PROBLEMS OF MANAGING RETINOBLASTOMA IN JOS UNIVERSITY TEACHING HOSPITAL A 6 YEAR REVIEW OF CASES

*P. D. WADE, C.D. MPYET, S.K. ALLI AND L. LANGNAP

Corresponding Author: Department of Ophthalmology
Jos University Teaching Hospital

ABSTRACT:
AIM: To review the cases of retinoblastoma patients who presented to Jos University Teaching Hospital during the 6year period, between 1997 to 2001 and to determine the problems in managing the patients.

METHODS: Patients records were retrieved from the library. Information on age, sex, clinical presentation, past medical history, family history, clinical examination, laboratory results and treatment received before and after presentation. A total of 27 cases of patients were reviewed.

RESULTS: 27 cases were reviewed out of which 20 (74%) presented with fungating mass and proptosis. Leukocoria was seen in 5 (18.52%) and only 2 (7.41%) patients presented with poor vision and redness. Late presentation was seen in 15 (55.55%) patients. This was attributed to visits to peripheral clinics by 18 (66.67%) patients and to alternative healers by 14 (51.58%). Only 8 (29.63%) patients benefited from surgery and cytotoxic drugs. The two years survival rate was 3.7%.

CONCLUSION: Ignorance of the disease, inadequate health education, poor financial status of most patients, late presentation to the hospital contributed to high mortality

KEY WORDS: Retinoblastoma, Problems, Management

INTRODUCTION

Retinoblastoma is a blast tumour of the outer nuclear layer of the retina. It is the most common intraocular tumour of childhood and second only to malignant melanoma of the choroids as the most common tumour of any age group [1]. Retinoblastoma exhibits an autosomal dominant trait with greater than 90% penetrance. It is recessively inherited since both retinoblastoma genes at the 13q14 locus must be abnormal before the cell becomes malignant. In the non hereditary form, both mutations occur only in the retinal cell that has become malignant [2]. Globally, there are estimated to be 1.4 million children who are blind, and around three-quarters live in developing countries [3]. The incidence is about 1 in every 20,000 live births. Retinoblastoma is one of the childhood tumour that has one of the highest cure rates. If untreated, the disease is invariably fatal [2]. In developing countries, late presentation, lack of facilities, personnel for scientific eye care, ignorance and poverty have all contributed to the high mortality rate seen in this disease [4]. The aim of this study is to highlight the difficulties faced in managing this disease which is considered to have one of the highest cure rates [2] in developed countries but almost 0% cure rate in developing countries.

MATERIALS AND METHODS:

A total of 33 patients were seen during the 6-year study period from January 1996 to December 2001. But only records of 27 patients could be retrieved from the library. The records of 6 patients could not be traced. Information on age, sex, clinical presentation, past medical history, family history, and treatment received before and at presentation were obtained. General and ocular examination of the patients was done. Various investigations available in the hospital were done. These involved full blood count, haemoglobin, genotype, orbital x-rays and history of enucleated/exentherated eyes. Treatments given in Jos University Teaching Hospital were recorded, and records of those being followed up available to us were reviewed. Even those who signed and left against medical advice were noted. The information so obtained was analysed and results presented.

RESULTS

Most of the patients seen presented late with proptosis 13 (48.15%) and fungating masses 7 (25.92%) (Table 1). Other forms of presentation

included leukocoria seen in 5 (18.52%) and poor vision and redness seen in 2 (7.41%) patients.

Although most of the parents were not sure of the duration of the illness before presentation, as some of the children lived with their grandmothers or foster parents as is the practice in this environment where when a child to be weaned is handed over to the grandparents or some other relatives, however duration before presentation ranged from one month to eight months as shown in Table II. About 15 (55.55%) patients showed delayed presentation, and this is attributed to visits to peripheral clinics 18 (66.67%) and alternative medicine 14 (51.85%) as shown on Table III.

The only forms of treatments available here are cytotoxic drugs, surgery and cryotherapy. Cryotherapy can only be done on patients who present early but no patient was qualified for this form of treatment (Table 1). Only 8 (29.63%) of the patients agreed to enucleation/exentheration, while 8 (29.63%) (Table IV) agreed to surgery and cytotoxic drugs but could not all complete the required 6 cycles because of financial constrains, or were lost to follow up or the child might have died at home in between the cycles. The survival rate was very low with two (7.40%) patients followed up for two months before they died while one (3.7%) patient has received the 6 cycles of cytotoxic drug spanning two clinic years. (Table V).

TABLE I: PRESENTING COMPLAINTS

S/NO	COMPLAINT	NUMBER	PERCENTAGE
1	White sport (leukokoria)	5	18.52
2	Protruding moss (fungating)	7	25.92
3	Proptosis	13	48.15
4	Poor Vision	1	3.7
5	Redness	1	3.7
6	Strabismus	0	0
TOTAL		27	100%

TABLE II: DURATION OF ILLNESS BEFORE PRESENTATION

S/NO	DURATION (days)	NUMBER	PERCENTAGE
1	1 - 30	12	44.44
2	31 - 60	4	14.81
3	61 - 90	4	14.81
4	91 - 120	4	14.81
5	121 - 150	1	3.7
6	151 - 180	0	0
7	181 - 210	1	3.7
8	211 - 240	1	3.7
9	>240	0	0
	TOTAL	27	100%

TABLE III: TREATMENT GIVEN BEFORE PRESENTING

S/NO	TREATMENT	NUMBER	PERCENTAGE
1	Orthodox only	7	25.93
2	Alternative medicine only	3	11.11
3	orthodox and Alternative medicine	11	40.74
4	No Treatment given	6	22.22
	TOTAL	**27**	**100%**

