ORIGINAL ARTICLE

ORBITAL TUMORS IN CHILDREN: A DESCRIPTIVE STUDY AT TERTIARY CARE CENTRE

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ABSTRACT

Incidence of orbital tumor is -3.5-4%. Tumors and tumorlike lesions of the orbit in children represent a different histologic spectrum than in adults. In the pediatric population, a delay in diagnosis of orbital tumors, even if benign, can lead to vision loss and deformity. Purpose of this study is presentation of epidemiology, clinical signs and symptoms, frequency and histopathology of orbital tumors in children. The most common clinical findings are leukocoria (white pupil) & proptosis. Out of 75 cases studied, retinoblastoma was the most common malignant tumor (61.33%) while dermoid as most common benign tumor(13.33%). Remaining cases include inflammatory pseudotumor, osteoma, dermolipoma, schwannoma, neurofibroma, meningioma, rhabdomyosarcoma, glioma. The variety of tumor can involve the orbit, neoplastic lesion exceeded the non-neoplastic lesion.

Key words: intraorbital tumors , tumorlike conditions, pediatric patients.

INTRODUCTION

Tumors of the orbit are rare diseases in ophthalmic pathology. A spectrum of tumors and pseudotumors can involve the orbit of children[1]. They are great challenge for the ophthalmologist as they are often difficult to diagnose at the initial stages. Orbito-ocular tumors are unsightly and may contribute to visual disturbances. Although thought to be rare, they contribute to significant cause of morbidity and mortality[2,3,4]. Visual loss in children has implications for all aspects of the child's development[3]. Both benign and malignant masses of the orbit can have bone destruction[5], that's why it is very important to diagnose early for these intraorbital tumors in children. Orbital tumors include tumors of the lacrimal gland, fibrous tissue, adipose tissue, muscle tissue, bone and cartilage, xanthomatous and histiocytic tumors, tumors of blood vessels and lymph vessels, tumors of peripheral nerves and autonomic nervous system, optic nerve. It also includes metastasis from malignant tumors of contiguous anatomical areas such as nasal cavity, paranasal sinus, parotid glands, skin of the forehead & eyelid[6]. The histopathological characteristics of these tumors are critical to their biologic behaviour, line of management, outcome and prognosis[2]. Malignant tumors in children and adults are biologically and histologically distinct. Aims and objective of this study is to find out relative incidence of intraorbital tumors in relation with age group and sex in children, to study the pathological features of these tumors and to compare relevant data with other series of different authors from different areas of the world.

MATERIALS AND METHODS

A present study consists of 75 cases and has been carried out at M & J Institute of Ophthalmology, Histopathology Department, New Civil Hospital, Ahmedabad.

All cases with age less than seventeen years were included in this study.The clinical details were obtained from original case record like, age and sex of patients, signs and symptoms and other investigations like X-Ray, Ultrasonography, Computed tomography scan, Contrast Enhancing CT scan, Magnetic Resonance Imaging.

The gross examination of each available specimen includes its size, shape, weight, consistency and appearance of cut surface, especially in regard to the presence of hemorrhage, necrosis and cystic spaces etc. All the specimens were kept in 10% formalin for fixation.

In histopathological study, numbers of sections were taken from different sites according to size of specimen, while in case of small biopsy specimen, it was whole given.

Then these representative sections were subjected for processing and stained with hematoxylin and eosin using standard procedures. Special staining procedures like (PAS) Periodic Acid Schiff, Reticulin stain were done as and when required. All space occupying lesions of orbital cavity were included in the study.

The final diagnosis in each case was established by a combination of history, clinical examination, radiological imaging and histopathological examination.

RESULTS

A study of 75 cases of intraorbital tumors and tumor like conditions in children presented here with data of age wise, sex wise distribution, nature of neoplasm and various morphological types of neoplastic lesions are tabulated.

Table 1 Various types of intraorbital tumors among study participants

Individual tumor type	Cases (n=75) (%)
Retinoblastoma	46 (61.33)
Dermoid	10 (13.33)
Inflammatory pseudotumor	6 (8.0)
Osteoma	1 (1.3)
Dermolipoma	2 (2.66)
Schwannoma	2 (2.66)
Neurofibroma	3 (4.0)
Meningioma	1 (1.3)
Pilocytic astrocytoma	1 (1.3)
Rhabdomyosarcoma	3 (4.0)

Common symptoms and signs encountered were proptosis, vision impairment, pain and leukocoria (white pupil).

