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Characteristics of retinoblastoma in patients of Jammu

[1]Dr. Avantika Kailu, Post Graduate, Department of Pathology, ASCOMS, Jammu.

[2]Dr. Kanavdeep Kapoor, Post Graduate, Department of Ophthalmology, ASCOMS, Jammu.

[3]Dr. Anisha Kapoor, Undergraduate, MBBS, ASCOMS Jammu.

[4]Dr. Dinesh Malhotra, Professor and HOD, Department of Opthalmology, ASCOMS, Jammu.

Corresponding Author: Dr. Kanavdeep Kapoor, Post Graduate, Department of Ophthalmology, ASCOMS, Jammu.

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Abstract

The incidence of retinoblastoma is increasing among children. Retinoblastoma is the most common intraocular malignant tumor affecting pediatric age group. The retinoblastoma develops after random gene (RB1 gene) mutation in retinal cell (photo receptor cells). The inheritance increases the risk of retinoblastoma. Literature suggested that the retinoblastoma is having good prognosis if it is detected at early stages. This prospective observational study was conducted in the Department of Ophthalmology at Acharya Shri Chander College of Medical Sciences and Hospital, Jammu, during the period of 2 years with the aim to determine the characteristics of retinoblastoma. A total of 100 eyes of 84 patients with retinoblastoma were involved in the study. The mean age of the patients at diagnosis was 22.8±15.68 months, females patients were affected mostly (60. 71%) with female male ratio 1.54:1, majority (80. 95%) of the patients had unilateral retinoblastoma and 5.95% patients had family history of retinoblastoma. Leukocoria as the most common clinical manifestation and most common mode of the treatment was enucleation. The study concluded that the leucocoria was the most common clinical feature seen in retinoblastoma patients. Undifferentiated Type was the most common Pathological Variant Seen on Histo pathological Examination.

It concluded majority of Retinoblastomas expressed RB as the most common marker on Immuno Histochemistry.

Keywords: Retinoblastoma, Children, Enucleation, Histopathology, Clinical Manifestations, Eye cancer, Tumor and Childhood cancer.

Introduction

Retinoblastoma is the intraocular malignant tumor generally affects children. The incidence of retinoblastoma is increasing among paediatric age group,

and it is estimated that 1/ 15000- 18000 children had retinoblastoma globally. In developing countries 10% of paediatric cancer patients were diagnosed with retinoblastoma. [1]

The retinoblastoma develops after random gene (RB1 gene) mutation in retinal cell (photoreceptor cells). The inheritance increases the risk of retinoblastoma.[2]

It was observed that approximately 3 out of 4 patients had unilateral retinoblastoma and 1 had bilateral retinoblastoma.[3]

The manifestations of retinoblastoma depend on its size and location of the tumor. The Cat's eye reflex (Leukocoria) is the commonest presenting manifestation of retinoblastoma and other manifestations are strabismus, Conjunctival hyperemia, blindness and glaucoma.[4]

The various studies have suggested that the retinoblastoma is having good prognosis if it is detected at early stages. In developing countries, the survival rates of patients are low than the developed countries.[5]

Thus, the present study was undertaken to determine the characteristics of retinoblastoma.

Aims and Objectives

1. To describe the clinical characteristics of retinoblastoma patients in Jammu region.
2. To evaluate the pathological patterns in retinoblastoma patients of Jammu region.
3. To assess the immunohistochemistry of retinoblastoma patients in Jammu region.

Material and Methods

The present prospective observational study was conducted in the Department of Ophthalmology at Acharya Shri Chander College of Medical Sciences and Hospital, Jammu, during the period of 2 years (June 2020 to July 2022) after obtaining approval from the institute ethical committee.

A total of 84 patients attending the Eye Out Patient Department were involved after obtaining the informed consent from all the study participants.

A detailed history was taken and ocular examination (slit lamp examination, indirect Ophthalmoscopy, computed tomography and magnetic resonance imaging) was done.

All the patients (eyes) were screened for presenting symptoms, type of tumor, Histopathology and immuno chemical staining and modes of treatment.

Inclusion Criteria

- Age group 0-15 years.
- Patients presenting with Leukocoria, red eyes and eye swelling.

Exclusion Criteria

- Study participants whose guardians were not willing to participate.

Data was analysed and recorded in a excel sheet. Statistical package for social science software (SPSS), version 20 was used to analyse and interpret the data.

Observation and Results

In the present study, 84 patients attending the Eye Outpatient Department were included. The 100 eyes of 84 patients were clinically diagnosed with retino blastoma.

