



Case report

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# Difficulty of Management of Retinoblastoma about Two Cases Report in North Benin and Literature Review



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## Abstract

Retinoblastoma is a highly malignant tumor and is the most common intraocular tumor in children. We make two cases report, the first one is about a 3-year-old female of ethnicity Peulh from Beninese origin who has been consulted for an intraocular tumor of the left eye, who is diagnosed retinoblastoma complicated by purulent melt. The symptomatology would have begun around the age of 2 and half years by a leucocoria of left eye associated 3 months later with an exotropia that required multiple treatments by traditional care givers before reference to hospital. The second case concerned a male of 3 years, ethnic of Beninese origin, who consulted for leucocoria of the left eye. The ophthalmological examination in the left eye noticed in the fundus examination a whitish mass superior to 5mm height invading the vitreous cavity. As concerned the right eye, the anterior segment and the fundus examination were normal. The diagnosis evocated is a retinoblastoma in its endophytic form of the left eye. We made literature review about the management of retinoblastoma and difficulties of this management in low income countries.

**Keywords:** Retinoblastoma; Leucocoria; Strabismus; Intraocular tumor; North Benin

## Introduction

Retinoblastoma is a highly malignant tumor and is the most common intraocular tumor in children. It is an embryonic retinal cancer occurring in very young children. It represents 3 to 4% of pediatric cancers in France [1]. In Canada, the disease is diagnosed in approximately 23 children per year [2] and against that affects approximately 40 children in Germany per year [3].

It is a complex tumor triggered by a genetic mutation in one or more cells in the retina and is initiated by mutation of the RB1 gene. The tumor may be unilateral or bilateral and can be inherited. Overall survival, eye salvage, and preservation of vision are largely dependent on the stage of disease at presentation. Despite a recently enhanced understanding of the etiology of retinoblastoma, the mortality associated with it remains high worldwide [4]. The RB is unilateral in 60% of cases and bilateral in 40% of cases. In developed countries, most children are diagnosed early with localized intraocular disease, and the overall survival rate exceeds 95% [3]. Its treatment in early stages holds a good prognosis for survival and salvage of

visual function. In very late stages, however, the prognosis for ocular function and even survival is jeopardized [5].

## Observations

### Case 1

We report the case of a 3 years female of ethnicity Peulh from Beninese origin who has been consulted for an intraocular tumor of the left eye, for whom we diagnosed retinoblastoma complicated by purulent melt. The symptomatology would have begun around the age of 2 and half years by a leucocoria of the left eye associated 3 months later with an exotropia that required multiple treatments by traditional care givers. The progressive installation of a left exophthalmos motivated consultations in several health centers including the Ophthalmological Hospital Saint André de Tinré (OHSAT) and from there, the child was referred to our ophthalmology unity at Centre Hospitalier Universitaire Departemental (CHUD) du Borgou et Alibori. There was no familial history of ocular tumors; medical and surgical histories have no peculiarities.

The ophthalmological examination finds

- a. In the left eye, an extra-orbital rounded tumor resting on the cheek, of about 5cm of diameter bleeding on contact and smelling nauseous with rupture of the ocular globe, purulent discharge.
- b. In the right eye, after dilation the anterior segment and the fundus of the eye are without particularities. Locoregional examination has not found any abnormalities.

We did not carry out an assessment of extension in the face due to the financial difficulties of the family and the unavailability of medical imagery in the center of care. There is neither possibility of oncological treatment nor radiotherapy in the center (Figure 1).



Figure 1: Left eye retinoblastoma complicated by purulent melt.

**Case 2**

J.P. a male of 3 years, ethnic of Beninese origin, consulted for leukocoria of the left eye. The ophthalmological examination in the left eye noticed in the fundus examination a whitish mass >5mm invading the vitreous cavity. As concerned the right eye, the anterior segment and the fundus examination are normal. The diagnosis evocated is a retinoblastoma in its endophytic form of the left eye. With clear consent of parents, we proposed and performed an enucleation of the left eye under general anesthesia. The control at one month and three months showed a clean orbital cavity. The Patient was reviewed 4 months after the last follow up, which means 7 months after enucleation for painful left orbital tumor. The right eye fundus examination was normal (Figure 2).