Leukocoria (white pupil) was observed in 57.33 % of the cases in this study. Proptosis observed in 49.33 % of cases in present study with 19 male and 18 female patients .

Table 1 shows that in total 75 cases , 49 cases of malignant tumors (65.33%), 26 cases of benign tumors (34.66%), and most common malignant tumor found to be retinoblastoma with 46 (61.33%) cases and the most common benign neoplastic lesion dermoid cyst with 10 cases (13.33%). After that come inflammatory pseudotumors with 8 cases. Inflammatory pseudotumor has been included in the series because it is invariably diagnosed clinically and by imaging studies as a tumor, from which it can only be distinguished on histologic examination. Remaining cases include osteoma, dermolipoma, rhabdomyosarcoma, meningioma and other neurogenic lesions including glioma, schwannoma, neurofibroma.

Table 2 Age wise distribution of Intraorbital tumor

Age(yr)	0-3(yr)	4-6(yr)	7-9(yr)	10-12(yr)	13-15(yr)	15-17(yr)	Total
Retinoblastoma	27	16	0	3	0	0	46
Dermoid	2	1	1	1	4	1	10
Inflammatory pseudotumor	1	0	0	2	1	2	6
Osteoma	1	0	0	0	0	0	1
Dermolipoma	0	1	0	1	0	0	2
Schwannoma	0	1	0	0	1	0	2
Neurofibroma	0	2	0	0	0	1	3
Meningioma	0	0	0	1	0	0	1
Glioma	0	0	0	1	0	0	1
Rhabdomyosarcoma	2	1	0	0	0	0	3
Total	33	22	1	9	6	4	75

Table 3 Sex wise distribution of intraorbital tumors

Tumor	**Male**	**Female**	**Total**
Retinoblastoma	26	20	46
Dermoid	4	6	10
Inflammatory pseudotumor	5	1	6
Osteoma	0	1	1
Dermolipoma	1	1	2
Schwannoma	1	1	2
Neurofibroma	1	2	3
Meningioma	0	1	1
Glioma	1	0	1
Rhabdomyosarcoma	2	1	3
Total	**41**	**34**	**75**

Table 2 shows the distribution of intraorbital tumors according to age, age range is 2 months to 17 years. Most of the tumors found below age of 6 years. Inflammatory pseudotumor occurs at any age. Rhabdomyosarcoma most common between age of 0 to 6 years. Most malignant cases were of retinoblastoma with age range of 2 months to 12 years. Maximum retinoblastoma cases noted between 0-6 years of age.

Table 3 shows sex wise distribution of intraorbital tumors. There were 41 males and 34 females in this study giving M:F ratio 1.2:1. It shows in total 46 cases of retinoblastoma, there is M:F ratio 1.3:1.

Table 4 shows that in total 42 cases of retinoblastoma, the tumor has bilateral involvement only in 4 cases. The incidence of unilateral occurrence of the tumor is high as compared to bilateral involvement. In bilaterality girls are affected more than boys.

Table 4: Distribution of 46 cases of Retinoblastoma according to laterality & sex

Gender	**Retinoblastoma affected eyes**		**Total**
	Unilateral	Bilateral	
Boy	25	1	26
Girl	17	3	20
Total	42	4	46

DISCUSSION

Orbital tumors and tumorlike lesions cover a wide range

of different types of tumors. The histopathological characteristics of these tumors are critical to their biologic behaviour, line of management, outcome and prognosis.

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,[5] and can lead to deformity and vision loss, that's why it is very important to diagnose early for these intraorbital tumors in children.

This series of 75 cases from our institute is based on histopathological examination of specimens and radiological and clinical correlation. We believe this study is fairly representative of the spectrum of orbital tumors in children worldwide encountered by ophthalmologist, radiologist and pathologist.

