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Pathology

Orbital Tumors and Pseudotumors – A Clinicopathological Study

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Abstract: Tumors of the orbit require a multidisciplinary approach and closer attention due to the specific anatomic structure of orbit. Orbital pseudotumors include a broad category of orbital inflammatory diseases and pose a special diagnostic problem. They mimic true orbital neoplasms. This study aims to analyze the clinical profile and histopathology of orbital tumors and pseudotumors in patients presenting at a tertiary eye care centre A retrospective clinicopathological analysis of 54 orbital tumors diagnosed at the Department of Pathology, Sarojini Devi Eye Hospital, Hyderabad was done over a period of three years. The clinical data was retrieved from the case sheets. Hematoxylin and eosin stained slides of the cases were reviewed. The age of the patients ranged from 5years to 62 years. Maximum number of cases occurred below 15 years. Most of the malignant tumors occurred in older age group above 60 years. Twenty two cases (40.74%) of orbital tumors occurred in children. Males constituted 33 cases (61%) and females constituted 21 cases (39%). (Table. 2) Histopathological spectrum showed 40 cases of benign tumors constituting 74% of the total and 14 cases of malignant tumors constituting 26% of the total orbital tumors. Our study is compared with other studies in literature. Majority of childhood orbital tumors are benign. Rhabdomyosacoma is the commonest malignancy in childhood. Malignancy should be considered in elderly patients presenting with proptosis.
Keywords: Orbital tumors, Pseudotumors, histopathology,malignant tumors.

INTRODUCTION

Orbit is an anatomically complex structure containing globe, extra ocular muscles, fat, vascular, nervous and glandular connective tissues. All these anatomical structures can give rise to neoplasms[1]. Orbital tumors constitute a heterogeneous array of lesions and as such, pose numerous challenges in terms of diagnosis, imaging and management[2]. Given the variety of structures within the relatively confined orbit, a systematic approach is necessary to understand the classification and clinical features of orbital tumors[3]. The majority of orbital tumors originate between the bony orbital wall and the extraocular muscle cone and 90% of the orbital tumors present with proptosis[4]. Close co-operation between an oculo-plastic surgeon, otorhinolaryngologist, neurosurgeon, radiologist, oncologist and histopathologist is required for the management of orbital tumors[5]. The World Health Organization classification of the tumors of the orbit provides a framework dividing them into groups according to origin as follows: benign soft tissue tumors, malignant soft tissue tumors, lymphoid and hematologic tumors, pseudotumors and other primary tumors of the orbit. Secondary and metastatic tumors of the orbit, are a separate group of orbital tumors[6]. Orbital pseudotumors are a nonspecific idiopathic inflammatory process that is histopathologically classified into three main types: granulomatous, lymphoid and sclerosing[7]. This condition was first described in 1905 by Birch-Hirschfeld[8].The present study aims to analyze the clinical profile and histopathology of orbital tumors and pseudotumors presenting at a tertiary eye care centre.

MATERIALS AND METHODS

A retrospective analysis of fifty four cases of orbital tumors was conducted over a period of three years from July 2013 to June 2016 at the Department of Pathology, Sarojini Devi Eye Hospital, Hyderabad. Intraocular tumors were excluded from the study. The clinical data such as age, sex, ophthalmic examination findings, X-Ray, CT and MRI findings etc were noted by reviewing the case records. The H & E stained histopathology slides of all the cases were reviewed. Special stains and immunohistochemistry done in relevant cases were reviewed. The data was analyzed

to know the clinical &histopathological characteristics of orbital tumors and pseudotumors.