Figure 2: Left eye retinoblastoma complicated by recurrence after enucleation

**Discussion and Literature Review**

**Frequency**

The incidence of retinoblastoma is high in Africa according to Bekibele et al. [6] and Zadnas et al. [7]. The early diagnosis of retinoblastoma is essential for its management, which is the cornerstone of a good functional and vital prognosis [8].

**Date of diagnosis**

The average age of discovery of the disease is variable. In industrialized countries the discovery is done before 3 years (50% of cases), and before 5 years (90%) according to Shields et al. [9]. For other authors, retinoblastoma is a cancer of the very young; two-thirds are diagnosed before 2 years of age and 95% before 5 years [10].

In Asia the discovery is made at 33.4 months reported Bhurgri et al. [11]. But against to these studies, Sow et al. [12] in Senegal reported that the discovery was done at 9 months. Other African studies reported 33.1 months as the date of discovery [6,7].

**Sex**

Against that, Finger et al. [13] reported that there is no gender preference in their series, but there is significant male predominance with sex ratio (M/F) of 1.6 according to Chebbi et al. [14].

**Presenting signs**

In most of the cases, the two major presenting signs are leukocoria and strabismus, but other ocular or general signs may be observed [5]. In the series of El Zomor et al. [15] the most common clinical presentation was leukocoria in 180 (73.8%) patients, strabismus in 32 (13.1%) patients and decreased visual acuity in 12 (4.9%) patients. Group D and E disease represented 62% of all affected eyes.

**Laterality**

Worldwide, researches showed a very great variability in the localization of the tumor. In their series, nineteen children had unilateral lesions (22.8%), and 64 (77.1%) had bilateral lesions. In Bamako, Mali republic, only 11% were bilateral versus 35% in France [16]. Similarly, unilateral form represented 79.7% according to Sow et al. [12]. In Egypt, one hundred thirty-nine (57%) patients presented with unilateral disease, while 105 (43%) suffered bilateral disease [15]. Against that, Shields et al. [9] reported that all of their 26 patients (100%) had unilateral sporadic retinoblastoma.

**Treatment**

In most developing countries with low income retinoblastoma appear to be a problematic malignancy of childhood associated with management problems often related to difficulty with patients accepting removal of the affected eye and financial constraint for treatment [6].

For many authors, systematic enucleation has been the starting point of true and structured management of the disease [8]. This last decade, new types of chemotherapy have shown spectacular results and are currently under study, one of them is the chemoreduction to make large tumors more manageable and enable less aggressive treatment of tumors located in delicate sites [8].

If the diagnosis of retinoblastoma is not established early it evolves towards the complicated form. Exophthalmos and tumor externalization are unfortunately the first manifestations of consultations in our regions. Chebbi et al. [14] found 10% of complicated form (exophthalmos). The primary goal of treatment is the survival of the child. The safeguarding of the eye, the preservation of vision and the achievement of good aesthetic results are also important objectives. In our region, if the disease is diagnosed very early, we performed enucleation only for the survival of the child. We didn't have any possibility of chemotherapy and radiotherapy.

In Bamako, cure rate was around 50%, but it is estimated only on the cases arriving in Bamako and with at least 20% lost of follow-up, against that the cure rate is over 95% in France within an exhaustive register [16]. In low income countries, Problems faced by the management of retinoblastoma included financial constraint regarding investigations and procurement of drugs, as well as availability of the chemotherapy [6].

### Prognosis

The overall cancer prognosis is based on the American Joint Committee on Cancer and the International Union against Cancer's Stage Classification System (TNM) Classification and determination of the severity of the disease requires examination under general anesthesia (EUGA) with relevant imaging at the same time. The natural evolution of the retinoblastoma is usually towards the complications and the death by locoregional and metastatic extension.

The survival rate is higher in familial forms due to earlier screening and diagnosis allowing a delay in early management. In industrial countries like France, Global survival is 98.5% [17] and exceeds 95% in Germany [3]. But in most low income countries like Benin Republic, the diagnosis is done in very late stages and then, the prognosis for ocular function and even survival is jeopardized [5].

### Conclusion

Like the case in many low income countries, our cases report show that retinoblastoma is a problematic malignancy of childhood associated with management problems often related to difficulty with patients accepting removal of the affected eye and financial constraint for treatment. Early diagnosis is essential

for better management and better prognosis of this disease. The education of health workers, parents for recognition of evocative signs (white pupils, strabismus) is essential for early diagnosis.

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