ORIGINAL ARTICLE

Childhood Retinoblastoma

by

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Abstract

Childhood retinoblastoma was studied in a period of January 1980 to December 1988. Forty six patients were included consisting of 21 males (45.65%) and 25 females (54.35%). Most of those patients (76.08%) were in the age group of 0 - 5 years. The main complaint was protrusion of eyes, found in 42 patients (91.30%). Bone marrow examination was done in 24 patients, and 52.17% of them showed sign of metastasis. Thirty seven cases had unilateral retinoblastoma (80.43%).

The statistical analysis indicated no significant differences in sex and age, site of tumor (left or right eye), abnormality of the eyes, bone marrow involvement, protrusion of the eyes either unilateral or bilateral.

Heredity was found in one patient (2.17%). The main treatment was radiation in 25 patients (50.43%).

Introduction

Retinoblastoma is a type of malignant tumor in children and it is uncommon in adults. This type of tumor originated from the retinic layer of the eye ball and it tends to lead to blindness (Hutchinson, 1972; Leonard et al., 1987); it is one of the three types of malignant tumor commonly found in children (Dunphy et al., 1977).

This tumor affected children under five years old (Reimenscheneiders, 1983). It can be hereditary (Kempe et al., 1970).

The clinical manifestations depend on the site and size of the mass in the retina. The mass in the macular area may disturb vision, and may produce cross vision. A tumor of bigger than half of the retinic area may result in cat's eye at night. This tumor extends through the optic nerves of emiseria sclera, while a tumor in the orbital space may suppress the tissues outwardly, tending to the formation of proptosis as a late stadium of retinoblastoma (Leonard et al., 1987).

The purpose of this paper is to describe the pattern of retinoblastoma in patients admitted to the laboratory of Pediatrics, Dr. Pirngadi Hospital, Medan, since January 1980 to December 1988.

Materials and Methods

Subjects of this study were patients with retinoblastoma admitted to the Laboratory of Pediatrics, Dr. Pirngadi Hospital, Medan in a period from January 1980 to December 1988. Sex, age, complaints, site of tumor at onset, family history, bone marrow examination and treatment were noted.

Patients were defined having metastasis when bone marrow examination indicated the presence of atypical cells, the qualitative data were analysed statistically by the Chi-square test.

Results

Those 46 patients studied were 25 females (54.35%) and 21 males (45.65%). Statistical analysis indicated no significant differences either in sex or age group ($p > 0.05$) (Table 1).

Most of the patients (76.08%) were on the first admissions in the age group of under five years.

Table 1 : *Distribution of sex and age group*

Age (years)	Sex		Number	%
	Male	Female		
0 - 5	15	21	36	76.08
6 - 10	4	2	6	12.04
11 - 15	2	2	4	11.88
Total	21 (45.65%)	25 (54.35%)	46	100.00

In a period of one year (from 1987 to 1988) the number of cases increased. Statistical analysis showed no significant differences between the earliness in the above periods and the age groups ($p > 0.05$) (Table 2).

Table 2 : *Correlation between age group and duration of hospitalization*

Age (years)	Duration of hospitalization				Number
	1980 - 1986		1987 - 1988		
	Number	%	Number	%	
< 5	9	69.23	27	81.81	36
> 5	4	30.77	6	18.19	10
Total	13		33		46

From 46 patients with retinoblastoma, 37 were unilateral and 9 bilateral. Bone marrow examination in the 46 patients indicated that 24 out of those patients had metastasis.

Proptosis appeared predominantly in 42 patients, unilateral cat's eye in 4 cases and unilateral proptosis in 33 cases. No significant differences were found between the bone marrow metastasis and eye disorder in the unilateral cases ($p > 0.05$). (Table 3).

Table 3 : *Correlation between unilateral eye disorders and bone marrow metastasis.*

Eye disorders	Bone marrow metastasis				Number
	+		-		
	Number	%	Number	%	
Proptosis	18	54.54	15	45.46	33
Cat's eye	2	50.00	2	50.00	4
Total	20		17		37

Bone marrow metastasis was found in cases of unilateral and bilateral proptosis (Table 4).

Table 4 : *Relationship between unilateral or bilateral proptosis and bone marrow metastasis*

Proptosis	Bone marrow metastasis				Number
	+		−		
	Number	%	Number	%	
Unilateral	18	54.54	15	45.46	33
Bilateral	4	44.44	5	55.56	9
Total	22		20		42

Whether the left or right eye, male or female sex and any age group were at the same risk for early symptoms of retinoblastoma. Statistical analysis showed no significant differences (Table 4 and 6).

