Original Article

Retinoblastoma

Frequency of Retinoblastoma According to Age, Gender and Laterality

1. Syed Muhammad Ishaque 2. Shahnaz Imdad Kehar 3. Saba Sattar

1. Demonstrator of Pathology, BMC, Quetta 2. Assoc. Prof. of Pathology, BMSI, JPMC, Karachi 3. Lecturer of Pathology, SMC, Karachi

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.

Citation of article: Ishaque SM, Kehar SI, Sattar S, Frequency of Retinoblastoma According to Age, Gender and Laterality. Med Forum 2015;26(10):20-23.

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 14[1-6]. Crucial period for mutations in retinal cells are retinal development between 4th to 8th gestational week and continuous up to six months of infancy[1]. Approximately 10% of affected children with retinoblastoma have positive family history[7]. 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 age[8].

Globally 5,000 new cases of retinoblastoma are diagnosed annually. Incidence ranges vary from 1 in 15,000 to I in 20,000 live births[9-10]. 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 annually[15]. 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 registry[18] data showed retinoblastoma being the 5th cancer type at the rate of 2.5%. Surveillance epidemiology and End Results (SEER) USA registry[19] showed that retinoblastoma is the 5th cancer type, at the rate of 3.2%. Shaukat Khanum collective cancer registry[20] 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 development[1].

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.

Correspondence: Dr. Syed Muhammad Ishaque,
Demonstrator of Pathology Bolan Medical College Quetta
Contact No.: 03003801784
E-mail: ilyas2927@gmail.com

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

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		
2010	648	26		
2011	550	19		2.34-3.62
2012	510	14		
2013	428	04		
Total	2726	80	2.93	

CI: Confidence interval

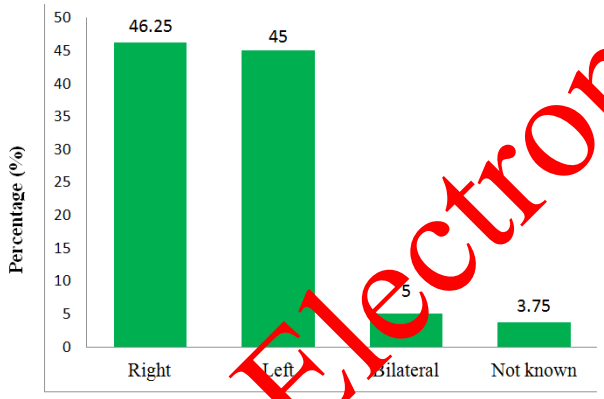

Figure No.1: Distribution of retinoblastoma according to laterality (n=80)

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.

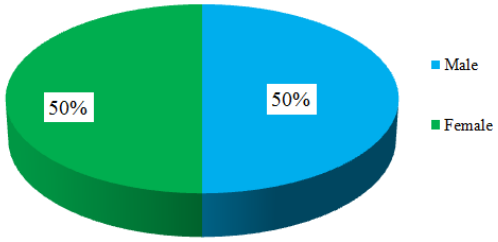

Figure No.2: Distribution of retinoblastoma according to gender (n=80)

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 ± SD were 3.64 years (43.25 months) ±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 al[12], Yeole and Advani[16], Jijelava and Grossinklans et al[11] and Li et al[9] 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 Advani[16] 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 al[10], Akhiwu and Igbe[17] and Chintagumpala et al[15]. 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 Igbe[17], Rodrigues et al[21] and Arif et al[22] reporting 1.1:1, 1.3:1 and 1.1:1 respectively. An Indian study by Yeole and Advani[16] 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 finding are comparable to the figures documented in Edo State of Nigerian Study by Akhiwu and Igbe[17] reporting 3 to 3.5 years and Tata Memorial Hospital Mumbai India study by Yeole and Advani[16] 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 al[15] 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 Igbe[17], reporting the mean age ranging from 2 to 4 years (24 to 48 months). But is dissimilar to Arif et al[22] and Antoneli et al[12] 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 out comes 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.

Acknowledgement: We are thankful to Dr. Shahid Zafar, Dept of Pathology, SMSI, JPMC Karachi for data collection.