OBSERVATIONS AND RESULTS

During the study period, a total number of 54 cases of orbital tumors and pseudorumors were diagnosed. The age of the patients ranged from 5 years to 62 years. Maximum number of cases occurred below 15 years. Most of the malignant tumors occurred in older age group above 60 years. (Figure 1).Twenty two cases (40.74%) of orbital tumors occurred in children and 32 cases (59.25%) occured in adults. (Table.1) Males constituted 33 cases (61%) and females constituted 21 cases (39%). (Table. 2) Histopathological spectrum showed 40 cases of benign tumors constituting 74% of the total and 14 cases of malignant tumors constituting 26% of the total orbital tumors.(Table 3).Dermoid cyst (Fig.1) was the most common diagnosis with 16 cases (29.6%) followed by seven cases (12.96%) of vascular lesions (Fig.2) and six cases (11.11%) of pleomorphic adenoma of lacrimal gland (Fig.3). Meningioma (Fig.4) constituted four cases (7.4%) and lacrimal gland cyst (Dacryops) (Fig.5) constituted three cases (5.55%). Schwannoma constituted two cases (3.70%). Solitary fibrous tumor constituted one case and Kimura's disease (Fig.6) constituted one case each (1.85%). Among the malignant tumors, there were four cases of small round cell tumors in adult patients (7.4%), referred to Oncology Institutute and proved to be Non-Hodgkin's lymphoma (Fig.7) on Immunohistochemistry. There were three cases of adenoidcystic carcinoma (5.55%) showing cribriform pattern (Fig.8), two cases of rhabdomyosarcoma (3.70%) (Fig. 9), two cases of extension of carcinoma from adjacent sites (3.70%). Of these one case of squamous cell carcinoma extended into the orbit from paranasal sinuses. (Fig.10) and another case from conjunctival squamous cell carcinoma. Two cases (3.70%) of metastatic deposits were detected, one case from thyroid in a known case of follicular carcinoma of thyroid. (Figs.11 & 12) and another case from unknown primary in a male patient aged 59 years (Fig.13).One case (1.85%) of orbital soft tissue plasmacytoma was reported (Fig.14) in a male patient aged 50 years.

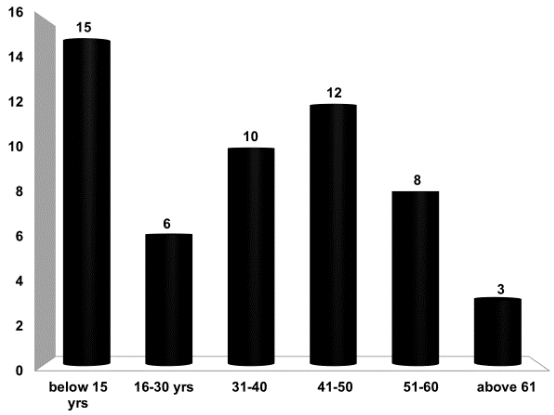

Diagram-1: Shows age distribution of orbital tumors

Table-1: Distribution of cases among adults and children

Age	Number	Percentage
Children	22	40.74%
Adults	32	59.25%

Table-2 : Sex distribution of orbital tumors

Sex	Number	percentage
Males	33	61%
Females	21	39%

Table-3: Histological spectrum and number of cases of orbital tumors

Benign	Cases	%	Malignant	Cases	%
Dermoid Cyst	16	29.6%	Non-Hodgkins Lymphomas	04	7.40%
Cavernous Hemangioma	07	12.96%	Adenoid cystic carcinoma	03	5.55%
Pleomorphic adenoma	06	11.11%	Rhabdomyosarcoma	02	3.70%
Meningioma	04	7.40 %	Direct extension	02	3.70%
Lacrimal gland cyst	03	5.55%	Distant metastasis	02	3.70%
Schwannoma	02	3.70%	Plasmacytoma	01	1.85%
Kimura's disease	01	1.85%			
Solitary Fibrous Tumor	01	1.85%			
Total no. and Percentage	40	74%		14	26%

Table-4: Histological spectrum of childhood orbital tumors

Histological type	Cases	%
Dermoid cysts	16	72.7%
Vascular tumors	04	18.18%
Rhabdomyosarcoma	02	9.09%
Total	22	100%

