RETINOBLASTOMA IN NWFP, PAKISTAN

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Background: Retinoblastoma is a malignant intraocular tumour of childhood, if not detected early, it results in loss of eye as well as life. The Objective was to study the pattern of presentation of Retinoblastoma in order to detect the disease at an earlier stage. It was conducted at Supra Regional Referral Centre for Retinoblastoma, Department of Ophthalmology, Khyber Teaching Hospital, Peshawar, Pakistan from 1st July 1999 to 30th September 2002. **Methods**: Seventy patients were registered and admitted for diagnostic evaluation. After initial interview a proforma was filled about history of disease. Examination under anaesthesia included indirect ophthalmoscopy with indentation. B-scan of the eye and CT scan of the Orbit was done when required. **Results**: Forty (57.14%) patients presented with Leukocoria; other presentation included hyphaema, strabismus, proptosis, 56.48% patients presented in stage VB (Rees Elsworth classification). **Conclusion:** Early detection through better understanding of presenting features of the disease and application of diagnostic skills can reduce the advanced stage of Retinoblastoma.

Keywords: Retinoblastoma, Leukocoria, strabismus, proptosis, B-scan, Rees Elsworth classification

INTRODUCTION

Retinoblastoma is the most common tumour of early childhood, both sight and life can be saved by early treatment. Epidemiological studies show the incidence in USA as 1 in 19,000, live births.[1,2] An increase in the incidence of Retinoblastoma was shown in Finland between 1912–1964, which might be due to improved diagnostic facilities during this period.[3]

The incidence of Retinoblastoma between 1968–1995 in Singapore was 11.10 per million for children less than 5 years of age.[4] Various studies reveal no significant difference between males and females.[4–6] In majority of studies, mean age at the time of presentation, which is 1–5 years in USA[7] and 2.52 years in Kenya.[8]

Common presentation of Retinoblastoma children is with leukocoria and strabismus. Less common presentations are hyphaema, glaucoma, endophthalmitis; proptosis although rare in developed world is still a frequent event in underdeveloped countries.[9,10] Worldwide studies show bilateral to unilateral ratio as 1:3. CT. Scan in majority of cases reveal calcification.[11,12] B-scan is helpful in most of the situations.[13]

The aim of the study was to deliberate on the profile and pattern of presentation in Retinoblastoma patients, because no definite statistical data and mode of appearance is available in Pakistan. It will help in setting guidelines for most effective and early treatment of these unfortunate children.

PATIENTS AND METHODS

The present study was carried out at Supra Regional Referral Centre for Retinoblastoma, Department of Ophthalmology, Khyber Teaching Hospital Peshawar Pakistan. Retinoblastoma patients are referred here from peripheral districts and Afghan Refugees' Hospitals as well as Afghanistan for diagnosis and treatment. Total number of patients, registered in the centre since 1st July 1999 to 30th September 2002, is 70. Total number of eyes included in the study was 108. The tumours in these eyes were classified according to Rees Elesworth system for Retinoblastoma classification.

All data was collected prospectively. Facial photographs, CT Scan, MRI and Ultrasonography were duly documented, after admission in the Eye Department. For each patient, age, gender, age at diagnosis, mode of presentation in terms of leukocoria, or squint was recorded on a printed detailed proforma, which is meant for registering the patient, collecting data and recording treatment plan.

After initial interview with the parents and baseline investigations, including complete blood count, ESR and serum LDH, a comprehensive examination was performed within 48 hours under general anaesthesia in the fully equipped operation theatre of Ophthalmology department.

Intraocular pressure was recorded (by Schiotz tonometer). Horizontal corneal diameter was measure with callipers. Condition of anterior segment, including cornea, iris for rubeosis and lens for presence of opacity was assessed under binocular microscope.

Indirect ophalmoscopy was done using indirect Binocular ophthalmoscope of OMEGA-100 of HEINE OPTOTECHNIK Germany) in a fully dilated pupil while using 1% atropine, or tropacanamide topical eye drops for this purpose.

Each eye was examined for location of tumours in relation to optic disc and number of tumours, size of tumours in disc diameters, vitreous seeding, subretinal deposits, and extent of detachment. Indentation method was used to see the peripheral retina. These findings were recorded diagrammatically in each case.

B-Scan was performed when fundus could not be seen by indirect ophthalmology CT Scan orbit and brain was carried out in each patient to confirm calcification and rule out intracranial extension of tumour.

RESULTS

Since July 1999 to 30th September 2002, newly diagnosed 70 retinoblastoma cases were registered for the study.

The regional distribution of Retinoblastoma cases in Retinoblastoma Registry is presented in Table-1. An average annual admission of 23 patients in the Retinoblastoma Supra Regional Centre was recorded.

Of 70 cases 65 (92.8%) were diagnosed before the patients were 5 years of age out of which 20 (30.7%) were diagnosed before one year of age in less than 5 years group. There was no difference in occurrence rate of Retinoblastoma by sex in either age group. The percentage of patients with Afghan origin was noted to be 17%. There was no significant difference in the incidence of Retinoblastoma among referring regional areas, within NWFP, Pakistan.