Results of the present series are compared with other series reviewed earlier. Variable incidence of primary orbital tumors has been reported by different workers in their series. Silva D reported that amongst the primary orbital tumors the pseudotumors followed by dermoids occurs most frequently[7]. Hemangioma was found to be the commonest by Ingalls and Reese. On the contrary meningioma was reported to be the commonest by Dandy and pleomorphic adenoma of lacrimal gland topped the list of primary orbital tumors in the studies of Forrest, Dass and Mohan et al. In the K Nath et al study pseudotumors (23.33%) and orbital dermoids (21.64%) were the commonest primary orbital new growths. These were followed by vascular (15.85), optic nerve (11.67%), mesenchymal (10.0%) peripheral nerve (9.16%) and epithelial tumors (8.33%). This included 103 cases of benign (85.83) and 17 cases of malignant orbital tumors (14.17%). Mohan et al have reported an incidence of 28% and 72% of benign and malignant tumors from a study of 68 orbital tumors. However, they did not include thirteen cases of pseudotumors in this category[8].

Table 5: Age and sex incidence of tumors of neural tissue pattern of neoplasia in children by Gracy Ramchandran et al[9] & Present study.

Histologic type	Gracy Ramchandran et al						Present Study					
	Sex			Age in years			Sex			Age in years		
	Total	Male	Female	0-5	6-10	11-14	Total	Male	Female	0-5	6-10	11-14
Neuroblastomas	12	7	5	6	4	2						
Gliomas	08	3	5	2	4	2	1	1			1	
Retinoblastomas	18	7	11	15	3	-	46	26	20	41	3	2
Diktyoma	01	1	-	1	-	-						
Neurofibroma	08	2	6	2	1	5	2	1	1	1	1	
Neurilemmoma	02	2	-	-	-	2	2	2			1	1
Neurofibrosarcoma	03	1	2	1	-	2						
Paraganglioma	02	2	-	-	-	2						
Total	54	25	29	27	12	15	51	30	21	6	6	3

Table 6:Analysis of Distribution of cases of retinoblastoma to laterality by Merrian G.R.Jr.[17] compared with present study.

Authority	Bilateral	Unilateral	Total	Percents (Bilateral)
Lawford and Collins	12	43	55	21.8
Marshall	12	20	32	37.5
Wintersteiner	97	308	405	23.9
Hirschberg	14	46	60	23.3
Vetschu	1	21	22	4.5
Devenport R.C.	5	22	27	18.5
Mc Urea W.B.E.	3	9	12	25.0
Berrisford	6	33	39	15.3
Owen	4	31	35	11.4
Merrian G.R.Jr.	56	15	72	77.7
Total	210	549	759	27.6
Present Study	4	42	46	8.69

Table 7 Active cases of retinoblastoma 1969 – 71 by laterality and sex by Susan S. Devasa18 compared with present study.

Retinoblastoma	Susan S. Devasa Study			Present Study		
	Total	Boys	Girls	Total	Boys	Girls
Unilateral	50	24	26	42	25	17
Bilateral	11	2	9	4	1	3

In the study by K Nath et al, primary orbital tumors are seen to be more common during the second (37 cases) and first (32 cases) decades and slightly less frequently in third fourth and fifth decades. Ingallas reported the first whereas Silva reported both second and third decades as the commonest age for the orbital tumors[8].

Comparing table 1 age distribution of 75 cases of present study with age distribution of 300 cases of D. Silva study, out of 300 cases 47.67% of cases were in first and second decade of life. In current study out of 46 cases of retinoblastoma, the children between 0-10 years of age are mostly affected. This compares favorably with the study of D. Silva and Gracy Ramchandran et al.[9] on a subject pattern of neoplasia in children in 1971-72 (table 6), and study by Chinda et al.[2] The most common sign was leukocoria (white pupil) which was observed in 57.33 % of the cases in current study while in Gianotti Antoneli et al study 68% and in study by Menon et al 72%[10]. Proptosis in 49.33 % of cases in present study and in Gianotti Antoneli et al study 75.9%[10]. In study by Mohan H et al, 42 pediatric patients out of 141 patients of different age groups presented with proptosis with highest contribution by the cases of retinoblastoma in pediatric patients[11].In current study both male and female presented with proptosis equally while proptosis was 1.4 times more common in males as compared to females in study by Nath K et al[8].

