Study of Eye ball and orbital tumors

Dr.Anjali P. Ingle[1] , Dr. Neeta Avinash Wahane[2]
[1]Assistant Professor, Department of Pathology, B.J.G.M.College, Pune, India
[2]Associate Professor, Department of Pathology, B.J.G.M.College, Pune, India
Email - drwneeta@yahoo.com

Abstract: *Introduction: Diseases of the eye and orbit create some of the most complex and perplexing problems. Aims and objectives: The present study aims at evaluating the tumors of the eye and orbit and to highlight various clinical, pathological and prognostic aspects of them. Methodology: This was a clinico-pathological study of tumors of eye and orbit carried out in tertiary care hospital. Histopathological samples of tumor and tumor like lesions of eye and orbit were studied. Results: Out of total 39 cases studied, 7 cases were benign and 32 cases were malignant. Intra-ocular tumors formed largest group. Retinoblastoma (26 cases) formed the largest group. Maximum number of tumors was in the age group of 1-10 years. There were 3 cases of benign neural tumor. There were 4 interesting cases with clinical presentation as tumor like lesions but on histology came out to be 3 cases of pseudo tumors and 1 case of coat's disease. Thus study emphasizes biopsy from tumor like lesions to save eye from excision. Conclusion: Expanding lesions in eye and orbit constitutes a diagnostic challenge. Early diagnosis and detection is the only answer to achieve a favorable prognosis.*

Key words: *Eye ball tumors, orbit.*

1. INTRODUCTION:

Ocular oncology differs from others since majority of the primary tumors of the eye do frequently manifest in infancy and childhood[1]. Of all these ocular neoplasm the most common and fatal is retinoblastoma. It is responsible for approximately 1% of all deaths from cancer in the age group under 15 years[2&3].Tumors of the orbit are not a common occurrence in ophthalmic practice[3]. Occasionally tumor like lesions present clinically as neoplasm's contributing to the histological curiosities. More than ever, the clinician must discuss the management of ocular and orbital tumors with pathologist before surgery, so that the tissue is handled in optimal fashion for the most reliable and precise diagnosis and realistic prognosis. The strides which we have made in our knowledge of eye and orbit tumors have been extra-ordinary. Considering this wealth in present institute for pathological examination, we have evaluated tumors of eye and orbit to highlight the actual incidence of eye and orbit tumors according to different age and sex. Also we evaluated cases according to their various clinical presentation, histo- pathological appearances and patterns of tumors with their prognostic aspects. We had also studied tumor like lesions which on histology came out to be cases of pseudo-tumors and coat's disease.

2. MATERIAL AND METHODS:

This was a clinico- pathological study of eye ball and orbital tumors carried out in tertiary care hospital. It was performed over a period 2 and ½ years from June 2009 to December 2012. The present study included analysis of 39 cases of eye and orbit tumors. Proforma of each case included detailed history, findings of local examination, and other ophthalmic investigations along with gross and microscopic examination of each specimen. Biopsies from lesions were processed. Enucleated Eye balls and exenterated specimens were fixed in 10% formalin 20-25 times volume for 48-72 hours. It was kept in 60% alcohol until restoration of color of the specimen. Complete description of eye including external appearance, scars, injuries and operated marks were recorded. Eye balls were opened with flat razor blade several millimeters from either side of the optic nerve passing through the cornea at limbus. Central block with optic nerve was used for processing[4]. In case of Retinoblastoma and other intra-ocular tumors cut surface was observed for size, gross appearance and extent of tumors. Section of eyeball for histology was included with the optic nerve. Haematoxylin and Eosin stained sections were studied.

3. RESULTS:

This was a clinico- pathological study of eye and orbit tumors carried out in tertiary care hospital. It was performed over a period 2 and ½ years from June 2009 to December 2012. The present study included analysis of 39 cases of eye and orbit tumors. Below table describes distribution of lesions as per their histological nature and WHO system.

Table 1: Distribution of lesions as per their histological nature and WHO system[4]

Group.	Types of tumor	Benign	Malignant	Total
Primary Intra-ocular	Retinoblastoma	0	26	26
	Melanoma	0	2	2
	Medulloepithelioma	0	1	1
Tumors of Orbit	Lymphoma	0	1	1
	Benign Neural Tumors I)Neurofibroma II) Schwannoma i) Neurilemmoma	2 2 1	0 0 0	3 3
Secondaries	Secondaries from ca Breast	0	1	1
	Leukemic infiltrates	0	1	1
Tumor like lesions	Coat's disease	0	0	1
	Pseudo tumors			3

Above table indicates that out of total 39 cases, retinoblastoma formed largest group followed other tumors in descending order of their incidence. The lesions were divided into three groups 1) Primary 2) Secondary 3) Cases clinically presented as tumor but diagnosed otherwise after histopathological examination. In the primary group of 33 cases, intra-ocular tumors formed largest group 30 cases. There were 3 cases of orbital neural tumors, of which 2 were neurofibroma. Both the cases of neurofibroma did not reveal any evidence of von-Recklinghausen disease. There were 2cases of secondary deposits in eye of which 1 was from breast carcinoma and other was from leukemic infiltrate. Study included 3 cases of pseudo tumor and 1 case of coat's disease as tumor like lesion.