Table 5 : *Distribution of retinoblastoma by early eye symptoms and sex*

Sex	Early symptom of retinoblastoma				Number
	Right eye	%	Left eye	%	
Male	12	42.85	9	50.00	21
Female	16	57.15	9	50.00	25
Total	28		18		46

Table 6 : *Distribution of retinoblastoma by early eye symptoms and age group*

Age (years)	Early symptom of retinoblastoma				
	Right eye	%	Left eye	%	
< 5	22	78.57	14	77.77	36
> 5	6	21.43	4	22.23	10
Total	28		18		46

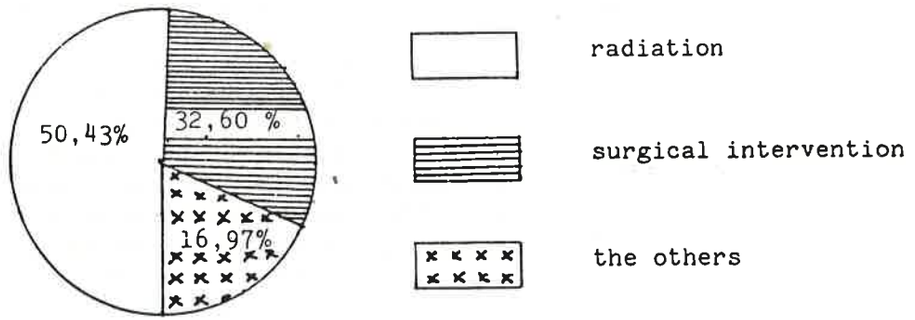

Figure 1 : *Treatments*

The main therapy was radiation of the tumor masses in 25 (50.43%) patients, surgical intervention in 15 (32.60%), radiation plus surgery in 4 (12.63%), radiation and cytostatics in 1 (2.17%) and cytostatics alone in 1 (2.17%) (Fig 1).

Of all cases, only one (12.17%) had the family history of retinoblastoma.

Discussion

In this study, although females appeared more than males for the age group of under five statistical analysis showed no significant differences. The mayority of the patients belonged to the age group of under five. This was similar to the earlier report that indicated no interferences of sex in retinoblastoma. Usually, it was found in children younger than 5 years (Reimenscheneiders, 1983). Marwoto (1984) in Jakarta, reported the opposite namely where males appeared more than females in the age group of under five. This difference might be due the differences in time and number of cases.

During 1987-1988 the number of patients with retinoblastoma increased which might be due to the functioning of the Team of Cancer of Dr. Pirngadi Hospital, Medan, where the cancer patients were mostly found in the age group of younger than 15 years, particularly patients with retinoblastoma admitted in Pediatric ward. Statistically, no significant differences in the age group of patients with retinoblastoma were found in the period of 1980 - 1986 compared to those in the period of 1987 - 1988.

Retinoblastoma is commonly bilateral, and patients usually present with proptosis of their eyes. Proptosis or cat's eye is not necessarily an indication of retinoblastoma with bone marrow metastasis (Table 3). So is it whether unilateral or bilateral (Table 4).

These findings seemed similar to an earlier report suggesting that the extension from one orbital space to the other one was through the subarachnoid space along the optical nerves while bone marrow metastasis was hematogenous. Proptosis

occurred in the late process of a tumor in the orbital space thus suppressing the eye balls outward which resulted in proptosis; bilateral cases appeared in one-third of retinoblastoma cases (Kempe et al., 1970).

A study in Jakarta in the year of 1982, suggested that proptosis is predominant (Moeloek et al., 1984). In contrast to the study in United States, Shields and Audburger stated that retionoblastoma with proptosis were rare (Binder, 1977). This difference might be due to differences in economic states, public knowledge, and the procedures of study.

The risk for contracting early symptoms of retinoblastoma of the right or left eye was similar when correlated to sex (Table 5) or age group (Table 6). This was in accordance with the literature (Kempe et al., 1970; Reimenscheneiders, 1983).

Radiotherapy is a common treatment, this is also in accordance with the literature stating that radiation is effective in retinoblastoma which indeed is radio sensitive (Dunphy et al., 1977). Surgical intervention is done for the patients without the signs of bone marrow metastasis, followed by radiation to minimize the tumors; cases with bone marrow metastasis in our study were treated with radiation although it should actually be radiation plus cytostatics. Cytostatics in our cases were given only to two patients; all cases had some irregular treatment thus we found it different in the evaluation of the course of the disease and treatment.

Only one case had a history of retinoblastoma in their family; Kempe et al., (1970) also reported the role of a hereditary factor in retinoblastoma.

Conclusion

The mayority of the patients with retinoblastoma were found in the age group of younger than 5 years. The increase in number of patients with retinoblastoma might be due to the functioning Team of Cancer of Dr. Pirngadi Hospital, Medan. The presence of the bone marrow metastasis is not correlated with either the symptoms found in the eyes, unilateral or bilateral. The risk of whether it is for retinoblastoma to the right or left eye is similar when correlated to sex and age group.

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