Case series

Clinical features and prognosis of retinoblastoma at the University Teaching Hospital of Yaounde Cameroon

Présentation clinique et pronostic du rétinoblastome au Centre Hospitalier et Universitaire de Yaoundé

Epée Emilienne[1,2], Moukouri Ernest[1,2], Koki Godefroy[2], Pondy Angèle[3,4], Mbassi K[2]

1) *University Hospital Center of Yaounde Cameroon*
2) *Department of Ophthalmology, Faculty of Medicine and Biomedical Sciences University of Yaoundé I Cameroon*
3) *Department of Pediatrics, Faculty of Medicine and Biomedical Sciences University of Yaoundé I Cameroon*
4) *Mother and Child Center of the Chantal Biya foundation(CME-FCB), Yaounde Cameroon*

- *Corresponding Author: Dr Epée Emilienne* epee 40@yahoo.fr, +237 99 54 90 41

ABSTRACT

PURPOSE / AI.: Retinoblastoma is the most common intraocular malignant tumor of childhood, an important cause of ocular morbidity and mortality. With early detection and new methods of management many affected children can survive, have the affected eyes saved and vision preserved. Nevertheless 90% of retinoblastoma patients live in developing countries with minimal hope for survival. In the current study, we aim at describing the natural history, clinical features, and management and outcome of patients with retinoblastoma at the University Teaching Hospital in Yaounde Cameroon

METHODS. From January 2004 to March 2011, we conducted a prospective and descriptive study including all patients with retinoblastoma received at the university teaching hospital. Variables studied included age of the patients at diagnosis, gender, history of the presenting complaints, mode of presentation, previous treatment, and time interval between diagnosis to surgery, diagnosis before surgery, management. Collected data were analyzed using Microsoft Excel 2007 software.

RESULTS. Eleven patients had retinoblastoma, 7 males and 4 females a sex ratio of 0.57. The median age at diagnosis was 43,6 ± 22,3 months . Leucocoria and exophthalmia were the common signs observed in 50% and 25% of the cases respectively. The median lag time was 38 ±67,7 days. Eight patients were operated and (6)75% underwent enucleation. Three patients received adjuvant chemotherapy. Death occurred in 6 cases (75%) within a year after surgery.

CONCLUSION. The prognosis of retinoblastoma remains poor due to late presentation. There was no change observed from the situation 10 years ago in all aspect of retinoblastoma in our hospital despite the advances in the field of retinoblastoma. Could education and raising awareness have helped in improving the prognosis?

KEY WORDS:
Retinoblastoma, childhood cancer, exophthalmia, leucocoria, enucleation,

RÉSUMÉ

OBJECTIFS. Le rétinoblastome est le cancer malin intraoculaire le plus fréquent de l'enfant. Grâce au dépistage précoce et de nouvelles méthodes de prise en charge de nombreux enfants atteints peuvent survivre, l'œil affecté sauvegardé et la vision préservée. Néanmoins, il est rapporté que 90%des patients atteints de rétinoblastome vivent dans les pays en développement avec l'espoir minime pour la survie. Dans cette étude, nous visons à décrire l'histoire naturelle, les caractéristiques cliniques, la prise en charge et les résultats obtenus des patients atteints de rétinoblastome au Centre Hospitalier et Universitaire de Yaoundé Cameroun et comparer la situation a un rapport antérieur il ya 10 ans.

MÉTHODES. De Janvier2004 à Mars 2011, nous avons mené une étude prospective et descriptive incluant tous les patients atteints de rétinoblastome reçus au Centre Hospitalier et Universitaire de Yaoundé. Les paramétres étudiés comprenaient l'âge des patients au moment du diagnostic, le sexe, les antécédents, motif de consultation, le mode de présentation, un traitement antérieur, l'examen du fond d'œil sous AG ; Les résultats de l'imagerie orbitaire (échographie, IRM ou TDM) ; les classifications ; la prise en charge en fonction des formes clinique et l'intervalle de temps entre le diagnostic à la chirurgie, le diagnostic avant la chirurgie, de la gestion. Les données recueillies ont été analysées en utilisant le logiciel Microsoft Excel 2007.

RÉSULTATS. Onze patients ont été recrutés, 7 garçonset4 filles. L'âge médian u diagnostic était de43,6 ± 22,3mois.La leucocorie et l'exophtalmie étaient les signes les plus courants,observés respectivement chez 50% et 25% des cas . Huit patients ont été opérés et 75%ont subi une énucléation. Trois patients ont reçu une chimiothérapie adjuvante. La mort est survenue dans 6 cas (75%) dans l'année suivant la chirurgie.

