

Frequency and Morphological Patterns of Malignant Intra Orbital Tumors in Various age Groups

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Purpose: To determine the frequency and morphologic types of intraorbital tumors.

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Material and Methods: A retrospective study conducted at the department of pathology Basic Medical Sciences Institute, Jinnah Postgraduate Medical Centre Karachi. All intraorbital lesions received over a period of 5 years i.e. from Jan 2005 to Dec 2009 were reviewed. All specimens were formalin fixed, routinely processed for paraffin embedding, sectioned and finally stained with haematoxylin and eosin using standard procedures, when required special stains were performed.

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Results: During study period total of 360 cases of intra orbital lesions were received. Out of, which 115 (31.9%) were non-neo plastic and 245 (68.0%) were neo plastic lesions. Retinoblastoma and squamous cell carcinoma were the most common malignant intraocular and intraorbital tumor with mean age of presentation 4 years and 55 years respectively. Remaining cases include 16 cases of intraepithelial dysplasia, 10 cases of rhabdomyosarcoma, 6 cases each of sebaceous carcinoma and malignant lymphoma, 4 cases each of malignant melanoma, malignant fibrous histiocytoma and adenoid cystic carcinoma, 3 cases of optic glioma and 1 case of mucoepidermoid carcinoma.

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Conclusion: A variety of tumors can involve the orbit. The incidence of neoplastic lesion exceeded that of non-neoplastic lesion. The data will be help full for ophthalmologist, pathologist and epidemiologist for comparison with other studies.

The orbit is an anatomically complex structure containing different types of tissues. The structures present within the orbital cavity are the globe, lacrimal gland, extraocular muscles, smooth muscle, fibroadipose tissue, blood vessels, peripheral nerves, sympathetic ganglia, optic nerve and cartilaginous part of trochlea. The lacrimal gland is the only epithelial structure present within the orbit^{1,2}.

The orbital tumors present with a variety of sign and symptoms and are a great challenge for ophthalmologist in terms of diagnosis, imaging and management³.

The presence of different structures within the

orbit causes great confusion therefore systematic approach is necessary to classify the orbital tumors. The orbital cavity lesions vary from inflammation to different types of neoplastic lesions and greatly affect quality of life^{3,4}. Schematically orbital tumors can be classified as primary, secondary and metastatic. The primary lesions originating from orbit itself include epithelial tumors such as squamous cell carcinoma arising from conjunctiva and mostly seen in elderly patients. Bone and soft tissue tumors including osteomas, fibrous dysplasia, aneurysmal bone cyst and osteogenic sarcoma are more common in younger age group. Among the lymphoid tumors and vascular tumors, hemangioma, lymphangioma, hemangioperi-

cytoma and AVM of vessels are most common tumors in childhood. Nerve sheath tumors within the orbit give rise to neurofibroma, schwannoma and malignant peripheral nerve sheath tumors. These tumors do not arise from optic nerve itself as schwann cells are not present, they arise from peripheral nerves which innervate the extraocular muscles. Secondary orbital lesion, which extend to orbit from neighboring structures include eyelid, conjunctiva, intraocular structures, paranasal sinuses and nasopharynx^{2,5}. Metastatic tumors in children include neuroblastoma, Ewing's tumor, Chloroma and Langerhan's cell and histiocytosis. In adults the tumors which metastasize to the orbit are from breast, prostate, kidney and carcinoid tumors of lung and gastrointestinal tract^{1,2,6}.

Our study aims at reviewing the morphological pattern and determining the frequency of various intraorbital tumors in different age groups.

MATERIAL AND METHODS

A retrospective study was conducted at the department of pathology Basic Medical Sciences Institute, Jinnah Postgraduate Medical Center to determine the frequency of intraorbital lesions received over a period of five years that is from January 01, 2005 to December 31, 2009. During this period total 399 cases were received. All specimens were formalin fixed, routinely processed for paraffin embedding, sectioned and finally stained with hematoxylin and eosin using standard procedures. When required, special stains were performed. All lesions, which occupied the orbital cavity regardless of its site of origin were included. The lesions arising from orbital sides such as skin lesions from eyelids, which did not extend into orbital cavity, were excluded.