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

REFERENCES

1. Orjuela MA, Titievsky L, Lui X, Ortiz MR, Castaneda VP, Lecona E, et al. Fruit and vegetable intake during pregnancy and risk for development of sporadic retinoblastoma. Cancer epidemiology, biomarkers Prev 20005; 14(6):1433-1440.
2. Oweye JFA, Afolayan EAO, Popoola DSA. Retinoblastoma-a clinicopathological study in Ilorin, Nigeria. Afr J Health Sci 2005;12:94-100.
3. Kleinerman RA, Tucker MA, Tarone RE, Abramson DH, Seddon JM, Stovall M, Li F, Pand JFF. Risk of new cancer after radiotherapy in long-term survivors of retinoblastoma; an expected follow-up. J Clin Onco 2005;23(10):2227-79.
4. Kleinerman RA, Schonfeld SJ, Tucker MA. Sarcomas in hereditary retinoblastoma. Available at http// www clinical sarcoma research. Com J Clin Sarc 2012;2-15.
5. Wu–D, Li Y, Song G, Zhang D, Shaw N, Liu ZJ. Crystal structure of human esterase D; a potential genetic marker of retinoblastoma. FASEB J 2009; 23:1-7.
6. Rosai J, Ackerman. Surgical pathology, Elsevier New Delhi India. 10th ed. Eye and ocular adnexae 2011;2;2490-93.
7. Diciommo D, Gallie Bl, Bremner R. Retinoblastoma: The disease, gene and protein provide critical lead to under stand cancer. Available at http// www. Ideal library, Com. J can bio .2000; 10:253-69.
8. Girardet A, Hamamah S, Anahory T, Dechaud H, Sarda P, Hedon B, et al. M First preimplantation genetic diagnosis of hereditary retinoblastoma using informative microsatellite markers. Available at http;//molehr.ox/ford journal.org, Mole Hum Rep J.2003; 9:111-16.
9. Li Z, Wu X, Li J, Yao L, Sun L, Shi Y, Zhang W, et al. Antitumor activity of celastrol nanoparticales in a xanograft retinoblastoma tumor model. Int J Nano Med 2012; 7:2389-98.
10. Islam F, Zafer SN, Siddiqui SN, Khan A. Clinical Course of Retinoblastoma. JCPSP 2013; 23(8): 566-69.
11. Jijelava KP, Grossniklans HE. Diffuse anterior retinoblastoma. A review, Available at www, Saudi Ophthal journal com. Saudi. J Ophth 2013; 27: 135-39.
12. Antoneli CBG, Steinhorst F, Ribeiro K de CB, Novaes PER, Chonjniak MMM, Arias V, et al. Extraocular Retinoblastoma; A 13-Year Experience. American Cancer Society 2003; 1292-98.
13. Jurkiewicz E, Rutynowska O and Perek D. Trilateral retinoblastoma: Diagnosis using magnetic Resonance Imaging. J Pedi Canc 2012; 3:185-92.
14. Jr BW, Schefler AC. Second malignancies in retinoblastoma; the real problem. Available at www.Intechopen.com 2014; 23-38.
15. Chintagumpala M, Barrios PC, Paysse EA, Plon SE, Hurwitz R. Retinoblastoma: Review of current management. Available at www. The oncologist com. J oncolog.2007; 12:1237-46.

16. Yeole BB, Advani S, Retinoblastoma: An epidemiological appraisal with reference to a population in Mumbai India, Bombay cancer registry Tata memorial Hosp Mumbai. Asian Pcif J Can Preven 2002;3:17-21.
17. Akhiwu WO, Igbe AP. Epidemiology of retinoblastoma. J Pedi Ophth Strab 2009;46: 288-93.
18. Idaho Pediatric Cancer incidence (1994-2004), Idaho Hosp Assoc Cancer data Registry of Idaho.
19. SEER, Stat fact sheet; 1994-2004, National Institute of Cancer, 1994-2004.
20. Shaukat Khanum Memorial Cancer Hospital Registry collective. Cancer Registry Report (1994-2011). Available at www. Shaukat Khanum. Org. PK/images /Skming / down loads/ Pdf/Ccrr-1994-2011.
21. Rodrigues KES, Latorre MDRO, de Camargo B. Delayed diagnosis in retinoblastoma. J De Pedi 2004; 80(6):511-16.
22. Arif M, Iqbal Z, Islam- Z-U. Retinoblastoma in NWFP (KPK), Pakistan .J Ayub Med Coll Abbottabad 2009; 21(4):60-62.

.