



RESEARCH ARTICLE

Retinoblastoma – A Retrospective analysis

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ABSTRACT:

Background: Primarily retinoblastoma occurs in children under the age of 5 years. The incidence of unilateral Retinoblastoma is higher in comparison to bilateral inherent. Some of the environmental factors and living conditions may be responsible for mutagenesis in the retinal cells. **Material and method:** All the cases who were attended radiation oncology department of IGIMS between Jan2000- Jan 2014 included in the analysis and analyzed according to their age, sex, laterality and type of presentation. **Results:** Most common age group was involved in our study 3-4 years; incidence of unilateral involvement was higher in our study. Male female ratio was 3.2:1. **Discussion:** Retinoblastoma is the most common childhood cancer of Retina. It occurs in two from hereditary and non hereditary. In our study male cases were 76.4% and female cases were 23.5%. Male were more in our study as compare to other studies. **Conclusion:** This retrospective study shows the distribution of retinoblastoma according to the age, sex, laterality and presentation.

Keywords: Retinoblastoma, Age, sex, laterality

INTRODUCTION

Retinoblastoma is the most common intraocular malignancy in children in India (Gogi & Nath¹, Dhir et al²). Studies from India show a two to three fold higher incidence of tumors of the eye majority of which will be retinoblastoma in children <15 years of age. Abra Rs³

It contributes almost 4% of all pediatric malignancies and each year approximately 250-300 children are newly diagnosed in the United States and substantially higher rates occur in developing countries^{4,5}Shield leal⁴. It is reported to affect 1 in 15000 to 1 in 18000 live births. Abra³

Retinoblastoma accounted for 2-4% of cancer in children in Europe, North America and Australia (parkin et al 1988)⁶. The relative frequency is similar in Asia (Ajikki et al 1994⁷ yeole el al 2001⁸).

In India it is estimated to be 4.4 times more common than that of United States. In developed countries extra ocular retinoblastoma is very rare (2-5%). In developing nations, extra ocular disease contributes 50% of all retinoblastoma cases presenting to a tertiary care referral centre Bakshi & Bakshi⁹. Survival rate is only 50% worldwide while over 95% patients with Retinoblastoma survived in developed countries (leal-lead⁵).



MATERIAL AND METHOD-

A retrospective analysis has been done at IGIMS, Patna. The study period included from Jan2000- Jan 2014. Data collected on retinoblastoma by sex, age religion and laterality. Total 51 patients were analyzed in this study.

Result:

	Surgery Done	Not operable	Total
Male	15 (38.4%)	24(61%)	39
Female	3 (25%)	9(75%)	12

Sex & Age :

Most common age group was 3 -4 years of age in both sexes.

In total 51 patients 39 patients (76.4%) were male and 12 patients (23.5%) were female.

AGE Group

Age group	Total No of Patients	Male(39)	Female(12)
0-2 years	11	9	2
3- 4 years	27	20	6
5- 7 years	13	7	4

Laterality

Out of 51 cases 37 were unilateral and 14 were bilateral. In unilateral cases right eye was affected in 22 cases. In bilateral disease started from right eye in 7 cases and in 2 cases, disease started from left eye.

Laterality	Total No of Patients	Male(39)	Female(12)
unilateral	37	29	8
Bilateral	14	10	4
Total	51	39	12

Presentation— Most of cases presented in advanced stage 56% in male and 66% in female.

Presentation	Total no of patient	Male	Female
Watering of eye	1	1	0
White pupillary reflex	6	5	1
Proptosis	16	13	3
Advance stage	26(56.9%)	18(46%)	8(66%)



Site of Involvement:

Total no patient	Right Eye	Left Eye
51	30(58%)	21(42%)

Most of the patient presented with involvement of right eye (58%).

Discussion-

Pawius described retinoblastoma as early as 1597⁵.lead In 1809. Flexner (1891) and Wintersteiner (1897) believed it to be a neuro epithelioma because of the presence of rosettes.⁵ Lead, there was a consensus that the tumor originated from the retinoblastoma and the American Ophthalmological Society accepted officially the term retinoblastoma in 1926.^{4shield}

More than 50% cases presented in advanced stage. Approximately 60-75% cases were inoperable in both sexes only one fourth of cases were operable.

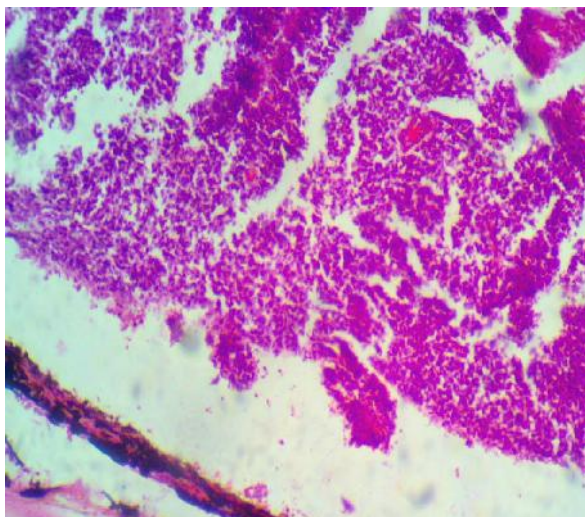


Figure 1: Section shows sheets of small round cells with hyperchromatic nuclei with scanty cytoplasm. Tumour cells have tendency to arrange around feeding vessels.-- Retionoblatoma

Retinoblastoma is the most common childhood intra ocular malignant neoplasm that arises in the retina (knudson¹⁰, 1971). It occurs in two form hereditary and nonhereditary forms, the hereditary forms transmit as an Autosomal dominant trail (Hogg et al 1992¹¹). The gene associated with the disease called as a tumor suppresser gene & retinoblastoma susceptibility gene RBI on chromosome 13 g 14 (Taumchida et al 1993)¹².

