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Intraorbital Tumors: Morphological and Clinicopathological Spectrum

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Abstract

Introduction: *Pathologies arising within the orbit produce significant symptoms in patients due to its confinement to narrow bony space. Most patients present with proptosis and finally have vision impairment. A variety of benign and malignant lesions affect orbit and the treatment strategies differ.With advancement in techniques both radiological and surgical, it is now easier to pick up orbital lesions at an early stage and help to reach a prompt and accurate diagnosis by pathologists. The present study was conducted to study the morphological types of orbital lesions and itscorrelation with age, sex and clinical features at a tertiary care institute.*

Materials and Methods: *A retrospective cross-sectional study was conducted at SMS Medical College, Jaipur during study periodJanuary 2017 to November 2018.*

Results: *35 cases of intraorbital tumors were reviewed out of which 20 were benign and 15 were malignant. Angiomatous lesions were the most common benign tumors (17.14%) with mean age of presentation 57.1 years. Lymphoma and BCC were the most common malignant lesions. Others included Schwannoma, melanoma and rhabdomyosarcoma. In females most frequent lesions were hemangioma and schwannoma while in males it was lymphoma.*

Conclusion: *Present study shows that benign tumors constitute major bulk of intraorbital tumors, hence a knowledge of all the benign tumors occurring within the orbit saves the patient from over treatment.*

Introduction

Orbit is a narrow space containing eye globe, extra ocular muscles, fat, vascular, nervous and glandular connective tissues, confined by bony cage. Pathologies can arise from any of these structures herein and lead to significant morbidity in patients.[1] Most of the lesions originate between bony wall and extraocular muscle cone and thecommonest presentation is proptosis.The World Health Organization classifies tumors of the orbit into groups according to origin as follows: benign soft tissue tumors, malignant soft tissue tumors, lymphoid and hematologic tumors, pseudotumors and other primary orbital tumors, secondary and metastatic tumors.[2] The epidemiological data of orbital tumors varies between different series. In present era, advancement in technologies both surgical and radiological have made it easier to pick up the orbital lesions at an earlier stage and diagnose accurately by pathologists. Close co-operation between an ophthalmologist,

otorhinolaryngologist, radiologist, oncologist and histopathologist is required for the proper management of orbital lesions.[3]

The present study has been conducted with the aim to study the morphological types of orbital lesions and its correlation with age, sex and radiology at a tertiary care institute.

Materials and Methods

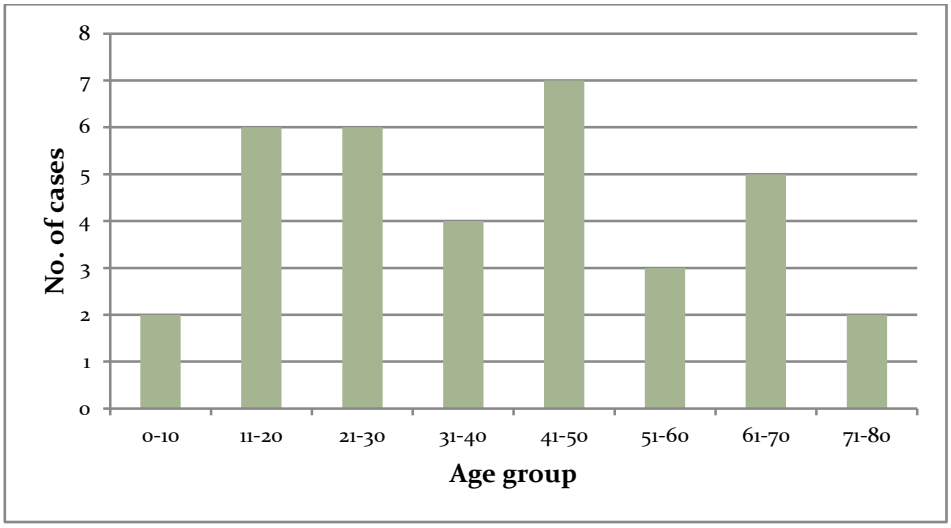
A retrospective cross sectional study was conducted at SMS Medical College Jaipur. Paraffin blocks of intraorbital tumors received during the time period from January 2017 to November 2018 were retrieved. Age and sex of each case was obtained from the case records. Blocks were sectioned and stained with Hematoxylin and Eosin, special stains and immunohistochemistry were applied where ever needed and were examined under light microscope.