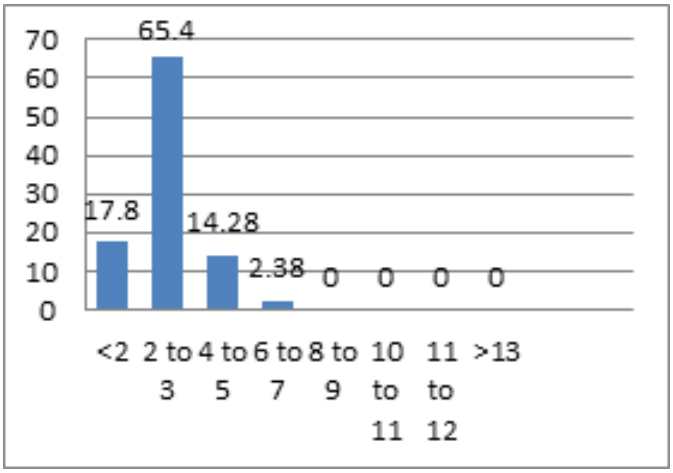
Age of the patient's ranges between 5 months to 7 years; the youngest patient was 5 months old and the oldest was 7 years old with maximum cases were found in the age range of 2-5 years as depicted in table no. 1. The mean age at diagnosis was 22.8±15.68 months.

Table 1: Age distribution of patients.

Age range	No. of cases	Percentage
<2	15	17.8
2-3	55	65.4

4-5	12	14.28
6-7	2	2.38
8-9	0	0
10-11	0	0
12-13	0	0
>13	0	0

Figure 1: Age distribution

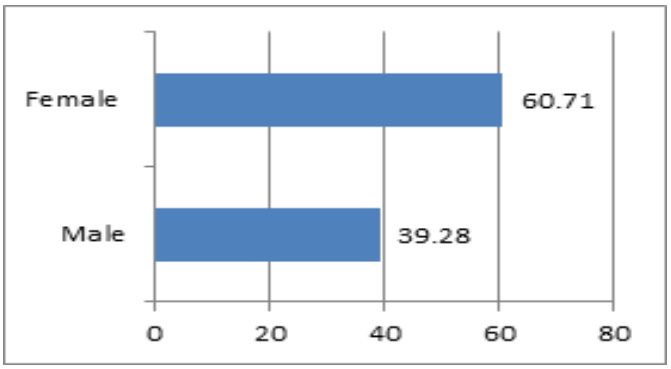


In the present study majority of the cases were female patients60.71% and 39.28% cases were males with the female to male ratio of 1.54:1 as depicted in table no. 02.

Table 2: Gender distribution of patients.

Gender	No. of cases	Percentage
Male	33	39.28
Female	51	60.71

Figure 2: Gender distribution

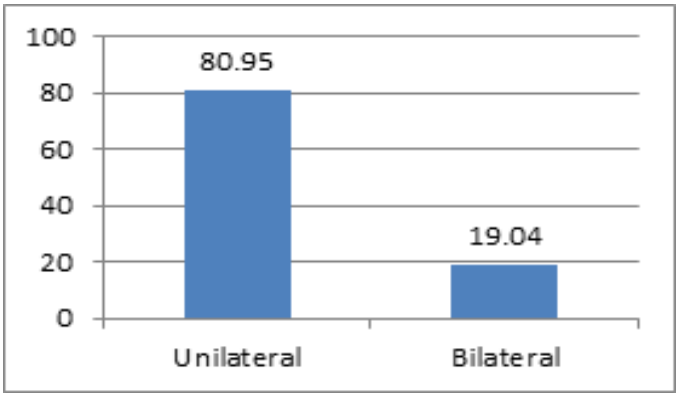


In the majority (80.95%) of the patients unilateral retinoblastoma was detected and 19.04% patients had bilateral retinoblastoma as depicted in table no. 03.