Table 2: Distribution of cases of Retinoblastoma

Age in years	Male		Female		Total	Total
	cases	Percent	cases	Percent	cases	Percent
<=1	1	3.84%	0	0%	1	3.84%
1-3	7	26.92%	7	26.92	14	53.54%
3-5	3	11.54%	3	11.54%	6	23.08%
5-7	3	11.54%	0	0.00%	3	11.54%
7-9	1	3.84%	0	0.00%	1	3.84%
9-11	1	3.84%	0	0.00%	1	3.84%
	16	61.54%	10	38.46%	26	100%

In the above table the present study showed that the incidence of retinoblastoma was more between 1-3 years, nearly 90% were diagnosed with retinoblastoma before 6th year of age. The reason for this can be said that though the tumor may present with growth within few days after birth, a sub acute course of this malignancy makes its detection only possible only when the parents detect some deformity in the eye. Males (61.54%) were commonly affected than females (38.46%). There were 24 cases which presented unilaterally in either of eye, out of which 14 cases in males and 10 cases in females. Mostly the patients of retinoblastoma present with proptosis and white reflex.

Table 3: Distribution of cases of Pseudo tumors

Case no.	Age	Sex	Signs and symptoms	Laterality	Histological typing	Nature of specimen
1	45yrs.	Male	Proptosis poor vision	Left eye	Excision biopsy	Predominantly Lymphocytes
2	20yrs.	Male	Proptosis poor vision redness	Right eye	Enucleation	Predominantly Lymphocytes
3	55yrs.	Female	Proptosis poor vision redness	Left eye	Enucleation	Predominantly polymorphous

Study included 3 cases of pseudo tumors presented with proptosis, redness of eye and poor vision. Out of which two were predominantly lymphocytic variety and one was of granulomatous variety. Cases didn't reveal any systemic

pathology. As coat's disease forms one of the differential diagnosis of Retinoblastoma, present study emphasizes biopsy from these lesions for definitive diagnosis and conservative management to save the eye.

4. DISCUSSION:

Eye and Orbital tumors are an important cause of proptosis which may result in loss of vision[5]. Present study was designed to evaluate the various morphological patterns of intraorbital tumors and assess the frequencies of various tumors in different age group. In our study the incidence of neoplastic lesions exceeded that of non- neoplastic lesions. Among the neoplastic lesions two peaks seen. Of the total ophthalmic malignant tumors 56.41% were males and 43.58% were females. A high percentage of malignant ophthalmic tumors were observed in pediatric age group due to retinoblastoma[6, 7, &8]. Detailed analysis of 39 cases of tumors of eye and orbit were carried out over a period of 2 and ½ year. Study included clinical evaluation, investigations, Histopathological and prognostic aspects of each case in detail. Retinoblastoma formed the largest group with 26 cases (59.06%) and data analyzed as to age, sex, and mode of presentation, laterality and extent of lesion in accordance with those classically described. In the present study the average age of presentation was between 1-3 years with male: female ratio was 1.6:1. Males (61.54%) were commonly affected than females (38.46%). Tahira et al found out 73 (37.6%) cases of Retinoblastoma with male: female ratio was 1:1[5]. Studies done in India and Pakistan also reported same age distribution but with male predominance[6&9]. It is stated that prognosis in Retinoblastoma is very favorable with solitary lesions less than 4 disc in diameter. It is obvious that the patients with retinoblastoma present with proptosis followed by white reflex only one case has orbital recurrence and these are consistent with previous studies. We wish to state that the duration of disease had no direct relation with the clinical presentation. It will require further detail analysis of other factors like family history, general health and anaplasia of the malignancy which may be contributing to rapid spreading in few cases and vice- versa. In our study all retinoblastoma tumors were endophytic type fig1. We divided them into two groups 1) differentiated and 2) Undifferentiated with other associated features like necrosis, calcification and optic nerve infiltration. In our study the histopathological correlation and clinical presentation was one of the aim. We had 26 specimens among which 8 were differentiated (Figure 1), 18 were undifferentiated (Figure 2) 4 cases showed necrosis and 4 cases showed calcification. We found in our study 8 cases of retinoblastoma with optic nerve involvement (Figure 3). It was concluded that there was no relation between histological grade and invasive capacity of the tumor. Thus in conclusion data analyzed were in accordance with those classically described. We also studied cases clinically mimicking like Retinoblastoma which on histology came out to be Coat's disease (Figure 4) and Medulloepithelioma (Figure 5). Ocular melanoma mostly effects white or lightly pigmented individuals and rarely affects Asian or black population[10]. We found 2.1% cases of Malignant Melanoma (Figure 6). These findings are closer to study in subcontinent[6]. Present study included 3 cases of pseudo tumors (Figure 7) with conservative management by steroids...Thus the present study emphasizes the importance of biopsy diagnosis of tumor like eye lesions to save the eye. One case of lymphoma (figure 8).

