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Original Research Paper

Pathology

HISTOPATHOLOGY OF 4 CASES OF RETINOBLASTOMA: A STUDY AT RIMS RANCHI

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ABSTRACT Retinoblastoma is the most common intraocular neoplasm of children. Retinoblastomas can present at birth but are usually diagnosed between 1 and 2 years of age, with heritable cases arising in younger infants. This is a retrospective study conducted at a Tertiary care centre, RAJENDRA INSTITUTE OF MEDICAL SCIENCES, RANCHI from May, 2018 to April 2019. 4 cases with histopathological diagnosis of retinoblastoma were included in the study. The age of the patients of these 4 cases were 20months, 24months, 31months and 68months. The mean age being 35.75months and median age being 22months. 3 out of 4 cases were male patients. Optic nerve as involved in 2 of the cases. It is important that attention be given to the details of the histopathological report in Retinoblastoma, particularly optic nerve invasion, corneal involvement, anterior and posterior chamber involvement to determine post-enucleation therapy and prognosis.

KEYWORDS : Retinoblastoma,Histopathology, Ranchi.

INTRODUCTION

Retinoblastoma is the most common intraocular neoplasm of children. It has long been thought to arise from retinal stem or progenitor cells, and recent studies suggest both early progenitors and cone precursors as cells of origin for distinct molecular groups of tumors.[1] Retinoblastomas can present at birth but are usually diagnosed between 1 and 2 years of age, with heritable cases arising in younger infants. Approximately 60% of the cases are unilateral, and the other 40% are bilateral, with all bilateral cases and 15% of unilateral cases representing a heritable form of the disease.[2] The responsible gene is located at chromosome 13q14 and designated *RB*.[3] Patients with hereditary RB have a germ cell mutation in one allele and develop the RB as a result of a somatic mutation in the second allele, whereas in patients with sporadic RB both mutations are somatic.[4] This model, known as Knudson's "two-hit hypothesis," has become a paradigm of tumorigenesis.[5] The yearly incidence of inherited RB mutations is 1 in 15,000 to 20,000 live births.[2]

RBs characteristically present as a leukocoria (white pupillary reflex) or less often as a strabismus. Rarely, extraocular extension with the formation of an orbital mass is the presenting manifestation.

Enucleation for retinoblastoma is done in patients with advanced intraocular disease and if there has been failure of conservative treatment. Histopathological reporting of retinoblastoma is critical because certain macroscopic and microscopic features contribute to the staging of the tumor that determines prognosis and post-enucleation therapy.

MATERIAL AND METHODS

This is a retrospective study conducted at a Tertiary care centre, **RAJENDRA INSTITUTE OF MEDICAL SCIENCES, RANCHI** from May, 2018 to April 2019. 4 cases with histopathological diagnosis of retinoblastoma were included in the study. Data related to age at the time of enucleation, sex, laterality, corneal involvement, anterior and posterior chamber involvement and optic nerve invasion and presence of necrosis and calcification was obtained.

RESULTS

Total 4 cases were included in the present study. The age of the patients of these 4 cases were 20months, 24months, 31months and 68months. The mean age being 35.75months and median age being 22months. 3 out of 4 cases were male patients. In all the cases, the tumor was unilateral. Optic nerve as involved in 2 of the cases. The cornea was not involved in all of the cases. Necrosis was present in 1 case while calcification was present in 3 cases.

- **DEMOGRAPHIC VARIABLES:**

AGE	
Mean age	35.75 months
Median age	22 months
SEX	
Males	3 (75%)
Females	1 (25%)
LATERALITY	
Unilateral	4 (100%)
Bilateral	0 (0%)

- **HISTOPATHOLOGICAL VARIABLES:**

VARIABLES	NUMBER OF CASES PRESENT IN
Corneal involvement	0
Anterior chamber involvement	1 (25%)
Posterior chamber involvement	4 (100%)
Optic nerve invasion	1 (25%)
Necrosis	1 (25%)
Calcification	3 (75%)

DISCUSSION

In our study the mean age is 35.75months and median age is 22months while it was 29.74 months and 24 months respectively in study conducted by Gupta et al[6] & 32.88 months and 30months respectively by Kashyap et al[7]. In our study, 75% of patients were males while in case of studies conducted by Gupta et al[6] and Kashyap et al[7] the percentage of male patients affected by retinoblastoma were 60% and 65.7% respectively. In our study all the cases had unilateral involvement while in case of studies by Gupta et al[6] and Kashyap et al[7], the unilateral cases were 65% and 73.7% respectively. In our study, optic nerve was involved in 25% cases, while the percentage for the same were 8% and 7.4% respectively in studies conducted by Gupta et al[6] and Kashyap et al[7]. The differences in these variables among our study and other studies could be due to smaller sample size in our study and also due to difference in the demography of different regions.

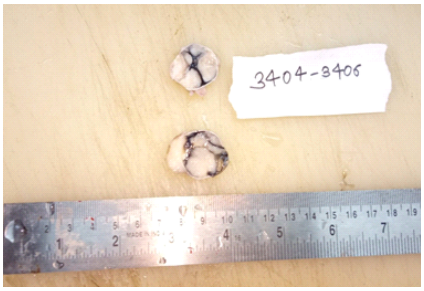

Figure 1: Gross appearance of retinoblastoma in the enucleated eye from a 24months old male patient.

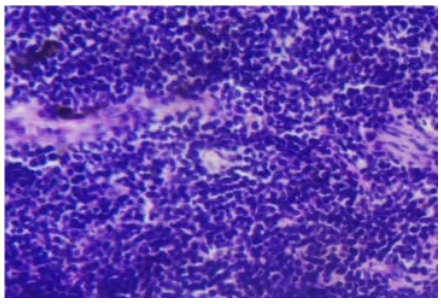

Figure 2: Small round blue cells in retinoblastoma from a 24months old male patient (High power, H&E)

CONCLUSION

Retinoblastoma is the most common intraocular tumor of childhood and it is important that attention be given to the details of the histopathological report in Retinoblastoma, particularly optic nerve invasion, corneal involvement, anterior and posterior chamber involvement to determine post-enucleation therapy and prognosis.

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