RESULTS

During the study period a total of 399 cases were received out of which 39 were inconclusive due to inadequate material. Of the remaining 360 cases, 115 (31.9%) were non-neoplastic and 245 (68.0%) were neoplastic lesions.

The bulk of the non-neoplastic lesions consisted of cystic lesions with 54 cases (46.9%) (Table 1). The rest of cases comprised of inflammatory lesions 44 cases (38.2%), degenerative disorders 11 cases (9.5%), hemorrhage 04 cases (3.47%) and reactive hyperplasia 02 cases (1.7%). Among the neoplastic lesions, 51 cases of benign neoplastic lesions and 194 cases of malignant lesions were found.

Table 2 shows benign neoplastic lesion.

The most common benign neoplastic lesion found to be angiomatic lesion 22 cases (8.8%) followed by 07 cases of pleomorphic adenoma (2.8%). Remaining cases include 6 cases of schwannoma (2.4%), 04 cases of compound naevus (1.6%), 03 cases of meningioma (1.2%) and 02 cases each of neurofibroma and intradermal naevus (0.8%). The rare cases include xanthomatous lesion, lipoma, cystic teratoma, hemartoma and dermolipoma.

The most common malignant tumor found to be retinoblastoma 73 cases (37.6%) with mean age was 3.76 years with equal male to female ratio 1:1 followed by squamous cell carcinoma 67 cases (34.5%) with a mean age 55 years and male to female ratio 1.79 (Table 3-5). Remaining cases included 16 cases of intraepithelial dysplasia (8.2%), 10 cases (5.15%) of rhabdomyosarcoma, 6 cases each of (3.09%) of sebaceous carcinoma, and malignant lymphoma, 4 cases (2.06%) each of malignant melanoma, malignant fibrous, histiocytoma and adenoid cystic carcinoma, 03 cases (1.5%) of optic glioma, one case (0.5%) of mucoepidermoid carcinoma.

Table 1: Non-neoplastic lesions, n = 115

Name of Lesion	No. of Patients n (%)
Cystic lesions	54 (46.9)
Inflammatory	44 (38.2)
Degenerative	11 (9.5)
Hemorrhage	02 (1.7)
Reactive hyperplasia	04 (3.47)

Table 2: Benign neoplastic lesion, n = 51

Name of Lesion	No. of Patients n (%)
Angiomatic	22 (8.9)
Pleomorphic adenoma	07 (2.8)
Schwannoma	06 (2.4)
Compound naevus	04 (1.6)
Meningioma	03 (1.2)
Neurofibroma	02 (0.8)
Intradermal naevus	02 (0.8)
Miscellaneous	05 (2.04)

DISCUSSION

Orbital tumors are an important cause of proptosis, which may result in loss of vision. Present study was designed to evaluate the various morphological patterns of intraorbital tumors and assess the frequencies of various tumors in different age groups.

In our study, the incidence of neoplastic lesions exceeded that of non neoplastic lesions. Among the neoplastic lesions, the age distribution of patients showed two peaks one around 04 years and another around 55 years⁸⁻¹⁰. Of the patients with malignant ophthalmic tumors 58.7% were males and 41.2% females. A high percentage of malignant ophthalmic tumors was observed in paediatric age group due to retinoblastoma, which constituted 37% of all malignant ophthalmic tumors^{10,11,14}. The average age of retinoblastoma was 3.8 years with equal male to female ratio. Studies done in both Pakistan and India also reported same age distribution but with male predominance,^{11,12} while in western countries the age of presentation is earlier. This earlier age of presentation in developed countries is probably due to better diagnostic facilities and increased awareness among public, which is lacking in our continent.