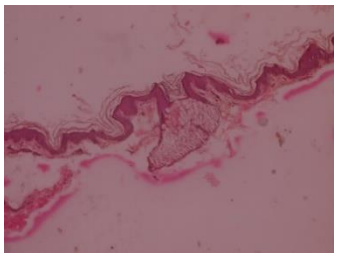

Fig-1: Dermoid cyst

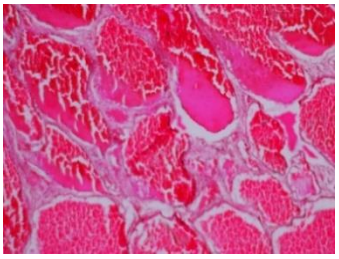

Fig-2: Cavernous Hemangioma

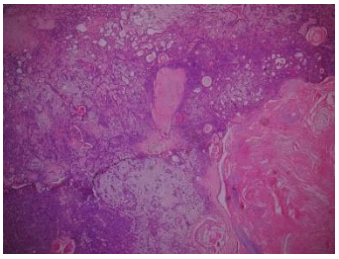

Fig-3: Pleomorphic Adenoma

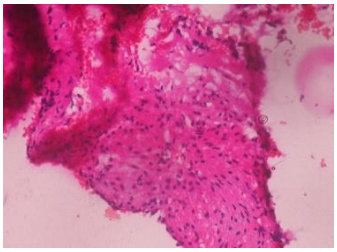

Fig-4: Meningioma

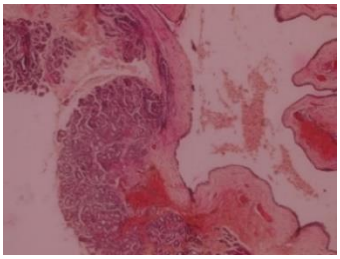

Fig-5: Lacrimal gland cyst

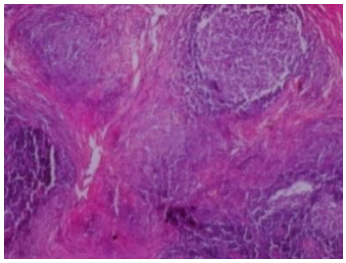

Fig-6: Kimura's disease

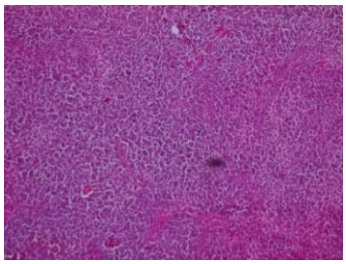

Fig-7: Non-Hodgkin"s Lymphoma

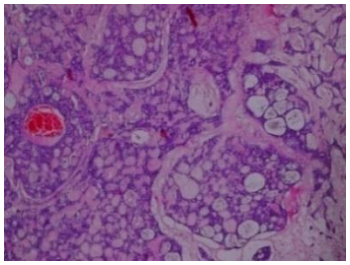

Fig-8: Adenoid cystic carcinoma

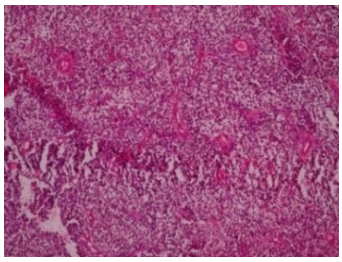

Fig-9: Rhabdomyosarcoma

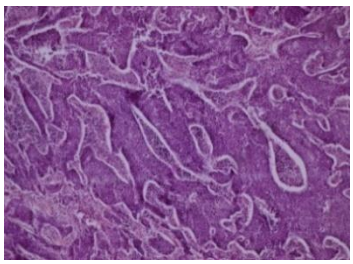

Fig-10: Squamous cell carcinoma

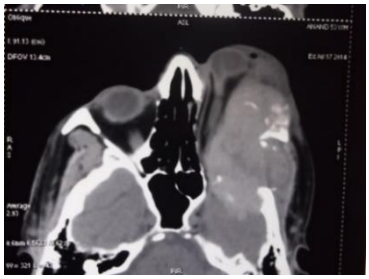

Fig-11: CT scan shows orbital mass

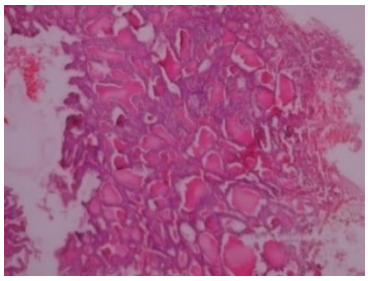

Fig-12: Metastasis of follicular ca thyroid

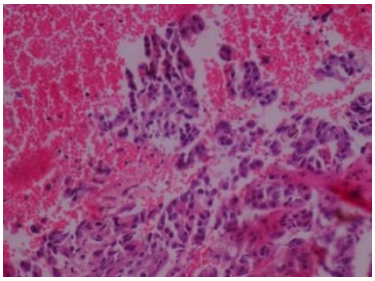

Fig-13: Metstatic deposits unknown primary

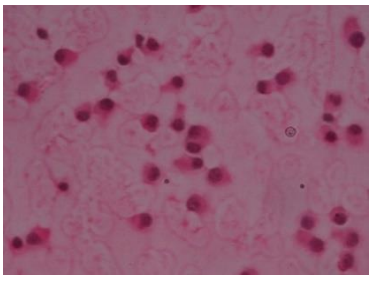

Fig-14: Plasmacytoma cytology

DISCUSSION

Orbital tumors can occur at various age groups, and because of their myriad presentation, their diagnosis poses a great challenge[2]. In our study, the age distribution of 54 orbital tumors showed developmental cysts to peak in the first decade of life, whereas secondary, metastatic and lymphoproliferative lesions had their maximum incidence afther the age of 60. These data are consistent with reports in literature.

Study	Age	Sex
Present Study	Below 15 years, 4th decade	M-61% F-39%
Boriana*et al*. 2007	6th and 7th decade	M-60% F-40%
Radha*et al.* 2005	6th and 7th decade	M-62.5%, F-37.5%
Jasna*et al*. 2004	1st and 7th decade	M-60% F-40%

In the present study, most of the cases occurred below 15 years and in the fourth decade. In the studies of boriana*et al*. [9] andRadha*et al*. peak incidence was in 6th and 7th decades. The incidence of neoplastic lesions has been found to show a bimodal age distribution curve with peaks in the first and seventh decades of life in the study of jasna et al. Our study similarly showed a bimodal peak with the second

peak in fifth decade. The sex distribution in our study correlated with other studies in literature.

Lesion	Present Study	Tanushree	Boriana	Radha	Sheilds
Dermoid cysts	29.6%	33%	12.5%	4.2%	2%