Table 3: Laterality

Laterality	No. of cases	Percentage
Unilateral	68	80.95
Bilateral	16	19.04

Figure 3: Laterality

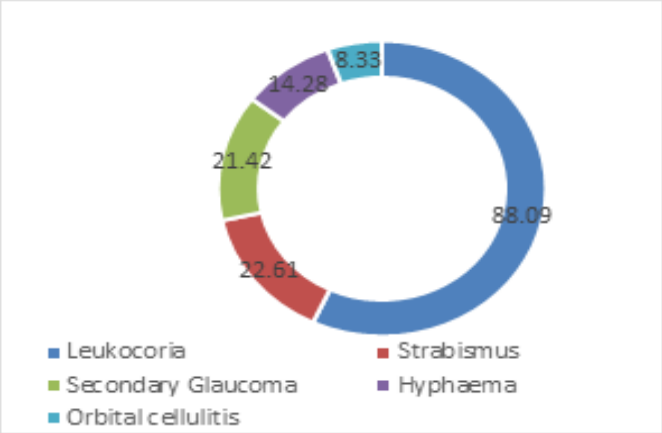


Present study showed that majority (88.09%) patients were presented with Leukocoria followed by strabismus (22.61%), secondary glaucoma (21.42%), Hyphema (14.28%) and Orbital cellulitis (8.33%) as depicted in table 4.

Table 4: Presenting manifestations

Presenting manifestations	No. of cases	Percentage
Leukocoria	74	88.09
Strabismus	19	22.61
Secondary Glaucoma	18	21.42
Hyphema	12	14.28
Orbital cellulitis	7	8.33

Figure 4: Presenting manifestations

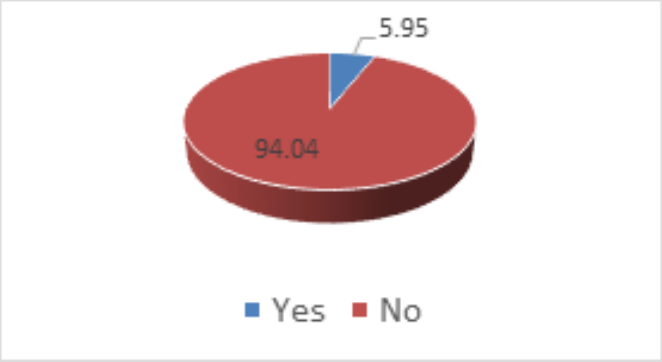


It was reported that 5.95% patients had family history of retinoblastoma as shown in table 5.

Table 5: Family history

Family History	No. of cases	Percentage
Yes	5	5.95
No	79	94.04

Figure 5: Family History

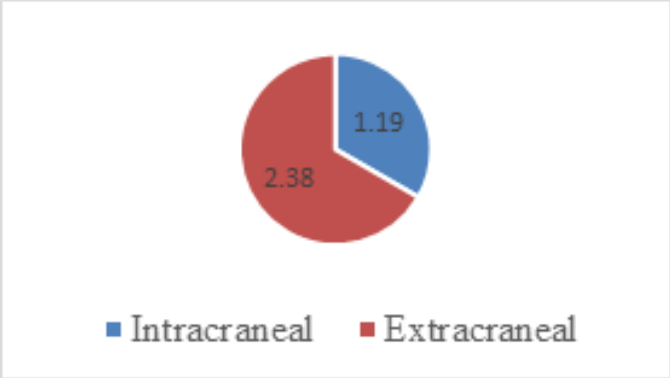


The present study found that 1.19% patients had intracranial spread of tumor and 2.38% patients had extracranial tumor as presented in table 6.

Table 6: Spread of tumor

Spread of tumor	No. of cases	Percentage
Intracraneal	1	1.19
Extracraneal	2	2.38

Figure 6: Spread of tumor

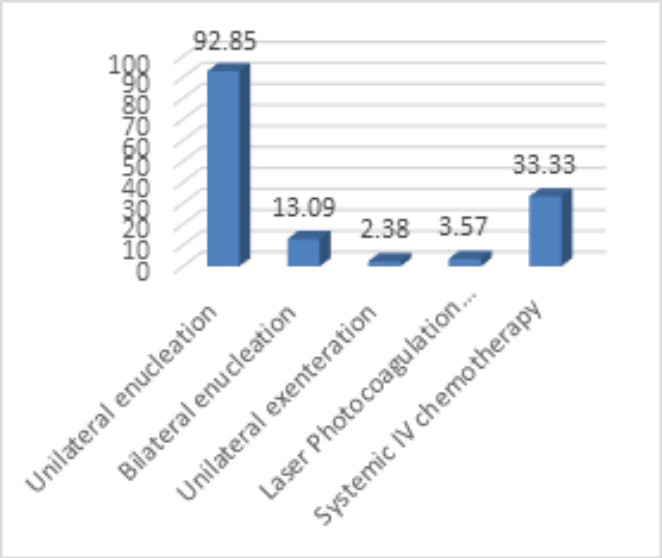


The most common mode of treatment was unilateral enucleation (92. 85%) followed by bilateral enucleation (13. 09%), Unilateral exenteration (2. 38%), Laser Photocoagulation Therapy (3.57%) and Systemic IV chemotherapy (33.33%) as presented in table 7.

