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Presentation of Retinoblastoma Patients in a Missionary Eye Hospital in Nigeria

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Authors' contributions

This work was carried out in collaboration between all authors. Author AAO designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors SOA and SAHC managed the analyses of the study. Author SAHC managed the literature searches. All authors read and approved the final manuscript.

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ABSTRACT

Background: The clinical presentation of retinoblastoma is essential in the diagnosis, treatment and prognosis for the survival of patients. Moreover, early detection and treatment of the disease can reverse the disease process and influence the modality of management with the eventual saving of the eye, vision and life of the patients.
Objective: To review the variants of presentation of retinoblastoma cases at Evangelical Church of West Africa (ECWA) Eye Hospital, Kano, Nigeria between December 2006 and January 2014.
Methods: Prospectively 120 cases of retinoblastoma were studied after obtaining consents from their parents. The patients' bio-data and relevant medical history were obtained and subsequently analysed using SPSS version 20.
Results: There were seventy (58.3%) male patients. The mean age at presentation was 2.7±1.4 years. The average time lapse between the manifestation of symptoms and presentation to the hospital was 7.1 months. The commonest form of presentation was proptosis (61.7%).
Conclusion: Patients with retinoblastoma presented late and with an advanced form of the disease

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in ECWA Eye Hospital, Kano. The commonest forms of presentation are proptosis, cat eye reflex and fungating mass. There were 19 cases of bilateral retinoblastoma and 101 cases with unilateral neoplasms.

Keywords: Retinoblastoma; presentations; ECWA Eye Hospital.

1. INTRODUCTION

Retinoblastoma is a rapidly developing cancer, arising from the immature neural retinal cells. A genetic mutation involving "Retinoblastoma gene 1 (Rb1) at chromosome 13 has a significant role in its aetiology [1]. In Nigeria, retinoblastoma accounts for about 1% of all childhood deaths from cancer [2-4]. It is the commonest paediatric intraocular tumour in children, occurring approximately 1 in every 20,000 live birth [5]. Timely presentation of patients with retinoblastoma to health facilities is a determining factor in timely diagnosis, choice of treatment modality, preservation of the eyeball and vision as well as the survival of the patient. This depends on the stage of the disease process at which the child is brought to the health facility and timely referral to the paediatric ophthalmologist. At an early stage the usual presentation is leukocoria, however, patients may develop strabismus, inflamed or painful red eye. Presentation of late cases varies from proptosis to fungating mass with secondaries. Other variants of presentation of retinoblastoma are inflammatory signs such as chemosis, hyphaemia; ptosis, aniridia, phthisis bulbi, nystagmus, anisocoria, heterochromia iridis vitreous haemorrhage microphthalmia/ buphthalmia [6-8].

The first presentation of retinoblastoma in children to health facilities differs in different localities because of different levels of awareness of the disease among the public, availability and accessibility of medical facilities as well as differences in cultural/religious beliefs. Parents/caregiver may not accept the diagnosis of cancer in the eye in a young child and thereby seek treatment from prayer houses and alternative medical practitioners [9].

The mean age at initial diagnosis was 18 months in this study, and the vast majority of cases become clinically obvious before the age of 3 years. Patients with bilateral tumours presented earlier than those with unilateral involvement [10].

With a simple pen touch or direct ophthalmoscope, obvious retinoblastoma cancer can be easily diagnosed. However, an examination under anaesthesia and with the help of B-ultrasound Scan & Computer Tomography Scan is often needed and used in facilities where the instruments are readily available [11]. Extraocular extension of a retinoblastoma tumour significantly reduces the chances of treatment, recovery of sight and survival of the patient [12]. Patients with the overt extraocular extension of retinoblastoma are first treated with higher doses of radiotherapy and chemotherapy to achieve the reduction of the bulk of a tumour followed by subsequent enucleation.

2. MATERIALS AND METHODS

This was a prospective study of 120 retinoblastoma patients who were treated at ECWA Eye Hospital, Kano from January 2006 to December 2014. All the children diagnosed with retinoblastoma and their parent(s) consent was recruited into this study. Patients whose parents did not give consent for the study and patients who lost to follow up after the initial diagnosis were excluded. Simple touch light examination, the direct and binocular indirect ophthalmoscope were used for the diagnosis. In few of the cases diagnosis was done under anaesthesia. All the diagnoses were made by the same paediatric ophthalmologist. The measurement of visual acuity was done using the E-tumbling chart, the Kay-pictures and the Central, Steady and Maintained (CSM) system, corneal light reflex where appropriate for the age and intellectual ability of the patient.