Total incidence is generally higher in developing countries especially in sub Saharan Africa, Black children have higher incidence than whites in the US. Incidence of retinoblastoma varies with the other regions of world and ethnic groups. These variations therefore presumably result from environmental factors operating after conception rather than from parental germ line mutation. Retinoblastoma incidence has been correlated with ultraviolet radiation in International data (Hooper 1999¹³), but the association was not



correlated after adjusting for ethnic group and tropical climate (Jamal et al)¹⁴. Bunin et al (1989)¹⁵ reported an inverse association between maternal use of multi vitamins during pregnancy and retinoblastoma. There was no association found with use of tobacco or alcohol intake during pregnancy.

Study conducted in African countries in which no bilateral case reported cobafunwe et al¹⁶ 1992 Tigani et al¹⁷ 1995. Study conducted at AIIMS the median age of presentation was 2.5 year. 69% of cases were unilateral and 30.5% of cases were bilateral. In this study there was male predominance and male female ratio was 1.6:1. In our study unilateral presentation was 72.5% and bilateral presentation was 28%. It was compared with study conducted at AIIMS. Study conducted in Oman the male female ratio was 0:7 (khandekan et al 2004¹⁸)

In Britain less than 2% cases were bilateral in Asian population (shltii et al 1991¹⁹).

Majority of the patients were presented in advanced stage more than due to illiteracy, poor socioeconomic condition, ignorance about disease, lack of health specialized services at rural areas.

Right eye involvement was more common (58 %) as compare to left eye (42 %).

In our study 39(76.4%) cases were males and 12 cases (23.5%) were female study conducted by B yoele & adnenis SH²⁰ showed 57.1% males and 42.9% female.

The average age of presentation was 2 years in our study while Rees has reported 13 years.

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Same pattern has been reported in earlier studies in India. (Yoele et al 2002²⁰, swaminathan et al 2008²¹ Hazarika et al 2014²²) and in developed countries (maccarthy et al 2009 mitra et al 2012)

Male female India reported unity in some of studies (parkin et al 1998⁶ mrc carthy et al 2009²³, mitra et al 2012²⁴). In India male female ratio has been reported 1.0 (swaminathan et al 2008²¹) and Mumbai 1.4 (yoele et al 2002²⁰). Most of the children presented in our study were in advanced stage.

In our study indicates 3.1% of retinoblastoma; Incidence of Retinoblastoma in pediatric cancer patient is higher (parkin et al 1988⁶, Mukibi et al 1995²⁵, weasels and Hesseling 1996²⁶). The male female ratio of Retinoblastoma in our study was 3.2:1. Showing higher proportion for males possibly due to higher proportion of boys in childhood population and in rural areas, female population is ignored by society, parents do not want to spend money for girl child. Maximum patients were in the age group of 3-4 years in both sexes. Youngest patient was of 18 months and oldest patient was 7 years of age.

The incidence of retinoblastoma varies according to socioeconomic status and between developed and developing countries (stiller & partend 1996²⁷). The rate of Retinoblastoma in the United States is 10.9 per million in children under six year of age (tambali et al 1990²⁸).

In comparison to non hereditary form of Retinoblastoma patient having hereditary form of retinoblastoma have higher risk to develop second primary (drapper et al 1985²⁹ derkinderson et al 1998³¹ eng et al 1993³²). Some studies shown most of the primary tumor are osteosarcoma and soft tissue sarcoma in short follow-up, melanoma in longer follow up. (moll et al 1996³²)



CONCLUSION

Objective of this study was to highlights the common age, laterality, presentation sex distribution of retinoblastoma. Genetic predisposition which was not assessed due to retrospective analysis. Prospective studies are required to assess the exact burden and genetic predisposition of disease in different parts of countries.

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Dr. Seema Devi, MBBS, MD (Radiation Oncology)

Biography

Dr. Seema Devi served 16 years in the field of Radiation Oncology; she started her journey from S.N. Medical College, Agra as a resident in the year 1997. Then after completing M.D in the year 2000 she moved to Safdarjang Hospital, New Delhi and worked there in the capacity of senior resident till 2002. Dr. Seema joined Department of Radio therapy, J.N. Medical College, Aligarh as an Assistant Professor in the year October 2002. She served there relentlessly for more than 12 years in the field of Radiation Oncology. There she was involved in the treatment of gastrointestinal cancers, breast cancer, sarcoma, and general radiation oncology and brain and central nervous system cancers. Dr. Seema now is working as an Associate Professor in Oncology Department of Indira Gandhi institute of Medical Sciences, Patna since October 2014, dealing with different type of radiation therapy techniques including accelerated partial breast irradiation and other protocol options.

Dr. Seema Devi Patient Care Philosophy

“I believe in individualized care and making decisions with my patients as a partnership.”