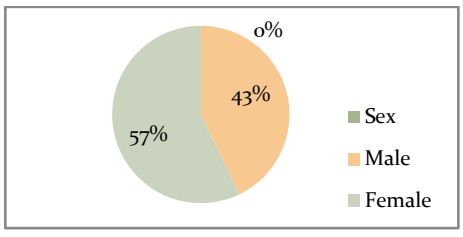
Results

During the study period a total of 35 cases of orbital lesions were obtained. There was a wide age range from1 month to 75 years with the mean age being 38.85 years.

Table1-Age distribution of cases

Age	No of Cases
0-10	2
11-20	6
21-30	6
31-40	4
41-50	7
51-60	3
61-70	5
71-80	2
Grand Total	35





Four cases (11.4%)of orbital tumors were children and 31 cases (88.57%) occured in adults. Males constituted 15 cases (42.85%) and females constituted 20 cases (57.15%) .

Most cases presented with proptosis (82.85%) and palpable mass (77.14%).

Table2- Clinical features

Clinical features	No. of cases	Percentage
Proptosis	29	82.85
Palpable mass	27	77.14
Pain	8	22.85
Defective vision	15	42.85
Restricted mobility	8	22.85
Ptosis	3	8.57

Histopathological spectrum showed 20 cases of benigntumors constituting 57.15%of the total and 15 cases of malignant tumors constituting42.85% of the total orbitaltumors.

Table 3 Distribution of benign tumors

Category	No. of cases	Mean age (years)	Sex distribution	
			Male	Female
Hemangioma	7			
Cavernous hemangioma	6	57.1	2	4
Capillary hemangioma	1	1 month	1	0
Schwannoma	5	27.6	1	4
Inflammatory pseudotumors	2	36.5	1	1
Meningioma	1	30	1	0
Osteoma	1	50	1	0
Chondroid syringoma	1	20	1	0
IgG4 related sclerosing lesion	1	28	0	1
Dermoid cyst	1	18	0	1
Benign lymphoproliferative lesion	1	69	0	1
Total	**20**		**8**	**12**

Angiomatous lesions were the most common benign diagnosis with 7 cases (20%) followed by5 cases (14.28%) of schwannoma. meningioma, osteoma, chondroid syringoma, dermoid cyst and benign lymphoid lesion constituted one case each. There were two cases of inflammatory pseudotumor and one case of IgG4 related sclerosing inflammatory lesion.

Table 4 Distribution of malignant tumors

Category	No. of cases	Mean age (years)	Sex distribution	
			Male	Female
Lymphoma	4	45.25	3	1
Basal cell carcinoma	3	66	0	3
Malignant melanoma	2	46.5	2	0
Embryonal rhabdomyosarcoma	2	10	1	1
Ewing's sarcoma	1	30	0	1
Adenoid cystic carcinoma	1	40	0	1
Round cell tumor	1	35	1	0
Poorly differentiated carcinoma	1	40	0	1
Total	**16**		**7**	**8**

Among malignant tumors, malignant lymphomas were the most common constituting 4 cases (11.42%) followed by 3 cases of basal cell carcinoma (8.57%) and 2 cases of malignant melanoma (5.71%). There were 2 cases of embryonal rhabdomyosarcoma,single case of Ewing's sarcoma, adenoid cystic carcinoma and poorly differentiated carcinoma and round cell tumor.

Table 5 Distribution of tumors in paediatric age group

Category	No. of cases	Percentage
Embryonal rhabdomyosarcoma	2	50%
Capillary hemangioma	1	25%
Schwannoma	1	25%

In paediatric population lesions observed were 2 cases of rhabdomyosarcoma, 1 case of capillary hemangioma and one case of schwannoma.

IHC was used to categorize lymphomas and round cell tumors.

Table 6 Immunohistochemical diagnosis

Morphological diagnosis	Immunohistochemistry	Final diagnosis
Non Hodgkins lymphoma	CD20+ BCL2+ CD3-CD23-CyclinD1-BCL6-MIB-25%	Marginal Zone Lymphoma
Malignant round cell tumor	LCA+ CD20+ CD3- CD5- CD23-MIB-20%	Marginal Zone lymphoma
Non Hodgkins lymphoma	CD20+CD10+BCL6+	DLBCL-GCB type
Malignant round cell tumor	Vimentin+Desmin+ Myogenin+	Embryonal RMS
Melanoma	Vimentin+ S-100+ Melan A+	Malignant melanoma
Malignant mesenchymal neoplasm	CD99+ MIB-15% Desmin-S100-CD34-EMA-	Ewings sarcoma

Discussion

Orbital tumors pose a significant burden of morbidity to population in all age groups. They present in varied ways making the exact diagnosis challenging.[4] We evaluated 35 cases of intraorbital lesions with age range of 1 month to 75 years. Maximum cases occurred in fourth decade. The case of lowest age was 1 month old having capillary hemangioma and highest age was 75 years old having basal cell carcinoma. In the study by Kavitha et al[5] bimodal peak below 15 years and in fourth decade was observed.Studies by Boriana et al[6] and Radha et a1[1]cases peaked at 6th and 7th decade of age. In present study 57.15% cases were females and 42.85% were males. It is in contrast to other studies in which cases were more frequent in males.