5. SUMMARY AND CONCLUSION:

Expanding lesions in eye and orbit constitutes a diagnostic challenge. Retinoblastoma forms the major group. Early diagnosis and detection is the only answer to achieve a favorable prognosis. Pseudo tumors are the clinical as well as pathological curiosities. Studies also emphasizes that this is a group which includes variety of lesions showing diverse histomorphology. Further studies may give clue regarding their etiology. Interesting cases like Medulloepithelioma, unsuspected melanoma, Coat's disease which was clinically confused with follicular keratosis. Hemangioma confused clinically as melanoma made the study even more fascinating.

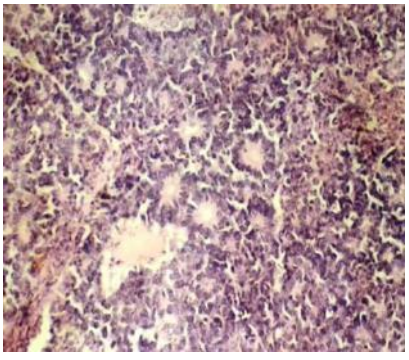

Figure1.Retinoblastoma –showing well formed true and pseudo rosettes x100

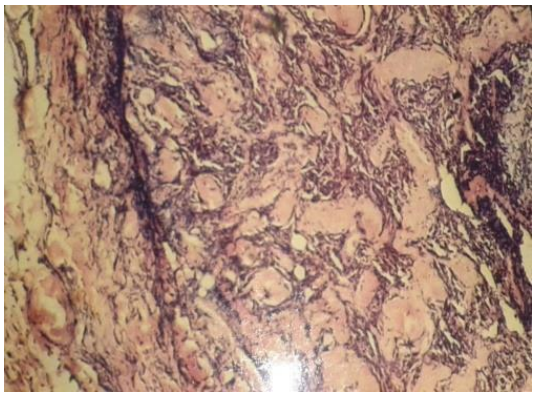

Figure2. Retinoblastoma showing optic nerve infiltration x100.

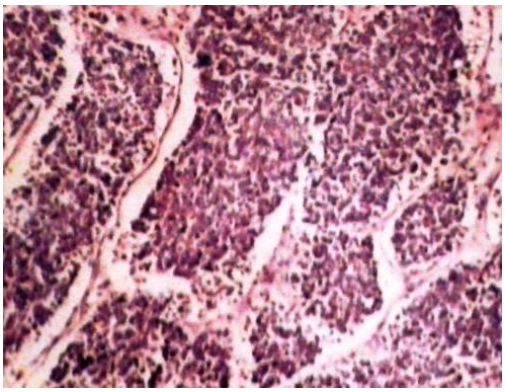

Figure 3: Undifferentiated Retinoblastoma showing round tumor cells arranged in groups and sheets separated by fibrous septae x100.

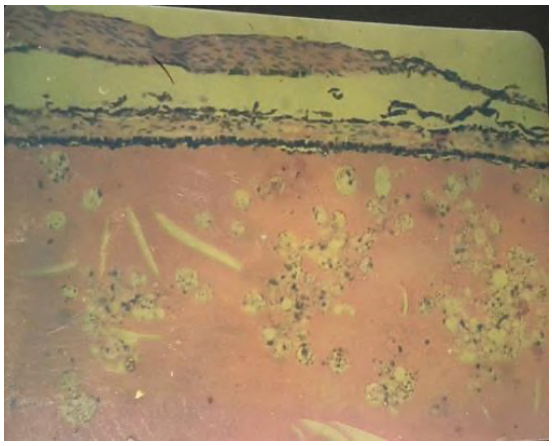

Figure 4: Coat's disease - showing sub retinal exudates containing cholesterol clefts and foamy histiocytes with choroid and sclera.x100.

