A Clinico-Pathological Study of Paediatric Primary Tumour of Eye & Orbit

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Abstract

Introduction: Tumors occurring in pediatric age group comprises about 2% of all tumors. Ocular tumors represent a different histological spectrum in children than in adults. Diagnosis of ocular tumors in initial stages in pediatric age group is a challenge for the opthalmologists. Early diagnosed tumors have higher cure rates due to immediate treatment. Incidence of orbital tumor is -3.5-4%. Orbito-ocular tumors are unsightly and may contribute to visual disturbances. Visual loss in children has implications for all aspects of the child's development.

Materials & Method: This is the prospective study including 80 registered cases below 12 years of age at M&J western regional Institute of Ophthalmology , B.J. Medical College & Civil hospital , Ahmedabad. Histopathological examination was carried out on all the specimens received and the results were noted and analysed.

Results: Out of 80 cases, 37 were benign (46.2%) and 43 were malignant (53.7%).43 were males and 36 were females. Most common site involved was Retina and Retinoblastoma was the most common tumor found. Rhabdomyosarcoma was the most common extra-ocular tumor. Dermoid cyst was the most common benign lesion.

Conclusion: In this study, maximum number of cases fall in the age group 0-6 years of age. Retinoblastoma is the most common tumor found in the pediatric age group followed by Dermoid cyst.

Keywords: *Pediatric age group, ocular tumors, malignant, benign, vision loss.*

Introduction

Tumors occurring in pediatric age group comprises about 2% of all tumors.Ocular tumors represent a different histological spectrum in children than in adults.Although thought to be rare, they contribute to significant cause of morbidity and mortality.[1]It includes many cystic and solid masses involving eye and orbit that may become apparent at birth or are acquired later on. Diagnosis of ocular tumors in initial stages in pediatric age group is a challenge for the ophthalmologists. Tumors that have been diagnosed earlier, have higher cure rates due to more immediate treatments.[2-4].How fast treatments can start depends on how early the patient is diagnosed, which can be performed even quicker if the characteristics of the pediatric eye tumor patients are known.[2,4-6]

An appropriate approach is by classifying the tumors as per their location: Eyelids, lacrimal glands, conjunctiva, cornea, intraocular tissues (iris, ciliary body , choroid,retina) and orbit (ocular globe,optic nerve and its meningeal coverings, extraocular muscles, blood vessels, fibroadipose connective tissue, tenon's capsule)

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Incidence of orbital tumor is -3.5-4%.[1]Orbito-ocular tumors are unsightly and may contribute to visual disturbances. Visual loss in children has implications for all aspects of the child's development.[1,7]Malignant tumors in children and adults are biologically and histologically distinct. [1]For example Melanomas which are most frequent primary intraocular tumors in adults are rare in children wheres retinoblastoma are most common in this age group. The most common benign tumor seen is dermoid cyst. Both benign and malignant masses of the orbit can have bone destruction[1,8].

The aim of this study is to find out the incidence of various Paediatric tumors of eye and orbit as per age ,sex and location and to study pathological features which may aid in early diagnosis of pediatric eye tumors and prevent visual morbidity and mortality.

Material and Method

80 prospectively registered cases from July 2017 to April 2019 were included in the study which was carried out at Histopathology department, M.&J.western regional institute of Ophthalmology, Civil Hospital, Ahmedabad.

All cases included were below or of 12 years of age. The clinical details including chief complaints, signs and symptoms of the cases were obtained.Cases showing tumor on investigations like X-Ray, Ultrasonography, Computed tomography scan, Contrast Enhancing CT scan, Magnetic Resonance Imaging were included.

Tumor tissue was fixed in 10% formalin. Gross examination was carried out and findings were noted. Multiple tissue sections were taken, then processed, embedded and slides were prepared. In case of small biopsy, whole specimen was given. Microscopic examination was carried outand the final result was noted after taking into consideration both gross and microscopic findings.

Results

The data of this study of 80 cases was tabulated as per agewise, sex-wise, location wise ,type of neoplasm as well as various morphological types of neoplasm.

Out of 80 cases, 43 cases were malignant tumors (53.75%) whereas 37 cases were benign lesions (46.2 %).

Out of 80 cases, the highest number of cases were seen in the age group 3-6 years (table 1) The highest age was 12 years whereas the lowest age was 2 months old.

Out of 80 cases, 44 were males while 37 were females. There was slight male preponderance seen in the age group of 3-6years which had highest number of cases.