The patients' ages, gender, relevant past medical and ocular history, family medical and ocular was recorded. Data were analysed with statistical package for social sciences (SPSS) version 20 (IBM Corp. Armonk, NY). Ethical approval for the study was obtained from the University of Port Harcourt Teaching Hospital Ethical Committee.

3. RESULTS

A total of 120 cases of retinoblastoma were studied. Seventy (58.3%) of them were males while 50 (41.7%) were females. Female: Male ratio was 1: 1.4. The age range of the study

population was 4 months to 7 years. The mean age of patients was 2.7 years (SD ±1.4 years). The difference in the age groups of participants in this study was not statistically significant (p=0.118).

The mean time that lapsed between the manifestation of symptoms and the presentation of patients in this study to a health care facility was 7.1 months (SD±11.97 months). The difference in the duration of symptoms before presentation to health care facility and the various forms of presentation of retinoblastoma in this study was statistically significant (p=0.016). The most common form of presentation was proptosis (61.7%). Other variants of presentation were cat eye reflex (26.6%) and fungating mass (11.7%).

There were 19(16%) cases of bilateral retinoblastoma, and 101 cases of unilateral neoplasms.

Table 1. Age and sex distribution of the study population

Age group	Sex		Total (%)
	F (%)	M (%)	
0-6 mths	2 (1.6)	4 (3.3)	6 (5.0)
>6-12 mths	7 (5.8)	3 (2.5)	10 (8.3)
>1-3 years	25 (20.8)	46 (38.3)	71 (59.2)
>3-6 years	13 (10.8)	15 (12.5)	28 (23.3)
>6 years	3 (2.5)	2 (1.6)	5 (4.2)
Total	50 (41.7)	70 (58.3)	120 (100)

Pearson Chi-Square Test = 7.396; p-value = 0.118

Table 2. Modes of presentation of retinoblastoma in the study population

Duration of symptoms before presentation	Form of presentation of retinoblastoma			Total (%)
	Cat eye reflex (%)	Proptosis (%)	Neglected retinoblastoma (Fungating Mass) (%)	
0-3 mths	18 (15.0)	35 (29.2)	5 (4.1)	58 (48.3)
>3-6 mths	10 (8.3)	16 (13.3)	4 (3.3)	30 (25.0)
>6-12 mths	3 (2.5)	17 (14.2)	3 (2.5)	23 (19.2)
>1-2 years	1(0.8)	3 (2.5)	1 (0.8)	5 (4.1)
>2 years	0 (0)	3 (2.5)	1 (0.8)	4 (3.3)
Total	32 (26.6)	74 (61.7)	14 (11.7)	120 (100)

Pearson Chi-Square Test = 21.395; p-value = 0.016

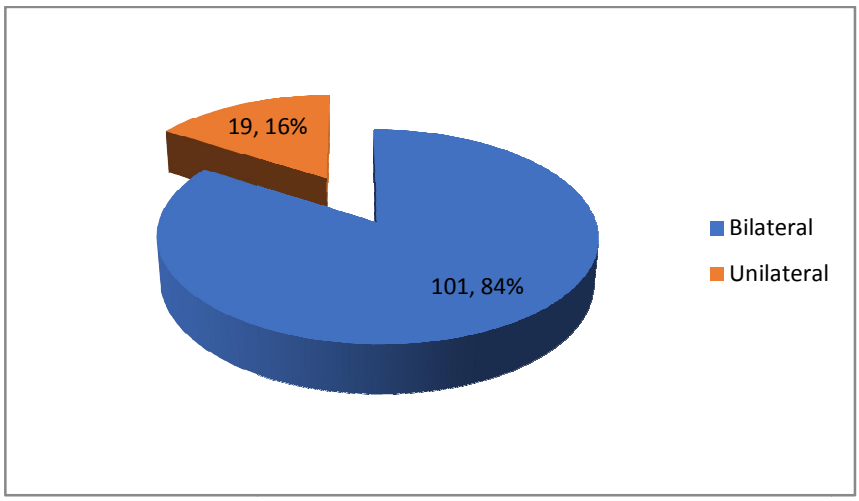

Fig. 1. Frequency of cases with unilateral and bilateral retinoblastoma involvement