CONCLUSION. Le pronostic du rétinoblastome reste faible en raison de la présentation tardive. En effet il n'y a pas d'améliorations, comparé au rapport antérieur il ya 10 ans et ceci malgré les avancées dans le domaine du rétinoblastome.

MOTS CLÉS
Rétinoblastome, exophtalmie, leucocorie, énucléation.

INTRODUCTION

Retinoblastoma is the most common and severe intraocular malignant tumor of childhood. It is an important cause of ocular morbidity and mortality particularly in the underdeveloped countries of sub-Saharan Africa .It represents approximately 3 to 4% of all malignancies in children and 1% of human cancers. The RB gene is localized in the long arm of chromosome 13, 13q14 which is the suppressor of tumors. Its inhibition or inactivation causes RB to develop. They are 2 forms of retinoblastoma , an hereditary form with autosomal dominant inheritance with complete penetrance usually bilateral and an and non-hereditary form with 2 mutations on the retinal cells mostly unilateral presentation in 94%.[1]. With early detection and new methods of treatment many affected children can survive and have the affected eye saved and vision preserved[2].For the past decades the survival rate of retinoblastoma has been more than 80-90% in the developed world, specifically in the United States and in Europe 90% [3].Although the disease is clinically easy to diagnose late diagnosis and intervention constitute the hallmark of several series described in developing countries where the overall survival rates remain low [4, 5]. When treated inadequately and lately, retinoblastoma uniformly leads to death. Despite the fact that 90% of retinoblastoma patients live in developing countries, their hope for survival is minimal. Due to scarcity of publications and reports, very little attention has been given to them and therefore, their impact is not taken into account by systematic reviews. [6].Reviewing a study carried out 10- years ago in the University Hospital Center of Yaounde (CHUY) by Moukouri et al in 1994 [7],we sought to find out the current status of retinoblastoma in the same hospital a decade after with existing improved techniques and methods of management of the disease. In this study we aimed at describing the natural history, clinical features and management retinoblastoma in CHUY Cameroon.

MATERIALS AND METHODS

We conducted a prospective and descriptive study over a period of 7 years from the 16th of January 2004 to the 30th March 2011 in CHUY. Included in our study were all the patients consecutively with clinically diagnosed retinoblastoma during the study period. The variables studied included thorough, demographic characteristics of the patients age and sex, clinical features of retinoblastoma, histopathological findings, treatments used and outcome. Data obtained were recorded in a predesigned follow up research form.The patients underwent a complete physical and ophthalmological examination. As far as possible, after observation, palpation and assessment of visual acuity slit lamp bio microscopy for the anterior segment was attempted or performed. In each case and for each eye we assessed the intraocular lesions with a fundus examination using a 90D or a 78 D lens. In Indirect ophthalmoscopy we used a 20 D lens. We emphasized ondetailed fundus examination under full mydriasis obtained using Tropicamide eye drops 0.5% to assess the extent of the tumor and to rule out bilateral involvement. The diagnosis of retinoblastoma was made on the basis of leucocoria in the history expressed as cat's eye or shiny spot in the eye, with or without exophthalmia and infiltrated periorbital space. During examination the presence of retinal mass particularly chalky areas and blood vessels guided the diagnosis. Computed Tomography of the brain and orbit was used to examine the characteristic calcified densities within the orbit and the retinoblastoma mass. The tumor or mass was classified using Reese and Ellsworth classification [8] and international retinoblastoma classification (IRCB)[9]. The assessment of general state of the patient and the systemic examination were completed by a pediatrician, anesthetist and pediatric-oncologist with whom we collaborated. After reaching a consensus with the pediatrician and oncologist, with the written consent of the parent or guardian, surgery, chemotherapy and or radiotherapy were carried out. The surgery performed was either enucleation or exenteration depending on the spread of the mass. Enucleation and exenteration are ocular mutilating surgical procedures used in advanced cases of ocular diseases. In enucleation only the eyeball is taken out of the orbit; the common indications are intraocular tumor chronic inflammations. In exenteration, the orbits are emptied of their contents eyeball muscles including the lids. It can be partial or total the main difference being the partial or total removal of all the lids. The main indications being tumors, extensive spread. [10]