Lacrimal Gland tumors	22.2%	10%	25%	8.2%	9%
Lymphomas	7.4%	04%	16%	33.3%	11%
Cavernous hemangioma	12.96%	10%	04%	16.6%2	6%
Meningiomas	7.4%	06%			4%

Dermoid cysts constituted 29.6% in the present study which correlates closely with the study of Tanushree*et al*. Lacrimal gland tumors in this study, both benign and malignant were close to the study of Boriana*et al*. Lymphomas in our study were 7.4% , the incidence of which is in between the studies of Sheilds*et al.*[10] and Tanushree*et al*. Meningiomas in our study are close to the study of Tanushree*et al*. Most orbital tumors of childhood are distinct from tumors that occur in adults. The most common orbital malignancy in childhood is rhabdomyosarcoma [11,12]. The majority of pediatric orbital tumors are benign and usually include developmental cysts and vascular lesions[13]. Dermoid cysts are the most common benign lesions and rhabdomyosarcoma is the commonest malignancy in our study.The term orbital pseudotumors has been generally accepted to describe inflammatory lesions of orbital tissue. A mixed inflammatory infiltrate with fibrosis of a varying degree is a histopathologic hallmark of orbital pseudotumor[6]. The histologic classifications of ocular infiltrates proposed by Knowles and Jakobiec in 1980 [14], and by World Health Organization in 1993[15] appear to be most acceptable. Kimuras disease i.e., lymphadenopathy with or without a soft tissue mass, also belongs to this group, shows a striking male predilection, and is characterized by dense aggregates of lymphocytes with prominent germinal centers. The lesion is benign although recurrence may develop after surgical excision [16]. One case of Kimura's disease is reported in our study.

CONCLUSIONS

Orbital tumors and pseudotumors occur in various age groups and because of their varied presentation, pose a great challenge in diagnosis. Pathological examination of the surgically obtained tissue specimens gives definitive diagnosis. Orbital tumors are classified into various histological types based on WHO classification. Majority of childhood orbital tumors are benign. Rhabdomyosacoma is the commonest malignancy in childhood. Elderly patients presenting with proptosis should be investigated for malignancy.

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