Table 1: Age and Sex wise distribution of Tumors

	Male	**Female**	**Total**
Age	**Male**	**Female**	**Total**
0-3 years	10	3	13
3-6 years	18	16	34
6-9 years	6	5	11
9-12 years	10	12	22
Total	44	36	

Out of this 43 cases, the most common malignant lesion was retinoblastoma followed by Rhabdomyosarcoma as shown in the table 3. The most common benign lesion was Dermoid cyst followed by Benign MelanocyicNevus(table 2).Slight male preponderance was noted for retinoblastoma (table 2). Out of 37 cases of retinoblastoma, 4 cases were bilateral and 33 were unilateral.. Bilaterality was found more common in female (i.e. 3 cases)

Table 2: Frequency of various tumors: Age and sex wise.

Diagnosis	**M**	**F**	**0-3 years**	**3-6 years**	**6-9 years**	**9-12 years**
Dermoid Cyst	9	8	3	4	5	5
Benign Melanocytic Nevus	2	5	0	1	2	4
Neurofibroma	3	1	0	0	0	4
Dermolipoma	2	1	1	0	0	2
Epidermal CYST	0	2	0	0	0	2

***Cont.*. Table 2: Frequency of various tumors: Age and sex wise**.

Lobular Capillary Hemangioma	2	0	0	0	0	2
Solitary Fibrous Tumor	1	0	1	0	0	0
Lipofibroma	1	0	0	0	0	1
Retinoblastoma	20	17	8	27	1	1
Embryonal Rhabdomyosarcoma	2	1	0	0	3	0
Medulloepithelioma		1	0	1	0	0
Myeloid Sarcoma	1	0	0	1	0	0
Adenoid Cystic Carcinoma	1	0	0	0	0	1

Table 3 shows the various tumors distributed as per the location where in the most common tumors are intra-orbital involving retina i.e. Retinoblastonma.

Table 3: Location wise distribution of tumors

Location	Total cases
Retina	37
Eyelid	30
Conjuctiva	6
Extra-ocular muscles	3
Orbit	3
Lacrimal Gland	1

Discussion

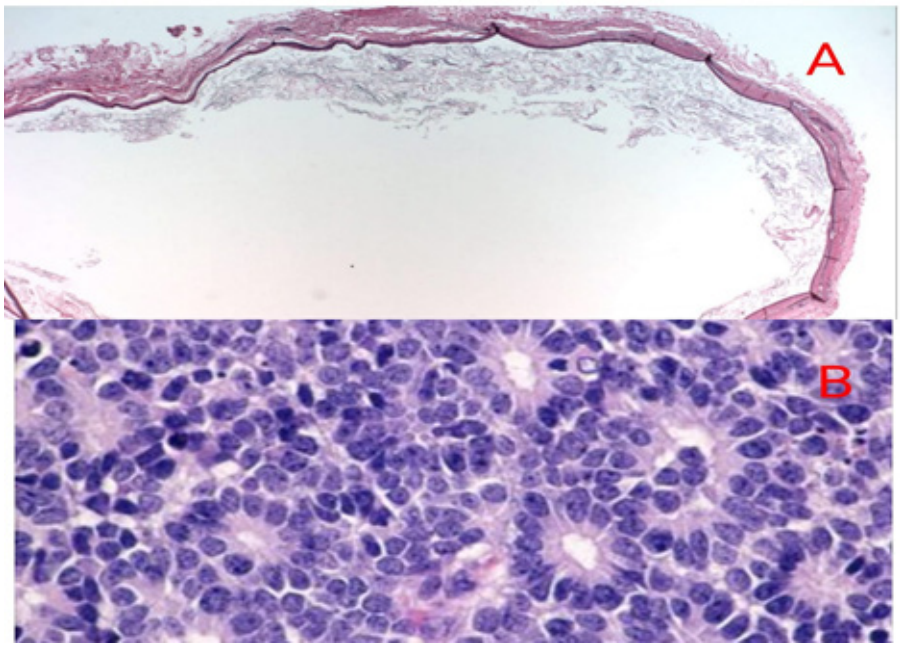
Tumor and tumorlike lesions of orbit in children represent a different histologic spectrum than in adults;both benign and malignant masses of the orbit can have bone destruction,[1,8]and can lead to deformity and vision loss, that's why it is very important to diagnose early for these intraorbital tumors in children.[1]

This study of 80 cases was based on histopathological examination of the specimens received and its clinical and radiological correlation. The results obtained after proper analyzing were compared with results of the earlier studies by various authors. As observed, 46.2% cases were benign and 54.75% cases malignant which was in concordance with Mohan et al[9] who had reported an incidence of 28% and 72% of benign and malignant tumors from a study of 68 orbital tumors and Modi PJ et al[1], study of49 cases of malignant tumors (65.33%), 26 cases of benign tumors (34.66%).