Examination of the fellow eye under anesthesia was done at this stage. The specimens were then taken to the laboratory for histological examination... Chemotherapy was done post operatively according to a protocol proposed by the pediatric oncologist. This, included a combination of Cyclophosphamide, Adriamycine, and Vincristine, The following protocol was applied cyclophosphamide (300mg/m^2 body surface in an isotonic solution given intravenously for one hour from day 1 to 5.adriamycine(300mg/m^2 body surface in an isotonic solution was given intravenously during 6 hours from day 4 to day 5. vincristine 1,5 to 2mg intravenous bolus on day1 and day 5.This regimen was administered in 2 to 3 cycles for 4 to 6 months depending on the response of each case. For each case a written informed consent was signed by parents or guardians for surgical intervention and the same for chemotherapy .The study was approved by the hospital administration who in addition delivered an ethical clearance. The results were compiled using software Microsoft Excel 2007 software. Descriptive statistics such as means and standard deviation were used to summarize quantitative variables.

RESULTS

From January 2004 to March 2010, 11 cases of retinoblastoma were admitted into our unit amongwhich7 males and 4 females, a sex ratio male to female of 0.57an incidence of 11 cases over 6 years, (1.83cases per year).

Age at diagnosis: A majority of patients5 (46%) were between the ages of 24 and 60 months, that is2 to 5 years. No patient was below 1 year of age at presentation. The mean age of the 11 cases was 43,6 ± 22,3 months , the median 36± 22months,theyoungest patient being 18 months the oldest one 7 years as shown in table 1 and 2**.**

Clinical presentation: A total of twelve eyes were examined. In 01 patient (9%) there was bilateral involvement, and unilateral in 10 (91%). Seven cases had the sporadic form of retinoblastoma .The only bilateral case did not give us contributive information concerning the family history of retinoblastoma. The most common complications encountered were local recurrences and whether under chemotherapy or not the patient came back for consultation with extensive diseases. Funduscopy was not contributory in all the immediate relatives we examined.

Among those with unilateral disease the left was affected in 07 cases (63%), and in the right eye in 03 cases (27%). Leucocoria was the main presenting sign in 6 eyes (50%) followed by exophthalmia in 3 eyes (25%).One patient had strabismus and another one hyphaema. At the time of management they were all at stage V of the Reese and Ellsworth classification or Group D and E of IRCB (international retinoblastoma classification [8].

Clinical history: By 75%, 9 eyes. The parents or guardians recalled having noted a whitish glow in the eye of the child at least once at night which disappeared the next day. In the absence of pain and discomfort medical care seemed unnecessary. The eye was not presented to specialized personnel.

Table 1: Demographic data of the 11 patients

Features	N (%)
Sex	
Males	4 (36%)
Females	7 (63%)
Age at diagnosis in years	
Birth to 1 year	0 (0)
1 yr to 2 years	3 (27)
2 to 5 years	5 (46)
After 5	3 (27)
Involved eye side	
Left	07 (63)
Right	03 (27)
Bilateral	01 (9)
Presenting complains/ sign (n=12)	
Leucocoria	5 (46)
Blurred vision	1 (9)
Strabismus	1 (9)
Exophthalmia	3 (27)
Hyphaema	1 (9)
Classification (IRCB 2005)	Group E 11 eyes Group D 1 eye

Figure 1 shows leucocoria or" cats eye" in the left eye of a child referred by pediatrician. The post segment is full of cancer cells. **Figure 2 shows** Fig 2 represents a case of recurrence 5 months after exenteration.

Figure 2: Extensive retinoblastoma of the left eye recurrence 5 months after exenteration

Diagnosis– surgery interval The median time interval was 38 days, the mean time 70.50 ±67.7 days the shortest being 10days and the longest186 days. In 3 cases 27%,the patient admitted to have seen at least 3 other care givers among which ophthalmologist but in addition traditional practionners for second opinion before deciding to comply, with the surgical management offered.

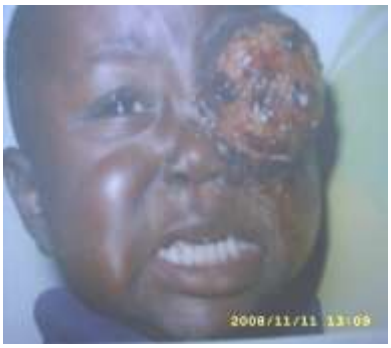

Figure 1: Leucocoria or" cats eye" in the LE of a child referred by pediatrician

Due to of financial constraints the screening for metastasis was not routinely performed for the patients before or after surgery.