Table-2 gives distribution of unilateral and bilateral Retinoblastoma according sex and age. Two thirds of the cases are bilateral. Unilateral cases are more common in males as compared to female. Bilateral cases were more in less than one year of age and unilateral cases were higher in 2–3 years of age.

Table-3 shows presentation pattern of Retinoblastoma in patient during 1999–2002. Forty (57.10%) of cases presented as leukocoria and 10 as fungating mass or proptosis (Photograph-1 and 2). Eight cases presented as squint, followed by glaucoma (Buphthalmos) and hyphaema. One case was diagnosed as part of familial screening. CT scan revealed calcification in 99% cases and was a confirmatory evidence of Retinoblastoma (Photograph-3). Table-4 shows distribution of various stages amongst Retinoblastoma patients. 61 (56.48%) eyes (for 108 eyes) presented in stage VB (Rees-Elsworth Classification).

Table-1: Regional Distribution of Retinoblastoma cases in children aged 0–12 years by Retinoblastoma Registry July 1999 through September 2002

District of Residence	Cases	%
Afghan origin	12	17.14%
Malakand and Bajawar	16	22.86%
Peshawar	11	15.71%
Southern region (Bannu, DI Khan)	8	11.43%
Mardan	9	12.86%
Kohat	6	8.57%
Kurram Agency	2	2.86%
Khyber Agency	3	4.29%
Chitral	1	1.43%
Haripur	1	1.43%
Karachi	1	1.43%

Table-2: Distribution of unilateral and bilateral Retinoblastoma in children aged 0–12 years by age and sex during July 1999 to September 2002

A: Based on sex distribution

Gender	Unilateral	Bilateral
Male	21	20
Female	14	15
Total	**35**	**35**

B: Based on age distribution

Age (in years)	Unilateral	Bilateral
0–1	5	13
1–2	6	8
2–3	11	5
3–4	6	6
5–12	7	3
Total	**35**	**35**

Table-3: Presenting features in Retinoblastoma patient, during 1999 through September 2002

Presenting feature	No. of cases	%
Leukocoria	40	57.1
Proptosis	10	14.3
Squint	8	11.4
Glaucoma/Buphthalmos	6	86
Hyphaema	4	5.7
Epiphora	2	2.95
Total	**70**	

Table-4: Staging of the Retinoblastoma at presentation according to Rees-Ellsworth classification for 108 eyes (70 patients)

Stage	No. of cases	%
IA	7	6.48
IB	2	1.85
IIA	3	2.78
IIB	3	2.78
IIIA	3	2.78
IIIB	1	0.93
IVA	2	1.85
IVB	5	4.63
VA	9	8.33
VB	61	56.48
METASTATIC	12	11.11
Total Eyes	**108**	

DISCUSSION

Retinoblastoma, in terms of its profiles and presentation, carry diverse clinical features. In this part of the world, the data in this regard is deficient because of lack of advanced facilities and expertise.

The aim of our present study is to project he gravity of situation in relation to late presentation and misdiagnosis of in retinoblastoma patients.

Our referral centres which received record number of cases of retinoblastoma in this region, helped in addressing the lacunae in management of these unlucky children.

According to our study the mean age at presentation is 28.17 months. Unilateral cases presented at mean age of 31.81 months.

Leukocoria was the most common presentation, which occurred in 40 (57.14%) patients.

Proptosis was noted in 10 (14.3%) patients and squint in 8 (11.4%) patients.

These figures are higher than the study done by Abramson, where the median age of presentation is 25 months for unilateral cases and 15 months for bilateral cases.[14]

In a multicentre study by Mexican retinoblastoma group, the age range was found to be 0–182 months. This study also emphasised the importance of referral centres to treat retinoblastoma patients.[15]

In study of 105 patients carried out in Turkey by Ozkan A *et al* the mean age at diagnosis was 25 months. The most common presenting sign has been leukocoria occurring in 82% of cases, while leukocoria was the presentation in 57.14% of cases in our study.[16]

The advanced age in our study is due to late presentation misdiagnosis and extreme poverty.

Majority of patients in our study presented in advanced stage of VB (Rees Elsworth classification). This is a major cause of high morbidity and mortality in these patients.

A study by Mullaney PB *et al* done in Saudi Arabia revealed 49.4% of retinoblastoma cases to be presenting in advanced stage VB disease. This study also supports our point of view that early diagnosis through establishment of referral centres and provision of expert facilities can increase survival in these children.[17]

In recent past, early detection in Europe and USA, with positive family history could save some useful vision in retinoblastoma patients. This strategy can prevent enucleation in 30% of unilateral cases.

At the start of the century, long term survival of Retinoblastoma patients was 25%. Now-a-days this rate has increased to more than 90% in developed countries.[18,19]

CONCLUSION

Understanding the common features of Retinoblastoma and timely diagnosis through diagnostic acumen and use of advanced equipment can save the sight as well as life of the child.

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