Benign tumors commonly presents with complaints of mild pain, dimness of vision or for cosmetic purpose whereas malignant presents with proptosis, leucocoria or vision loss. Dermoid cyst is smooth, painless,slowly progressing cystic mass which typically involves the upper eyelid along the lateral brow margin. It is lined by stratified squamous epithelium and filled with keratin debris,sebum and hair (Figure 1 A). Dermoid cyst (45.9%) was the most common benign tumor reported in this study which is in concordance PieterJuanarta et al [2]where it was 31.0%.

Benign Melanocytic Nevus is the second most common Benign tumor observed in this study.They are observed in cutaneous or conjunctival side of eyelids. All variants can occur but most of them are junctional nevus.

Neufibromas can present as isolated cases or associated with von Recklinghausen disease.Although believed to be present from birth, these tumors frequently show accelerated growth during childhood or later[18]. In our study, 4 cases were found in the age of 9-12 years of age.A rare case of Solitary fibrous tumor was observed arising in orbit in the study. It is the tumor consisting of alternate hypercellular and hypocellular areas along with keloid like collagen.



[Figure :1 Microscopic Examination of (A) Dermoid cyst (B) Retinoblastoma]

A high percentage of malignant ophthalmic tumors was observed in pediatric age group due to retinoblastoma[10]which is the most common intraocular malignant tunor in this age group. In this study retinoblastoma was the most common malignant tumor found which is in agreement with various studies like Broaddus E et al[11], Pieter Juanarta[2], Modi PJ et al[1]. Patients presents with leukocoria or strabismus.It may be endophytic or exophytic type. Flexner-Wintersteiner rosettes, Homer-Wright rosettes and fluerettes can be seen on microscopic examination(Figure 1B). Necrosis is often extensive and small foci of calcification in areas of necrosis can be appreciated by X-Ray.

It can present at birth but is usually diagnosed around 1-2 years of age Highest number of cases are in the age group 3-6 years(34 cases).In the study Pieter Juanarta et al [2] highest incidence was among 1-4 years age group.

Table :4 Distribution and comparison of retinoblastoma in most common age group 0-5 years.

	Present study (Out of 37 cases)	**Gracy Ramchandran et al12 (Out of 18 cases)**	**Modi PJ et al1 (Out of 46 cases)**
No.of cases	35	15	41
Percentage	94.5%	83.3%	89.13%

0-5years is the most common age for Retinoblastoma and the results in this age group is compared with other studies (table: 4) and the results are in agreement. Out of 37 cases of retinoblastoma 20 were male and 17 were female in discordant with study of Susan S. Devasaet al[13] where out of 61 cases, 26 were males and 35 were female.

Retinoblastoma can be unilateral or bilateral. Incidence of unilateral cases is more than bilateralas shown in (table 5). Bilaterality was found more in females as compared to males which is in concordance with various studies (Table 5).

Table 5: Analysis of laterality of retinoblastoma as per sex distribution.

Laterality	Susan S. Devasa Et Al[13] Study			Modi Pj Et Al[1] Study			Present Study		
	Total	Male	Female	Total	Male	Female	Total	Male	Female
Unilateral	50	24	26	42	25	17	33	19	14
Bilateral	11	2	9	4	1	3	4	1	3

Retina was the most common site for primary tumors followed by eyelid in pediatric age group. In Pieter Juanarta et al [2],66.7% retinal tumors were noted.

Embryonal Rhabdomyosarcoma has been described in the literature as the commonest soft tissue malignancy of the orbit among the pediatric patient [14]accounting for 10% of all rhabdomyosarcoma cases with mean age of 6-8 years of age[15,16] which is similar to the results of present study. Embryonal and alveolar types are seen .Male to female ratio is 2:1 in the present study while it was 1:1 in series by Tahira Soomro et al[17]

One case of Myeloid Sarcoma also known as Granulocytic Sarcoma presented with proptosis and normal CBC findings. One of Medulloepithelioma was also seen which is 2nd most common Intra-ocular round cell tumour tumor in pediatric age.Adenoid Cystic Carcinoma seen in this study is relatively common in lacrimal gland.

Conclusion

To conclude, in this study 80 cases of pediatric ophthalmic tumors were studied. Of which 46.2% were benign and 52.75% were malignant.Dermoid cyst was the most common benign tumor..

Retina was the commonest site for malignant primary ophthalmic tumors and so is Retinoblastoma , the commonest malignancy with maximum number of cases between 0-5 years..Rhabdomyosarcoma was the most common orbital tumor

In case of pediatric opthlamic tumors,as the symptoms are not well appreciated, early diagnosis of tumors by histopathological examination can help in preventing vision loss.

The obvious incidence and features of certain eye tumors and the rarity of others can become helpful in the field of research pertaining to etiology, pathology and epidemiology of tumors.

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