Management: The management was surgical in 8 patients. Six (75%) out of the 8 patients had enucleation while 2 (25%) others had total exenteration. Three patients underwent adjuvant chemotherapy. Only one case had radiotherapy

With the presence of calcification, fleurettes and rosettes, the clinical diagnosis of retinoblastoma was confirmed by histology in all the 8 cases operated upon. The optic nerve was involved only in one case in the series.

Outcome*:* 2 patients with unilateral disease are alive and on follow up, 5 patients died despite surgical management and adjuvant treatment. Two of them after the first and fourth cure of chemotherapy respectively, one of them after radiotherapy3 patients who needed surgical intervention died before surgery. One of them got lost to follow up and the two others died before surgery.

DISCUSSION

The rate of retinoblastoma in our serieswas1.83 cases per year. Consistent with findings reported in Cameroon1994 [7], and others reports in Nigeria2010 [11] and in Taiwan2006 [12]. However higher incidences have been reported in Brazil in2004 [13]; in Algeria in 2002[14] and in Ghana in 2004 [15].The lack of a cancer centers and registers can explain the apparent low incidence .Erwenne et al [16] wrote that age specific incidence of retinoblastoma was higher in developing countries.

Sex: We found a predominance of girls in our series, with a sex ratio M/F of 0.57. This is not in line with others [4, 7, 13, 15] who reported a male predominance in their series. Neither gender nor racial predilection has been described or confirmed even in previous cumulative reports [17].

Age: The mean age at diagnosis was 41 months which is above the 38 months reported by Moukouri et al ten years ago [7]. Our findings are in line with those of other African studies were late consultation and delayed diagnosis were the hallmarks [5,15]. Our results are different from the 30.6 months by Bekibele et al in Nigeria [4], the 26,3 months, by Chang in Taiwan [12] and the 25months by Karla et al in Brazil [13], who mentioned that their cases present earlier for management. It was equally observed that the higher the mean age at diagnosis the higher the risk of advanced disease and metastasis [18].

Eight, (73%) of our patients were diagnosed retinoblastoma before the age of 5years. Delayed diagnosis is commonly encountered in developing countries with 90% of cases diagnosed before the age of 5 years [19].None of our patients was diagnosed before the age of one while in Brazil, 53% of her patients were seen between 0-24 months [13].

Given that the disease can be present at birth, diagnosis of tumors after months of evolution with anterior segment involvement is a risk factor of poor prognosis [14, 16]. Screening and surveillance are best ways of detecting retinoblastoma in its early stages [20]. According to Linn [9], patients with high risk of having inherited the Retinoblastoma RB1 mutation must have dilated retinal examination as early as possible.

Clinical presentation: Leucocoria also known as cat's eye was detected in50% of the cases. It remains the most common presenting sign of retinoblastoma reported in several studies in up to 89 percent [4 13 15].Exophthalmia came next in 3cases (27%) detected and strabismus in one case,(9 %) of cases. Chung in Taiwan observed squint in 14,3 % of cases less often than red eye. In Brazil Karla in addition noted that leucocoria was closely followed by strabismus such as in our series [13]. Ghana recorded a higher percentage of strabismus at presentation, (21,7%)[15],27% of our cases presented with tumor masses at advanced stages associated with proptosis. This finding is comparable to Essurams (34%) but higher than for the results of Brazil (3.4%) [13]. Essuram et al [15] reported when comparing several. Africans studies, that proptosis; a sign of late presentation is common in retinoblastoma compared to the developed world. Whether in the case of leucocoria or proptosis .Late presentation remains a major risk factor. Using both classifications Reese Ellsworth [08] and IRCB international Classification of retinoblastoma [09] the prognosis of these cases with late diagnosis is unfavorable. Leucocoria strabismus and exophthalmia are common signs of the disease. [21].

Fig 1 shows leucocoria or" cats eye" in the left eye. The posterior segment is full of cancer cells.

The mean **lag time** from diagnosis to surgery was 2.4 months comparable to the findings of Chang 2.59 -2.96 [12], but lower than those of Brazil and Taiwan5.8 months and 5.3months respectively. Nevertheless Karla [13] noted that for patients older than 2 years at diagnosis the lag time was longer than for the younger ones. In fact, it is directly influenced by advanced disease [12].