A high percentage of malignant ophthalmic tumors was observed in pediatric age group due to retinoblastoma,[12] which constituted 61.33 % of all intraorbital tumors in present series is comparable to the study by Oyin Olurin et al[13].

Table 5 shows present series observations are very favorably comparable to the Gracy Ramchandran et al study, particularly retinoblastoma and neurilemmoma cases.

One case of meningioma reported in the present series. It is reported in first decade of life. This incidence is contradictory to incidence in study by D. Silva as more number of cases reported in third, fourth, fifth and sixth decades of life.

Peripheral nerve tumors in the present series out of 75 cases, 5 cases are reported. This compares favorably with study of Daniel Silva where more number of cases of this tumor reported in first and second decades of life.

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[4,7] which is corroborated by the results of the present study. It is also comparable to the study by Soomro T et al where 9 out of 10 cases were in age group of 1-10 years.[12]

In present study dermoid is the most common benign tumor like study by Somnath Saha et al[14].Dermoid in present study falling more numbers in childhood, first decades of life compares favorably with study by chung et al[15].

Inflammatory pseudotumors can occur at any age. For retinoblastoma there appears to be no particular predilection for person of either sex[16]. Individual reports may show a predominance of cases in one or the other sex, as in current study predominance in male.

Gliomas categorized as juvenile pilocytic astrocytomas, they account for 4-6% of all brain tumors in children, with the median age of diagnosis 5-9 years,[4] corroborated with morphological pattern of glioma in present study.

Meningioma: the tumor occur more in females than males. Here one case reported in female patient. This compares favorably with incidence of tumor according to sex, reported in series by Daniel Silva[7] (13 cases out of 16 reported in female).

Rhabdomyosarcoma: Male to female ratio is 2:1 in the present study while it is 1:1 in series by Tahira Soomro et al.[12]

For retinoblastoma two eyes are involved about equally. The chief difference of opinion is concerned with the relation of unilateral to bilateral occurrence. This tumor is usually in unilateral occurrence, in present involvement 4 out of 46 cases with bilateral retinoblastoma. This compares favorably with the study of 759 cases reviewed by Merrian G. R. Jr.[17] in table 6. Bilateral tumors occur in patients with hereditary retinoblastoma and the age at diagnosis is younger[16]. The mean age for unilateral tumor is 3.6 years and 3.3 years for bilateral tumors in current study.

In his analysis the tumor was bilateral in 210 cases (27.6%) out of 759 cases. It also compares favorably with a study of 61 cases of retinoblastoma by Susan S. Devasa[18]. In his study the bilateral involvement is in 11 cases (18%) from total 61 cases in table 7.

The comparison of laterality of present study of 75 cases with Daniel Silva and Susan S. Devasa Study shows bilateral involvement is uncommon.

CONCLUSION

In conclusion, total 75 cases of intraorbital tumor and tumor like conditions in pediatric patients are studied. In the study 65.33% cases are of malignant tumors and 34.66% of benign tumors. Retinoblastoma is observed to be the most common malignant tumor (61.33%). Dermoid is the most common benign tumor. Followed by pseudotumors, neurofibroma, rhabdomyosarcoma, schwannoma, dermolipoma, meningioma, osteoma and pilocytic astrocytoma are also encountered.

The least common tumors are glioma, osteoma and meningioma. The most common presenting symptoms in all cases are proptosis, visual changes and pain and clinical finding leukocoria (white pupil).

We have come across only 4 cases of bilateral

retinoblastoma out of 46 cases, which is most commonly found, as bilaterality is not common in retinoblastoma.

In this study we have come across one case of glioma with morphological pattern of pilocytic astrocytoma. Maximum cases of retinoblastoma fall in 0 – 6 years of age. There is no sex predilection. Rhabdomyosarcoma is most common in childhood. For intraorbital tumor and tumor like conditions sex distribution do not vary widely, except meningioma which is common in females. In the present study except retinoblastoma all types of tumor and tumor like conditions are unilateral lesions.

In the case of orbital tumors, the relative rarity of certain types of tumors and the preponderance of other types offer invaluable opportunities for research into etiology, pathogenesis and geographical pattern of neoplasms in general.

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