Table 1: Malignant tumors

Name of Lesion	No. of Patients n (%)
Retinoblastoma	73 (37.6)
Squamous cell carcinoma	67 (34.5)
Intraepithelial dysplasia	16 (8.2)
Rhabdomyo sarcoma	10 (5.15)
Sebaceous carcinoma	06 (3.09)
Malignant lymphoma	06 (3.09)
Malignant melanoma	04 (2.06)
Adenoid cystic carcinoma	04 (2.06)
Malignant fibrous histiocytoma	04 (2.06)
Optic Glioma	03 (1.5)
Mucoepidermoid carcinoma	01 (0.5)
Total	194 (79.18)

Table 5: Distribution of malignant neoplastic lesion according to sex

Name of Lesions	No. of Patients	Male	Female	M:F Ratio
Retinoblastoma	73	36	37	1:1
Squamous cell carcinoma	67	43	24	1.79:1
Intra epithelial dysplasia	16	11	05	2.2:1
Rhabdomyosarcoma	10	05	05	1:1
Sebaceous carcinoma	06	02	04	1:2
Malignant lymphoma	06	06	0	6:0
Malignant melanoma	04	03	01	3:1
Adenoid cystic carcinoma	04	02	02	1:1
Malignant fibrous histiocytoma	04	03	01	3:1
Optic glioma	03	03	0	3:0
Mucoepidermoid carcinoma	01	01	0	1:0

Among the epithelial tumors, Squamous cell carcinoma originating from conjunctiva in adults contributing to 34% of malignant ophthalmic tumors closer to some studies^{12,13}. However Singapore reports melanoma⁹ and Nepal reports Basal cell carcinoma¹¹ as most common tumors. The mean age of Squamous Cell Carcinoma in our study was 55 yrs with male to female ratio 1.79:1¹³.

Intraepithelial dysplasia was seen predominantly in males mostly between 51-60 years as reported in other studies¹⁰. Sebaceous gland carcinoma mostly arises from meibomian gland within eyelid and also from the conjunctival epithelium constitutes (3.09%) of intra orbital malignant tumors. Due to its invasive nature, it is difficult to ascertain the exact origin.

In the western population melanoma is one of the most common ocular malignancy. The frequency of malignant melanoma was relatively low in the present

Table 4: Distribution of malignant neoplastic lesions according to Age

Name of Lesion	No. of Patients	01 - 10	11 - 20	21 - 30	31 - 40	41 - 50	51 - 60	60 >
Retinoblastoma	73	73	—	—	—	—	—	
Squamous cell carcinoma	67	—	2	7	6	9	19	24
Intra epithelial dysplasia	16	—	—	1	3	2	8	2
Rhabdomyo sarcoma	10	9	1	—	—	—	—	—
Sebaceous carcinoma	06	—	—	—	—	2	1	3
Malignant lymphoma	06	—	1	—	1	1	—	3
Malignant melanoma	04	—	—	—	—	1	2	1
Malignant Fibrous Histiocytoma	04	—	—	1	1	1	1	—
Optic glioma	03	3	—	—	—	—	—	—
Adenoid cystic carcinoma	04	—	—	—	2	—	2	—
Mycoepidermoid carcinoma	01	—	—	—	—	—	—	—
Total	194	85	04	09	13	17	33	33

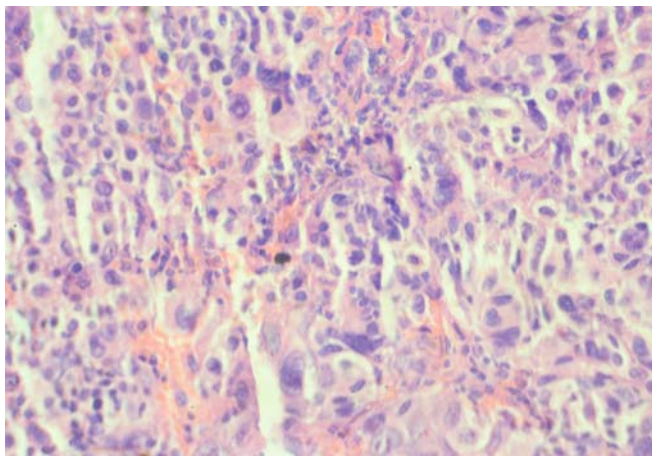


Fig. 1: Malignant Melanoma x 40

study. Ocular melanoma mostly effects white or lightly pigmented individuals and rarely occurs among Asian or black population¹⁴. We found 2.1% of our study cases to be malignant melanoma. These findings are closer to study in subcontinent¹⁰ but in contradiction to studies done in Singapore and Taiwan^{9,14} the frequency is lower.

