144 biopsy proven cases of orbit involving tumors were diagnosed at Menellik II referral hospital with 52.1% of them benign. Dermoid cysts were the most frequent benign tumors. The most common orbital malignancies were RB and SCC for age categories of 15 or under and for those

The majority of patients seek medical attention late. Designing ways to create awareness to the public and health professionals about noninfectious ocular conditions like tumours, especially secondary tumours, is important for early detection and referral. Most patients with SCC were seropositive for HIV. It is thus recommended that all patients presenting with squamous cell carcinoma should be screened for HIV.

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