4. DISCUSSION

In this study, the most common form of presentation of retinoblastoma was proptosis (61.7%). Other variants of presentation were leukocoria (white pupillary reflex or cat's eye reflex (26.6%) and fungating mass (11.7%) as shown in Table 2. This finding is similar to that of Arif and Islam [13], in Pakistan, who found that 47.3% of retinoblastoma cases were presented with proptosis and 30.14% cases were presented with leukocoria [13]. Also, this study is consistent with the work of Khan et al[14] who on clinical examinations, found that 53.8% patients were presented with proptosis 38.5% cases were presented with leukocoria while 5.8% with strabismus and 1.9% with painful red eye due to secondary glaucoma. This study had a lower yield of presenting patterns that the previous two studies [13,14] probably because the population of their studies had a greater number of participants. In a study by Balasubramanya et al. [15], the clinical presentation of retinoblastoma cases was different from the patients of this study, where a total of 392 cases of retinoblastoma reviewed; 72.2% had leukocoria, 13% had proptosis, 10% had strabismus, 1.5% were asymptomatic (detected on screening), 3.3% had a typical presentations and secondary glaucoma were 0.76%. These clinical findings are not consistent with the present work probably due to the difference in the geographical setting and difference in culture and level of awareness of the disease among the various populations.

In this study, the minimum age was 4months while the maximum was 7 years with mean age of 2.7±1.4 years. The difference in the age groups of participants in this study was not statistically significant (p=0.118). The maximum numbers of cases with retinoblastoma were of 1-3 years range (59.2%), while the minimum number of cases was in the age 6 years and above (4.2%), as shown in Table 1. Seventy cases (58.3%) were males while 50 (41.7%) were females. Female: Male ratio was 1: 1.4.

The findings of this study compared well with that of Khan et al. [14] in which the minimum age was 1 year while maximum age was 10 years (age range 1 - 10 years) with a mean age of 3.54 ± 1.686 years. The maximum numbers of cases with retinoblastoma eye were of 3 years (34.6%). This study is consistent with the study by Arif and Islam [13], Bhurgri et al. [16] and Saiju et al. [17] where predominance of males tumour burden was seen as compared to females. This study, however, differed from the study of Khan et al. [14], Nabie et al. [18] which noted the female preponderance of retinoblastoma with a male to female ratio of 1:1.5 and 1:1.4 respectively.

The study revealed that there were 16% cases of bilateral retinoblastoma and 84% of unilateral retinoblastoma Fig. 1. This finding slightly differed from that of Khan et al. [14] in which they noted 28.8% cases of bilateral retinoblastoma and 71.2% cases with unilateral retinoblastoma. This difference could be as a result of the difference in the number of cases studied. Also, Arif and Islam [13] found that 64.51% had unilateral tumours and 35.4% presented with bilateral disease. However, in a Chinese study by Zhao et al. [19], the frequency of bilateral retinoblastoma was greater than the present study. Bilateral retinoblastoma was present in 32% of the patients. These inconsistencies in the findings could be as a result of differences in the population studied especially when compared with the present results.

Researchers in different geographical regions of the world have noted the variations in the time of the presentation of retinoblastoma patients to healthcare facilities due to the differences in awareness of the disease among the various populations, the availability and accessibility of medical facilities and personnel and possibilities of obtaining satisfactory treatment [13,14,19]. In this study, the mean time lapse between the manifestation of symptoms and patients' presentation to the hospital was 7.1 months (SD±11.97 months) as shown in Table 2. The difference in the duration of symptoms in patients before their presentation to the health facility and the forms of presentation of retinoblastoma in this study was statistically significant. This late presentation to the hospital facility could be related to ignorance of the symptom of the disease on the parents' part, financial constraints and preference to alternative healthcare at prayer houses or traditional healers. This study was compared with the study of Abramson et al. [20] in Ohio, the United States of America who observed that the mean age of diagnosis is 12 months for bilateral tumours and 24 months for unilateral tumours. Also, in Malaysia, Jamalia et al. [21] observed a 3- 4 months duration of symptoms of retinoblastoma before presentation to a health facility. In Argentina, Navo et al. [22] noted that patients presented at older ages were reported from high-income countries [mean age 24 months (range, 0 to 165 months), 31 months for unilateral (range, 0 to 165 months), and 13.3

months (range, 0 to 62 months) for bilateral disease.

5. CONCLUSION

In ECWA Eye Hospital, Kano, Nigeria, retinoblastoma patients presented late, more frequently with advanced disease- the most familiar being proptosis. There is, therefore, the need for awareness campaign among the populace for early detection of the disease.

CONSENT

Prospectively 120 cases of retinoblastoma were studied after obtaining consents from their parents.

ETHICAL APPROVAL

Ethical approval for the study was obtained from the University of Port Harcourt Teaching Hospital Ethical Committee.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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