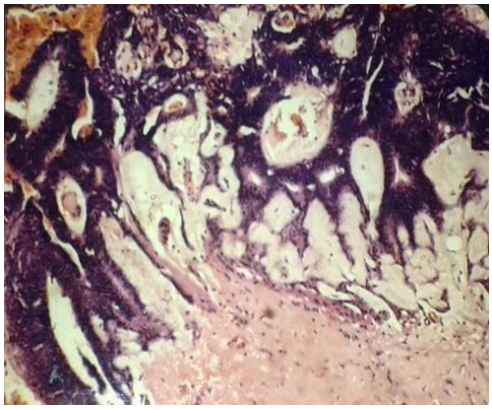

Figure5. Medulloepithelioma- showing tumor tissue comprised of single and multilayered epithelium that forms festoons and tubular structures x100.

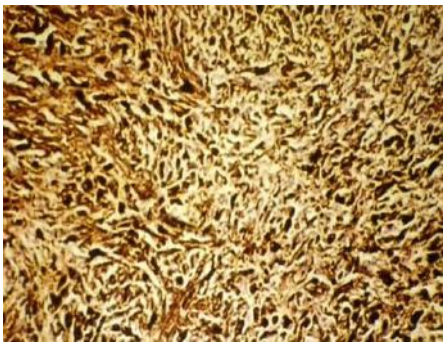

Figure 6: Malignant Melanoma- shows tumor tissue formed by oval and spindle shaped cells arranged diffusely with massive deposits of melanin x 100.

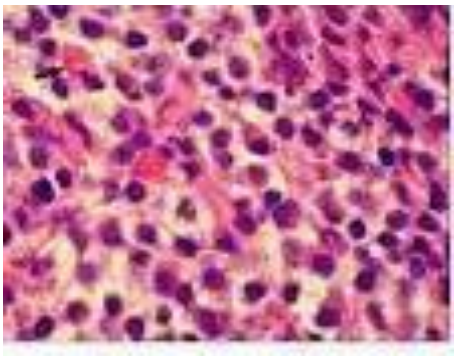

Figure 7: Pseudo-tumor of eye- showing sheets of small round cells having scanty cytoplasm invading fibro-collagenous tissue x100.

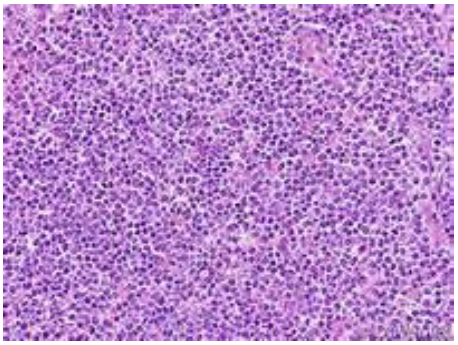

Figure 8: Lymphoma of eye –showing diffuse sheets of small round cells with oval nuclei and scanty cytoplasm with little variation in size and shape x100.

REFERENCES:

1. KincaidM.C.: Ocular and orbital tumors, Ophthalmology: 95, 1588-1595, 1988
2. Bukhari S et al: Presentation pattern of Retinoblastoma. Pak J Opthalmol, 27, 142-5.
3. Shridevi Subramanian et al., Presentation of Retinoblastoma Patients in Malaysia Asian Pac J Cancer Prev, 15 (15), 7863-7867.
4. Zimmerman L.E, et al: Histopathological typing of tumors of the eye and its adnexa World health organization: No. 24, 1980.
5. Táhira Soomro et al, Frequency and Morphological Patterns of Malignant Intra- Orbital Tumors in Various age groups. Pak J Opthalmol 2011, Vol 27 No. 4
6. N.Uddin, Mushtaq N, Khan MAH, et al. Morphological pattern of ophthalmic tumors in Northern Pakistan. J.Pak.Med Assoc. 2001; 51:19-22.
7. Sunderraj P. Malignant tumors of eye and adnexa. Indian J. of Ophthalmol.1991; 39:6-8.
8. Lee SB, Eong KG, SawSM, et al. Eye cancer incidence in Singapore.Br J Ophthalmol. 2008; 84:767-70.
9. Lavaju P, Arya SK, Sinha A, et al. Pattern of ocular tumors in the eastern region of Nepal.NepJ Ophthalmol. 2009; 1:9-12.
10. Cheng CY, Hsu WM. Eye – Incidence of eye cancer – an 18 year review.2004; 18: 152-8.