Diagnosis to surgery Interval None of our patients was diagnosed before one year of age, although 70% of parents or guardians recalled having noted the whitish glow in the eye of the child at least once. This occurred at night but since this was not clear enough it was neglected. Even in cases where it was mentioned at the next pediatric visits it did not raise interest.

The prognosis for survival is related to the age at diagnosis [22]; lack of knowledge and taboos on general beliefs about the tumors. In developing countries even in cases of early diagnosis the lag time before surgery has an impact on the prognosis only such as missed diagnosis is a reason for delay in treatment [23]. According to Chantada, early diagnosis of retinoblastoma is influenced by socioeconomic and maternal educational factors [20].

Management: Out of the operated cases, 6 patients (75%) had enucleation while 2 patients (25%) were exenterated. Enucleation versus exenteration depended on the stage of the disease; Yang [24] reported 76 % of enucleation. In the 80's as in our series surgery was the most common treatment advocated even in the United States [17]. In line with improved management protocol a chemo reduction is highly recommended before surgery now our days [25].The high incidence of patients requiring surgery in our series is explained by the late diagnosis and referral. The stages D and E of the disease at which all our patients were at the time of intervention determined the management. With IRCB, the bilateral cases of group D or E are managed by chemoreduction thermotherapy or low dose of external beam radiotherapy but E often requires enucleation [9].

Survival rate: The follow- up period was a year. Out of our 11 patients, 5 died within 3 weeks to 6 months, 2 others died within a year **(table 2).**Two cases were lost for follow up. The survival rate and the outcome of attempted treatment depend on the delay and denial of treatment only as observed by Chung [12].On the average 20% to 30% of patients abandon the treatment according to the results of a review by Canturk[26].

CONCLUSION

Retinoblastoma is still an enormous challenge for ophthalmologists; pediatricians and oncologists in Yaounde. Late diagnosis and increased lag time before surgery are frequent features of retinoblastoma. Leucocoria and exophthalmia are the most common signs of the disease at presentation. Despite availability of surgery and chemotherapy, due to of poor classification at presentation and increased lag time before surgery the prognosis remains poor. Substantial changes and advances in methods of management in the field of retinoblastoma have occurred but the overall outcome and patient behavior have not changed. Consequently an emphasis on interventions such as public awareness campaigns, physician and maternal education could be keys to the solutions of the retinoblastoma survival.

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Table 2: Features of 11 patients with retinoblastoma

N°	Age (y)/Sex	Eye	Interval diagnosis and surgery In days	Clinical presentation	Mass/ CT-scan Confirmation of diagnosis Spread of tumor	ICRB	Reese	Management	Outcome
1	2 /M	LE	93	Leukocoriamucopurulent discharge lid oedema	Intraoccular Homolateral optic nerv involvement	E	V	Enucleation +chemotherapy	Died of Pancytopenia after 2 course of chemotherapiy
2	6/F	LE	17	Painful red eye Strabismus Leucocoria at birth	Intraocular US retinal detachment CT-scan no on involvement	D	V	Enucleation	alive +prosthesis
3	3/M	RE	23	Red eye proptosis Necrotic vascularised tumor mass	Extraocular calcified mass 42 mm With normal optic nerve	E	V	Exenteration and Chemotherapy	Died of metastasis after 8months
4	2/F	RE	-	Leucocoria	Intraocular Ct – scan	C		No op	Loss for follow up
5	3/F	LE	10	Leucocoria Hyphaema Iris neovascularisation	USVitreous seeding and hemorrhage	E	V	Enucleation	Alive at last visit but reported dead by relatives 2years post op
6	4/F	LE	36	Exophthalmia	ExtraocularUSvithe CT scan on involvement	E	V	Enucleation + Radiotherapy	Died after radiotherapy
7	7/M	LE	-	Proptosis extensive Necrotic mass	ExtraocularExophytiq mass on involvement	E	V	Planned exenteration	Died before surgery
8	3/F	RE	157	Proptosis+leucocoria	Extraocular	E	V	Exenteration	Died 2weeks post op
9	2/F	LE	42	Leucocoria	Intraocular	E	V	Enucleation	Alive
10	6/M	LE	186	Leukokoria	Intraocular Endophy	D	V	Enucleation-	Died of undefined anaemia
11	2/F	LE	-	Leukocoria	Intraocular	E	V	No surgery no chemotherapy	Lost for follow up
11' *		RE	-	Leukocoria and Strabismus	US+CT Intraocular US+CT	E	V		

***Case number 11 was bilateral**