Table 7: Modes of treatment

Modes of treatment	No. of cases	Percentage
Unilateral enucleation	78	92.85
Bilateral enucleation	11	13.09
Unilateral exenteration	2	2.38
Laser Photocoagulation Therapy	3	3.57
Systemic IV chemotherapy	28	33.33

Figure 7: Modes of treatment

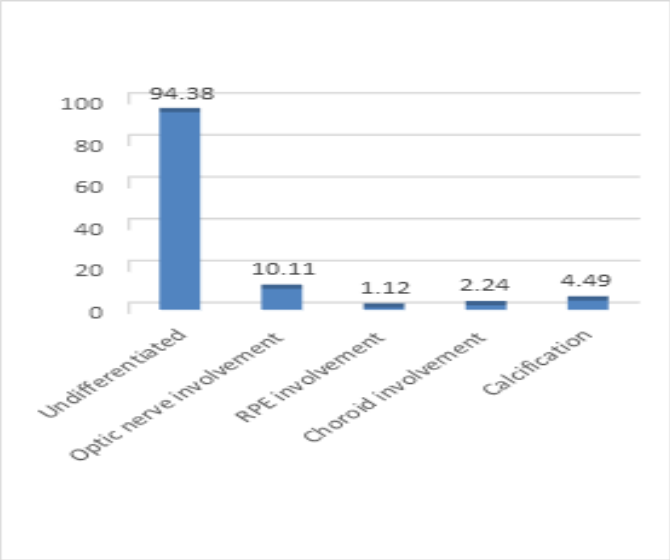


On histopathological examination of eyes it was observed that most of the eyes had undifferentiated pathological type (94.38%) followed by optic nerve involvement (10.11), RPE involvement (1.12%), choroid involvement (2.24%) Calcification (4.49) as depicted in table 8.

Table 8: Histo pathological examination of enucleated eyes

Parameters	No. of Eyes	Percentage
Pathological type (Un differentiated)	84	94.38
Optic nerve involvement	9	10.11
RPE involvement	1	1.12
Choroid involvement	2	2.24
Calcification	4	4.49

Figure 8: Histo pathological examination of enucleated eyes

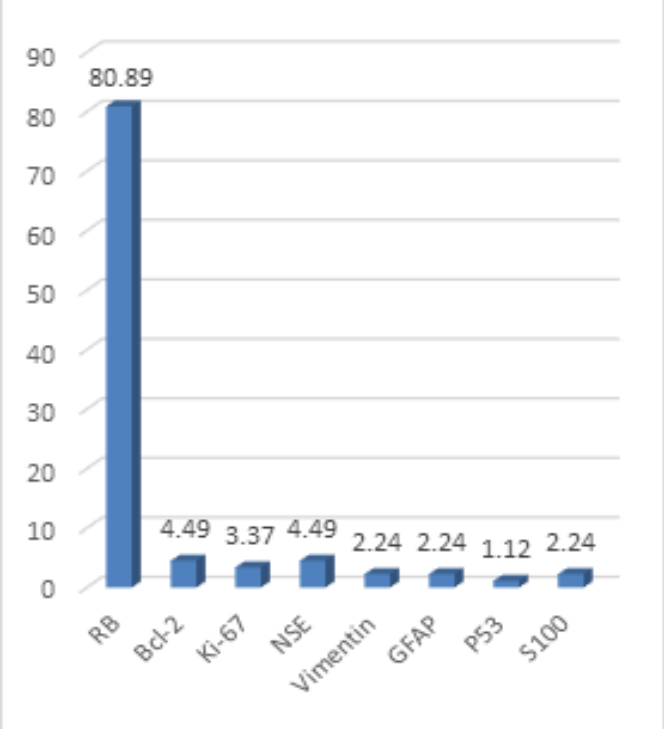


Immunohistochemistry study showed that RB was the most common marker in majority of the cases (80.89%) followed by B-cell lymphoma 2 (Bcl-2) and NSE (4.49% each), Ki-67 protein (3.37), vimentin, GFAP & S-100 (2.24% each respectively) and P-53 (1.12%) as depicted in table 9.

