

Frequency of Retinoblastoma According to Age, Gender and Laterality

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ABSTRACT

Objective: To evaluate the frequency, age, gender and laterality related distribution of retinoblastoma.

Study design: Cross sectional study.

Place and Duration of study: This study was conducted at the Department of pathology BMSI, JPMC Karachi from 1st Jan 2009 to 31st Dec 2013.

Materials and Methods: A total of 80 cases of retinoblastoma were received in the department of Pathology, BMSI, JPMC, Karachi during the period of 1st January 2009 to 31st December 2013. The cases were reviewed and morphological diagnosis done on H&E. Information regarding laterality of lesion, age and sex were recorded from archives. The data analyzed by using SPSS version 22.

Results: Frequency of retinoblastoma was 2.93%. Right sided lesions were 46.25% & 45% were Left sided, with a M: F ratio of 1:1. The mean age of retinoblastoma patients was 3.64 years. It was relatively more common in 3 to 4 years (53.75%) of age group compared to other age groups.

Conclusion: Frequency of retinoblastoma was 2.93%, with almost equal i-e 46.25% Right, 45% Left sided origins. The M: F ratio was 1:1 while more common age group was 3 to 4 years.

Key Words; Retinoblastoma. Frequency, Age, Gender, Laterality.

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INTRODUCTION

Retinoblastoma is a neuroectodermal embryonic tumor of developing retina, caused by mutation in RB1 tumor suppressor gene, presented at long arm of chromosome 13 q 14¹⁻⁶. Crucial period for mutations in retinal cells are retinal development between 4th to 8th gestational week and continuous up to six month of infancy¹. Approximately 10% of affected children with retinoblastoma have positive family history⁷.

Retinoblastoma is seen as both, hereditary as well as in non hereditary form. The former has increased risk of subsequent malignancies i.e. bone & soft tissue sarcoma, melanoma and brain tumors^{3,4}. Retinoblastoma is usually recognized under 3 years of age while majority of cases are diagnosed before 5 years of age⁸.

Globally 5,000 new cases of retinoblastoma are diagnosed annually. Incidence ranges vary from 1 in 15,000 to 1 in 20,000 live births⁹⁻¹⁰. Frequencies of retinoblastoma have been reported varying as 2 to 4%^{9,11} 3%^{12,13} and 6%¹⁴ respectively amongst all childhood malignant neoplasms.

In USA approximately 300 children are affected by retinoblastoma annually¹⁵. African countries showed higher incidence accounting for 10-15% of childhood malignancies whereas Australia, North America,

Germany, UK and Asian subcontinent accounts for 1.5 to 4% of neoplasms in children^{16,17}. Idaho pediatric cancer registry¹⁸ data showed retinoblastoma being the 5th cancer type at the rate of 2.5%. Surveillance epidemiology and End Results (SEER) USA registry¹⁹ showed that retinoblastoma is the 5th cancer type, at the rate of 3.2%. Shaukat Khanum collective cancer registry²⁰ data showed that retinoblastoma accounts the 6th most common malignancy with 5.09% frequency.

Environmental factors like poverty and low maternal education are associated with increased risk whereas multivitamins supplementation have protective role for retinoblastoma development¹.

This study was designed to assess the frequency of retinoblastoma in our population and to evaluate it with laterality of the lesion, age and sex.

MATERIALS AND METHODS

The study was performed at the department of Pathology Basic Medical Sciences Institute, Jinnah Postgraduate Medical Center Karachi. A total of 80 cases of retinoblastoma were received from 1st January 2009 to 31st December 2013. These patients were operated at Ophthalmology department of JPMC Karachi. All enucleated eye specimens were included, whereas poorly fixed & inadequate tissue, ocular tumor other than retinoblastoma and metastatic tumors were excluded. Formalin fixed, paraffin embedded blocks, surgical pathology & clinical records and Hematoxyline & Eosin slides were used. The relevant clinical

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information and data were collected. Sections were taken and stained with H&E. All slides were studied under light microscope using scanner (4x), low power (10x) followed by high power (40x). The data was analyzed by using Statistical Package for Social Sciences (SPSS) version 22.

RESULTS

A total number of 80 cases were received in the department of Pathology, BMSI, JPMC, Karachi over a period of 5 years. The data showed that total malignancies from all sites were 2726 out of which 80 cases were reported as retinoblastoma. Frequency of retinoblastoma was 2.93% over a period of five years.

Table No.1: Frequency of Retinoblastoma (n=80)

Years	Total no malignancy of all sites	Retino-blastoma	%age	95% C.I.
2009	590	17		2.34-3.62
2010	648	26		
2011	550	19		
2012	510	14		
2013	428	04		
Total	2726	80	2.93	

CI: Confidence interval

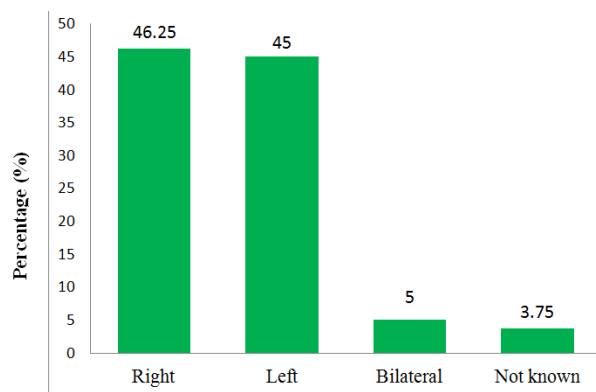


Figure No.1: Distribution of retinoblastoma according to laterality (n=80)

Figure 1 elaborates the laterality. Out of 80 cases, 37 [46.25%] cases were seen in right eye, 36 [45%] cases were seen in left eye, 04 [05%] cases were bilateral and in 03 [03.75%] cases laterality was unknown respectively.