Angiomatous lesions especially cavernous hemangioma was the most frequent lesion in present study accounting for 17.14% of all cases which is in accordance with study by Radha et al[1] that reported 16.66% cases and Kavitha et al[5] reported 12.96% cases of hemangioma. Whilethe studies by Tanushree et al[4] and Shields et al[7]reported 10% and 6% cases respectively. The mean age for the cavernous hemangioma was 57.1 years. The other variety capillary hemangioma was observed in 1 month old child. Capillary hemangioma occur in first few months of life and usually regress at puberty. They have a risk of developing amblyopia when deeply seated.

The next most common benign tumors were schwannomas constituting 14.28% of all cases.These are slow-growing peripheral nerve sheath tumors that originates from schwann cells and are rarely located in orbit. These cause progressive painless proptosis and are treated by excision.[8]

Two cases of inflammatory pseudotumor (5.71%) were observed. Orbital inflammatory pseudotumor is a nonspecific, non-neoplastic inflammatory process of the orbit.[9] It is the 3rd most common ophthalmologic disease of the orbit after Graves' disease and lymphoproliferative disorders.It mimicks variety of lesions and is a diagnosis of exclusion. mixed inflammatory infiltrate with fibrosis of varying degree is a histopathologic hallmark of orbital pseudotumor.

Chondroid syringoma is a benign mixed tumor characterized by sweat gland elements in a cartilaginous stroma. This rare tumor accounts for only 0.01% of all primary skin tumors and occurs only rarely in the periorbital region.[10] There was one case of chondroid syringoma of eyelid in our study. Also one case of orbital meningioma, osteoma and dermoid cyst each was observed.

Single case of Immunoglobulin G4-related disease (IgG4-RD) was observed which is immune-mediated disorder affecting almost all major organs of the body including orbit and is characterised by a lymphoplasmacytic infiltrate having IgG4-positive plasma cells, and a variable degree of fibrosis that has a characteristic “storiform” pattern along with elevated serum IgG4 levels.[11]The treatment strategies are still evolving.

Lymphomas were the most common among malignant lesions accounting for 14.28% similar to a study by Boriana et al that reported 16% cases of lymphomas. Shields et al[7] reported 11% cases while Radha et al[1] reported 33.3% cases of lymphoma in their studies. The mean age for the lymphomas was 50 years. Out of 4 cases of lymphoma 3 cases were categorized as extranodal marginal zone lymphomas and one as diffuse large B-cell lymphoma following immunohistochemical studies. Orbital lymphomas have worse prognosis.

The second most frequent malignant tumor was basal cell carcinoma constituting 8.57% of all the cases and a mean age of 66 years. BCC is one of the most common malignant tumors of eyelids but the orbital invasion is rare. There is slight male preponderance and exposure to UV radiation is a known risk factor.[12]One case of malignant melanoma and Ewing’s sarcoma each were observed and confirmed by application of immunohistochemistry. One case of round cell tumor was reported and IHC was advised for categorization. Single case had adenoid cystic

carcinoma arising from lacrimal gland which is a malignant neoplasm occurring in adults, having poor prognosis and treated by exentration of eye.

In females most frequent lesions were hemangioma and schwannoma while in males it was lymphoma. Most orbital tumors of paediatric age group are distinct from tumorsoccuring in adults. The majority are benign and usually include developmental cysts and vascular lesions.[13] The most common orbital malignancy in childhood is rhabdomyosarcoma.[14] In our study out of 4 cases of paediatric age group, 2 were embryonal rhabdomyosarcoma and other 2 cases were benign namely capillary hemangioma and schwannoma. Both the cases of embryonal rhabdomyosarcoma were confirmed by using IHC and showed positive expression for desmin and vimentin.

Though radiological studies provide information about the extent but definite nature of the lesion of can only be assessed by histopathological studies. Benign tumors are treated by simple excision while for malignant tumors, exentration is the treatment modality used. So histopathology becomes essential to decide the line of treatment.

Conclusion

Orbital tumors have myriad clinical presentation. Present study shows that benign tumors constitute major bulk of intraorbital tumors, hence a knowledge of all the benign tumors occurring within the orbit saves the patient from overtreatment.

Limitation

Our study sample size is small. However, a larger sample size will help in recognizing the trends of orbital tumors in local population.

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