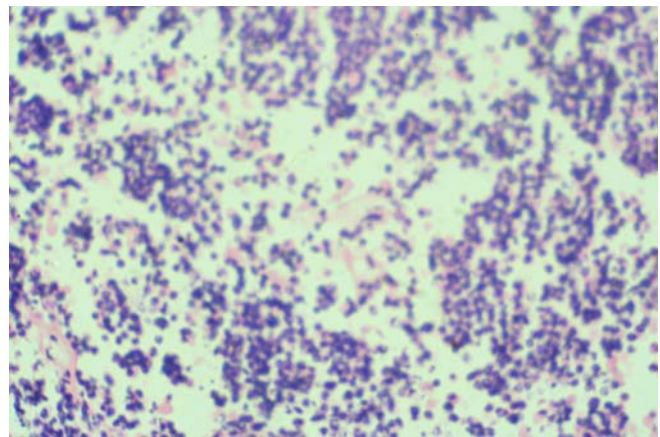


Fig. 2: Retinoblastoma x 40

Among the mesenchymal tumors, Rhabdomyosarcoma was observed as 5.15% in 1 to 10 yrs age group, which is lower in comparison with other Pakistani studies¹². The reason for lower number of cases might be due to the fact that our study is limited to a single medical institute. In adults, the most common

mesenchymal tumor was found to be malignant fibrous histiocytoma. Our experience regarding malignant lymphoma show only 3.06% with mean age at diagnosis was 49 yrs. These results are also lower than other studies^{12,14}.

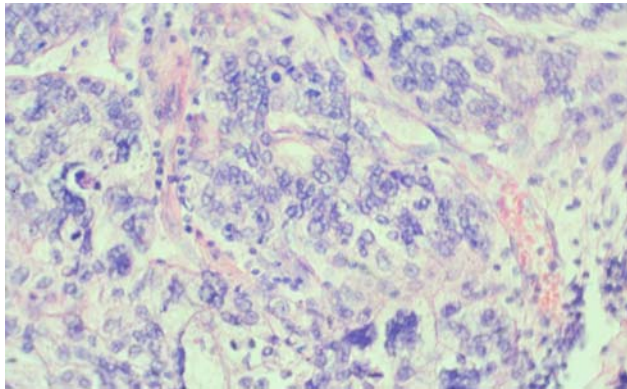


Fig. 3: Hemangiopericytoma x 40

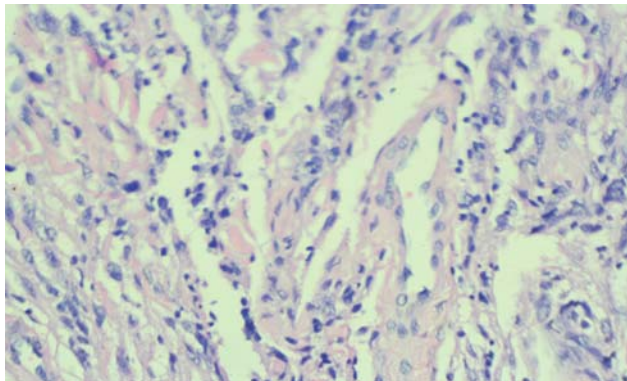


Fig. 4: Benign Angiomatous lesion x 40

Among the neuroepithelial tumors, optic glioma constitutes 1.5% of malignant cases and found to be closer to other study in Pakistan⁸.

CONCLUSION

Retinoblastoma was the most common ocular malignant tumor in paediatric age group in our study. Squamous cell carcinoma is the commonest intra orbital malignancy in the older population. This data

provides help and is also useful for clinicians and ophthalmologist for future references.

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