Table 9: Immuno Histochemistry

Markers	No. of cases	Percentage
RB	72	80.89
Bcl-2	4	4.49
Ki-67	3	3.37
NSE	4	4.49
Vimentin	2	2.24
GFAP	2	2.24
P53	1	1.12
S100	2	2.24

Figure 9: Immuno Histochemistry



Discussion

The present study evaluated the 100 eyes diagnosed with retinoblastoma of the 84 patients attending the eye OPD. Data was analysed and discussed with previous literature.

The present study showed that the mean age of the patients at diagnosis was 22.8±15.68 months, females patients were affected mostly (60.71%) with female

male ratio 1.54:1, majority (80.95%) of the patients had unilateral retinoblastoma and 5.95% patients had family history of retinoblastoma. The findings are correlated with the study conducted by Butros JL, et al. 2013, found that the median age of the patients at diagnosis of retinoblastoma was 23.3 months and 72% patients had unilateral retinoblastoma.[6] In similar study conducted by Nabie R, et al. (2012), found that the mean age of the patients was 29.5±13.2 months and 18.5±10 months, majority of the patients were females (57.7%) followed by 42.5% females and most of the patients (57.7%) had unilateral retinoblastoma and bilateral retinoblastoma was found in 42.5% patients.[7]

Present study reported that majority (88.09%) of patients were presented with Leukocoria followed by strabismus (22.61%), secondary glaucoma (21.42%), Hyphema (14.28%) and Orbital cellulitis (8.33%). The intracranial spread of tumor was found in 1.19% patients and extracranial spread of tumor was reported in 2.38% patients. Findings are in accordance with the study conducted by Rodrigues KSE, et al. (2004) observed that majority of the patients had leukocoria (78.9%) presenting manifestation followed by strabismus (10.7%), tumor (3.4%) and other symptoms (7%).[4] In similar study conducted by Faranoush M, et al. (2014) found that the most common presenting symptoms among patients with retinoblastoma were leukocoria (67.51%) and strabismus (25.4%).[8]

In our study the most common mode of treatment was unilateral enucleation (92.85%) followed by bilateral enucleation (13.09%), Unilateral exenteration (2.38%), Laser Photocoagulation Therapy (3.57%) and Systemic IV chemotherapy (33.33%). The findings are consistent with the study conducted by Subramaniam S, et al. (2014) found that the retinoblastoma was treated with different treatment strategies. The unilateral retinoblastoma was treated with enucleation (79% patients), focal therapy was administered in 7% patients 1% patient was treated with EBRT, 95% patients received systemic chemotherapy (mean, 6.5 cycles; minimum, 2; maximum, 13 cycles). The bilateral retinoblastoma was treated with enucleation 41%, focal therapy was performed in 41% patients, radiotherapy for 6% patients and 72% patients were received systemic chemotherapy.[9]A similar study conducted by Kao LY, et al. (2002) observed that majority of the patients with retinoblastoma were treated with enucleation (75%), followed by exenteration (3.12%), radiotherapy (43.75), chemotherapy (33.33), laser photocoagulation (2.08%) and cryotherapy (1.04%).[10]

The histo pathological examination and Immuno histochemistry study of eyes reported that most of the eyes had undifferentiated pathological type (94.38%) followed by optic nerve involvement (10.11), RPE involvement (1.12%), choroid involvement (2.24%) Calcification (4.49). Further RB was the most common marker in majority of the cases (80.89%) followed by B-cell lymphoma 2 (Bcl-2) and NSE (4.49% each), Ki-67 protein (3.37), vimentin, GFAP & S-100 (2.24% each respectively) and P-53 (1.12%). The findings are correlated with the study conducted by Chuluunbat T, et al. (2016), performed histopathological examination and Immuno histochemistry study of 5 enucleated eyes and reported that 47% eyes had undifferentiated pathological type, optic nerve infiltration was seen in 7% eyes, 5.5% had calcification and necrosis or haemorrhage was found in 1 eye. As well as 80% retinoblastoma were classified as undifferentiated RB and 20% was well-differentiated RB, NSE-, Ki-67 protein-, and B-cell lymphoma 2 (Bcl-2)-positive cells were found in all the eyes, and Rb

protein was detected 60% eyes. Vimentin was found in two cases, glial fibrillary acidic protein in three cases, P-53 in two cases, and S-100 in three cases.[8]

Conclusion

The study concluded that the Leukocoria was the most common clinical feature seen in retinoblastoma patients. Undifferentiated Type was the most common Pathological Variant Seen on Histo pathological Examination. It concluded majority of Retinoblastomas expressed RB as the most common marker on Immuno Histochemistry

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