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Vitreous Seeding, Choroid and Sclera Involvement in Retinoblastoma Patients

Dr. Himanshu Jodhani

Abstract: ***Vitreous seeding, Choroid and Sclera Involvement in Retinoblastoma patients is the aim of the study. This study has been conducted in the M&J Western Regional Institute of Ophthalmology, Civil Hospital, Ahmedabad, Gujarat between the period of January 2013 to December 2014.In this study 40 cases were included. A detailed histopathological examination was done. Choroid involvement is most common in retinoblastoma patients while scleral involvement is least common.***

Keywords: Retinoblastoma, Vitreous seeding, choroidal involvement, scleral involvement

1. Introduction

Retinoblastoma is a rare eye tumor of childhood that arises in the retina and represents the most common intraocular malignancy of infancy and childhood. It may occur at any age but most often it occurs in younger children, usually before the age of two years. The incidence is 1 in 15,000–20,000 live births. In 60% of cases, the disease is unilateral and the median age at diagnosis is two years. Of these cases, 15% are hereditary. Retinoblastoma is bilateral in about 40% of cases with a median age at diagnosis of one year. All bilateral and multifocal unilateral forms are hereditary.

Leukocoria (white reflection in the pupil) and strabismus are the most frequent clinical manifestations of retinoblastoma. Leukocoria is initially inconstant, visible only at certain angles and under certain light conditions. This sign may be seen on flash photography. Strabismus, when present, becomes rapidly constant, reflecting impairment of the vision. These signs are still all-too-often overlooked and justify an ophthalmological consultation with ocular fundus examination. Some other signs may be observed, including iris rubeosis, hypopyon, hyphema, buphthalmia, orbital cellulitis, and exophthalmia. Some children with retinoblastoma may have no symptoms. Screening in case of familial history or dysmorphic syndrome with a 13q14 deletion may lead to diagnosis of retinoblastoma. Most affected children are diagnosed before the age of five years.

2. Pathology

Retinoblastoma is characterized histopathologically by malignant neuroepithelial cells (retinoblasts) that develop within the immature retina. The retinoblasts typically appear to have a large basophilic nucleus and scanty cytoplasm. Cellular necrosis, intralesional calcification and sleeve growth surrounding feeding retinal blood vessels are frequent associations, especially in larger tumors. In some cases, tissue differentiation occurs, often producing Flexner-Wintersteiner rosettes or Homer Wright rosettes. Retinoblastoma has a strong tendency to invade the optic nerve and choroid and extend out of the globe via either the optic nerve or scleral emissary canals.

3. Materials and Methods

This study has been conducted in the M&J Western Regional Institute of Ophthalmology, Civil Hospital, Ahmedabad, Gujarat between the period of January 2013 to December 2014.In this study 40 cases were included. A detailed histopathological examination was done. Thorough clinical examination was done. A detailed history was taken from parents. Relevant medical and surgical history was obtained.Under the ocular examination following parameters were recorded:

1. Visual acuity: if child was co-operative
2. Anterior segment examination: on torch light/ on slit lamp if child allows
3. Posterior segment examination: (with direct and indirect ophthalmoscopy.)
4. B-scan USG (if fundus is not visible)
5. Routine Investigation of the patient (hemogram, ESR, S.HIV, S.HBsAg, RBS, Bl.urea, S.creatinine, urine-routine & micro)
6. MRI to rule out optic nerve and extraocular spread
7. Whole eyeball was sent for histopathological examination

4. Results

This study has been conducted in the M&J Western Regional Institute of Ophthalmology, Civil Hospital, Ahmedabad between the period of January 2014 to March 2015.There were 24 male children and 16 female children. Age distribution of patients is given in table no. 1 below:

Age of patient	*Number of patients*
<1 yr	9 (22.5%)
1-3 yrs.	17(42.5%)
4-6 yrs.	12 (30%)
>6 yrs.	2 (5%)
Total	40

Vitreous, choroid and scleral involvement was present in following number of patients as shown in table no. 2

Table 2

	Present	*Absent*
Vitreous seeding	29(72.5%)	11
Choroidal involvement	32(80%)	8
Scleral involvement	12(30%)	28