TABLE IV: TREATMENT GIVEN IN JUTH

S/NO	COMPLAINT	NUMBER	PERCENTAGE
1	Enucleation (only)	5	18.52
2	Exentheration (only)	3	11.11
3	Cytotoxic drugs only	0	0
4	Surgery + cytotoxic drugs	8	29.63
5	No Treatment given	11	40.74
	TOTAL	**27**	**100%**

TABLE V: PATIENTS SURVIVAL RATE

S/NO	MONTHS	NUMBER	PERCENTAGE
1	0 - 6	2	7.41
2	7 - 12	0	0
3	13 - 18	0	0
4	19 - 24	1	3.7
5	25 - 30	0	0
6	Loss to follow up	24	88.89
	TOTAL	**27**	**100%**

DISCUSSION

Retinoblastoma is a common childhood tumour which accounts for only 3% of childhood cancer [5]. The outcome of patients with retinoblastoma is strongly related to the extent of the disease at diagnosis. This is why of the 27 patients seen only one (3.70%) has been followed up for the past 2 years.

In developing countries, retinoblastoma is still associated with high mortality often approaching 100%[4] This is due to late presentation as seen in our studies where only 2 (7.40%) of the patients presented with redness and poor vision, even then the tumour size had exceeded Reese-Ellsworth grade V.The single most important factor responsible for improvements in retinoblastoma survival rate in . The single most important factor responsible for improvements in retinoblastoma survival rate in developed countries has been the use of highly effective chemotherapy regiments as an adjunct to surgery and to radiation [6]. Although enucleation remains the standard treatment for advanced unilateral retinoblastoma and for the worse eye in most bilateral cases, many eyes can now be preserved by conservative therapy [5]. Only

surgery and cytotoxic drugs combination of vincristine, actinomycin D and cyclophosphamide is offered in this centre. Even then not all patients could afford the cost of a single cycle which is about N4, 000.00 (four thousand naira) or $30 (thirty dollars). A report on the use of external beam radiation for retinoblastoma from St. Bartholomew's Hospital, London showed that of the 175 eyes treated, the overall ocular cure rate was 57% and these children were followed up for 2 17 years [7]. While in our study and others [4] [8] done in this country, most patients were lost to follow up. Considering the high fatal rate of the disease especially when presented late, coupled with ignorance about the disease, these patients' studied are presumed dead.

Many people are so deeply rooted within traditions that at time of trouble, they first seek alternative medication before presenting to the ophthalmologist even when medical services are available nearby [7]. Besides in Nigeria, there are few ophthalmologists, most of who reside in urban areas. The alternative medicine is easily accessible in the rural areas is the first line of medical attention. [9]. In this review, 3 (11.11%) of patients admitted seeing a traditional healer before presentation while 11 (40.74%) saw traditional cum orthodox practitioners like patent medicine vendors, and peripheral clinics and hospitals. The visits to these traditional healers, and non-specialist clinics and hospitals led to waste of valuable time and hence late presentation. In this country there are few centres with radiotherapy facilities and patients have to travel long distances across the country for treatment.

Health education is the most important way of reaching to people. This involves using electronic media to inform people about the existence of this disease. Even among the educated little is known about this disease. Pregnant women attending antenatal clinics should be educated on the existence of the disease and hence the need to pay adequate attention to the eyes as the child grows.

Community extension workers who see most of these patients in the rural areas and other semi-urban and urban centres should be trained to recognize the early forms of the disease, and advice the patients for immediate and early presentation in the treatment of the disease as it's primordial stage.

- Facilities for diagnosis and treatment should be located at strategic places where they can be accessible to people in various parts of the country.
- Government and NGOs should assist by supplying cytotoxic drugs which should be given free to children 0-15 years. This will go a long way to help the poor who cannot afford the cost of these drugs
- If one patient is affected with familial retinoblastoma or bilateral sporadic retinoblastoma, each of the offspring will have a 50% chance of inheriting the tumour [2]. Genetic counseling is very essential especially in survivors of the disease and where there is a family history.

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REFERENCES:

1. Newel F.W; Ophthalmology Principles and Concepts. Mosby yearbook. 7th edition, St. Louis; (1989) 314 317
2. Pavan-Langston D; Manual of Ocular Diagnosis. Third edition. Place of Publication Little Brown and Company (Inc). BOSTON; 281-283
3. Gilber C; New issues in Childhood Blindness. *Community Eye Health. 1991 4:53.*
4. Ezepue U.F; Maduka-Okafor C; Retinoblastoma a review of occurrence, available treatment and prognosis at Enugu, Nigeria. *Nigerian Journal of Ophthalmology 1995; 3:*
5. Hungerford J.L; Toma N.M.G; Plowman P.N. and Kingston J.E; External beam radiotherapy for retinoblastoma One whole technique. *British Journal of Ophthalmology 1995; 79: 109-111*
6. Hungerford J.L; Factors influencing metastasis in retinoblastoma. *British Journal of Ophthalmology, 1993;9:541*
7. Dudges J; Retinoblastoma Trends in Conservative Management. *British Journal of Ophthalmology, 1995;9: 104*
8. Adejor G.O; Retinoblastoma as seen in National Eye Centre, Kaduna A 4 Year retrospective study. *Nigerian Journal of Ophthalmology 1998;6:9-15*
9. Osahon A.I; Consequences of traditional eye medications in UBTH Benin City. *Nigerian Journal of Ophthalmologist, November 1995, 3:51-54.*