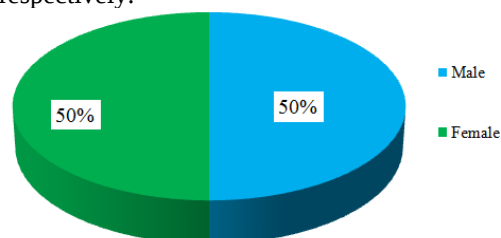


Figure No.2: Distribution of retinoblastoma according to gender (n=80)

Figure 2; shows distribution of retinoblastoma according to gender. Out of 80 cases 40 [50%] cases were seen in each gender. Thus male: female ratio of 1:1.

Table No.2: Distribution of retinoblastoma according to age (n=80)

Age (years)	No of cases	Percentage %	Cumulative index
<1 year	04	05	05
1-2 years	11	13.75	18.75
3-4 years	43	53.75	72.5
5-6 years	16	20	92.5
7-8 years	02	02.50	95
9-10 years	01	01.25	96.25
Unknown	03	03.75	100
Total	80	100	

Table 2; demonstrates distribution of retinoblastoma according to age. Amongst 80 cases, majority i.e 53.75% cases were found between ages 3-4 years followed by 20% cases in 5-6 years. The mean $\pm$ SD were 3.64 years (42.25 months) $\pm$ 1.74, median age was 4 years (48 months). The minimum age was 02 months while maximum age noted was 09 years.

DISCUSSION

In the present study the frequency of retinoblastoma was 2.93 %. Our findings are comparable to the figures documented in the studies by Antoneli et al¹², Yeole and Advani¹⁶, Jijelava and Grossinklans et al¹¹ and Li et al⁹ reporting the frequency as 3%, 2 to 4%, 2.4% and 4% respectively.

Regarding laterality this study shows that nearly equal distribution i.e 46.25 % and 45% of retinoblastoma were seen in right and left eye respectively. These finding are comparable with the Indian study by Yeole and Advani¹⁶ who reported 41.3 % and 40% retinoblastoma originated from right and left eye respectively. In this study 91.25% cases were unilateral and 05% cases were bilateral, these figure are comparable with the Al Shifa Trust Eye Hospital study by Islam et al¹⁰, Akhiwu and Igbe¹⁷ and Chintagumpala et al¹⁵. They reported 72.7%, 82%, 70 to 80% unilateral and 27.3%, 18% and 29 to 30% bilateral cases respectively. The record on laterality was missing in some cases which explains the variation seen in the present study.

In this study male to female ratio was 1:1, which is similar to the studies by Akhiwu and Igbe¹⁷, Rodrigues et al²¹ and Arif et al²² reporting 1.1:1, 1.3:1 and 1.1:1 respectively. An Indian study by Yeole and Advani¹⁶ reported 4:3.1 showing male predominance. This discrepancy could be due to the cultural difference, where only males are brought preferentially to medical attention than females.

The most common age group in our study was 3 to 4 years in 53.75% cases followed by 20% cases in 5 to 6 years age group. These findings are comparable to the figures documented in Edo State of Nigerian Study by Akhiwu and Igbe¹⁷ reporting 3 to 3.5 years and Tata Memorial Hospital Mumbai India study by Yeole and Advani¹⁶ who reported 76.5% to 78 % cases under 4 years of age, but is dissimilar to the studies by Rodrigues et al.²¹, Chintagumpala et al¹⁵ who reported 53% cases under 2 year and 80% cases under 3 year respectively. This discrepancy may be due to poverty, lack of awareness, alternative treatment such as Hakeem's, leading to late presentation in our population.

In this study the mean age was 3.64 years (42.25 months). This finding is in total agreement with the work by Akhiwu and Igbe¹⁷, reporting the mean age ranging from 2 to 4 years (24 to 48 months). But is dissimilar to Arif et al²² and Antoneli et al¹² who showed 32 months and 28.7 months respectively. The reason for this could be late presentation in our population or could be due to environmental and genetic differences.

CONCLUSION

Majority of cases were seen in later age groups i.e 3 to 4 years with high histological grades, so wide scale awareness through education to the community, parents, physicians and counseling program may help to ensure early presentation in initial stage of the disease, in order to improve clinical outcomes with less morbidity and mortality.

True frequency of retinoblastoma in our community may be higher than reported, but due to lack of good health services and weakness in referral system, it is impossible to know the true scenario.

Further studies based on larger sample size, with complete clinical and radiological examination with long term follow up is recommended.

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Conflict of Interest: The study has no conflict of interest to declare by any author.

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