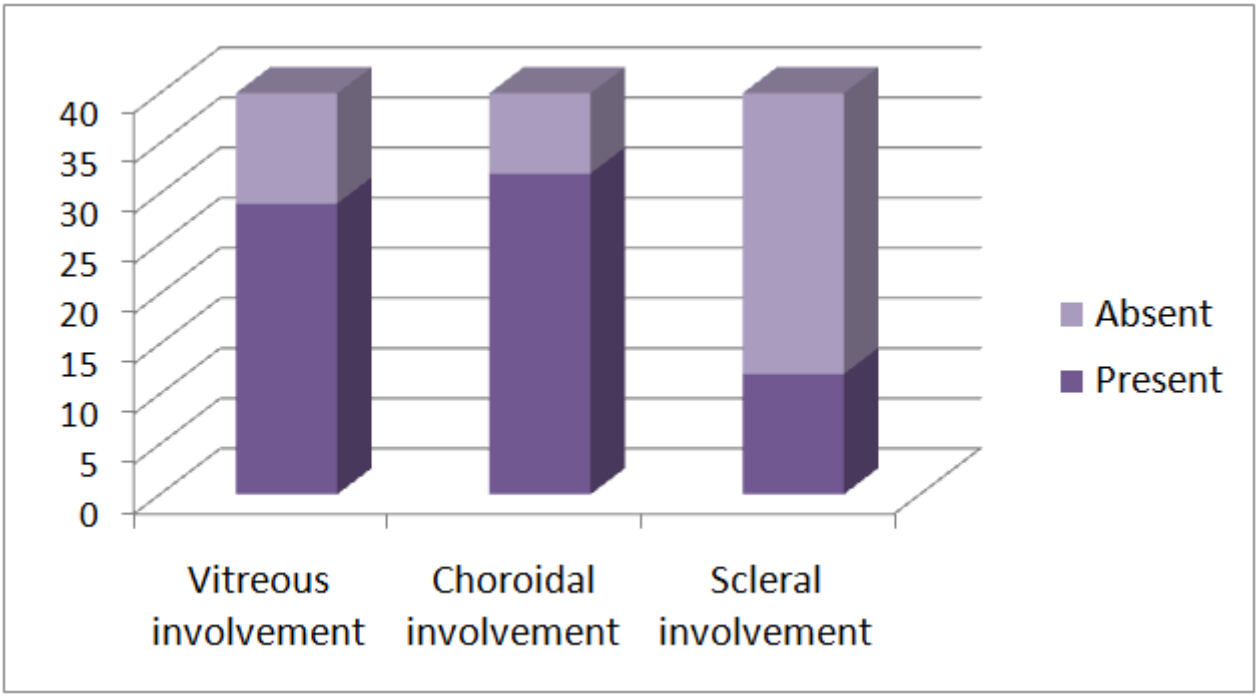

Figure 1

Multiple sites were involved as shown in table no. 3.

Table 3

Involvement	*Total number of patients*
Vitreous + Choroid	26(65%)
Choroid + Sclera	11 (27.5%)
Vitreous + Sclera	10 (25%)
Vitreous + Choroid + Sclera	10(25%)

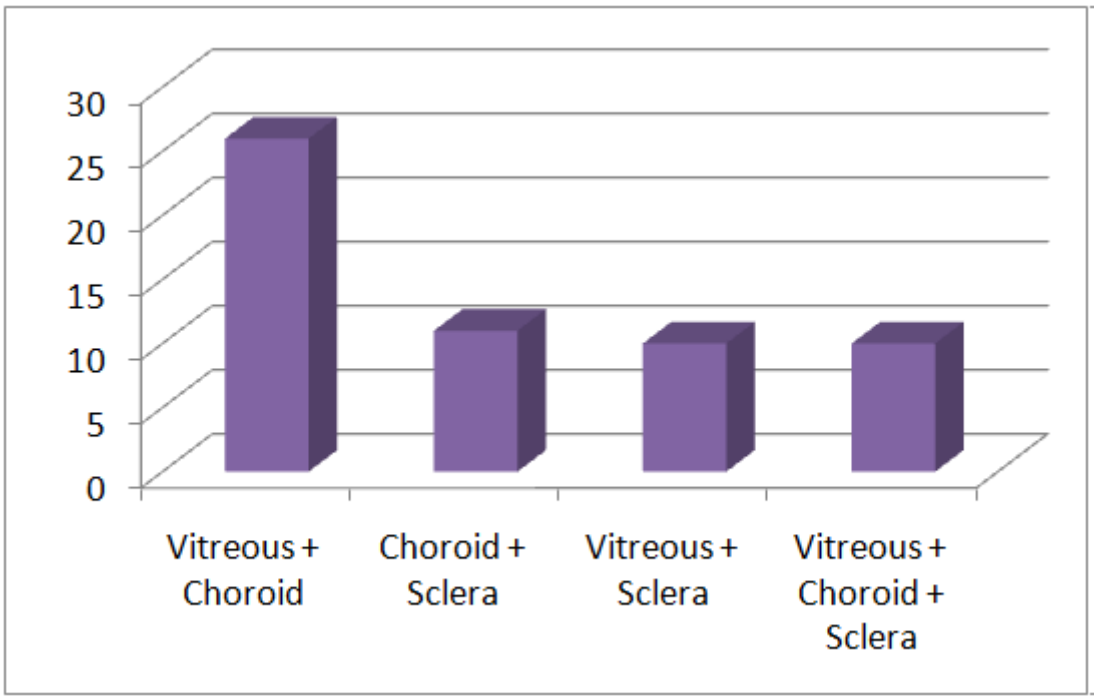

Figure No. 2

5. Discussion

Retinoblastoma is a primary malignant neoplasm of the retina that arises from immature retinal cells.Retinoblastoma with vitreous seeding has been one of the most challenging conditions for eye-preservation therapy. Several modalities for treating vitreous seeding were reviewed in order to analyze the problems associated with them. External beam radiotherapy has been the most reliable method to treat vitreous seeding. However, recurrence after external beam radiotherapy needs other types of treatments to preserve the eyeballs. Due to the progress of investigations concerning retinoblastoma, chemotherapy has become the most promising method to cure not only recurrence but also primary tumors. Systemic chemotherapy can rarely cure vitreous seeding, but local chemotherapy using vitreous injections of melphalan can preserve about 50% of the eyeballs with vitreous seeding. Currently, animal experiments are being conducted to study the efficacy and safety of vitreous surgery combined with infusion of anticancer drugs for eradication of vitreous seeds and maintenance of visual function. Thus, study of vitreous, sclera and choroidal involvement is important for management of retinoblastoma patients.

6. Limitations of Our Study

We conducted study for 2 years at tertiary care centre. So, we could include patients coming to our institute only. We studied vitreous, choroid and scleral involvement only in enucleated eyes of patients.

7. Conclusion

Choroid involvement is most common in retinoblastoma patients.Scleral involvement is least common.Vitreous + choroid involvement is seen in maximum number of patients.

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