

# Retinoblastoma Revealed by the Symptomatology of Orbital Cellulitis in a Little Girl of 02 Years at the University Hospital Center of Lomé

Kassoula Batomaguella Nonon Saa<sup>1</sup>, Fokam Olive Kamgno<sup>1</sup>, \*Dadjo Amouzou<sup>1</sup>, Nidain Maneh<sup>1</sup>, Diori Adam Nouhou<sup>1</sup>, Kodjo Gamele<sup>1</sup>, Koffi Didier Ayena<sup>2</sup> and Komi Balo<sup>1</sup>

<sup>1</sup>Department of Ophthalmology, Faculty of Health Sciences, University of Lomé (Togo)

<sup>2</sup>Department of Ophthalmology of Bè secondary Hospital, Lomé (Togo)

\*Auteur correspondant: Amouzou Dadjo/E-mail: dadjooson@gmail.com

**Received:** August 25, 2019

**Accepted:** September 27, 2019

**Published:** October 10, 2019

## Abstract

**Objective:** Report the clinical case of retinoblastoma with unusual symptomatology. **Observation:** This was a unilateral retinoblastoma manifested by the characteristic symptoms of orbital cellulitis in a two-year-old girl. A first hospitalization in paediatric emergencies with standard management had allowed the regression of the infectious signs, thus allowing a easier examination of the patient and revealing a leucocoria suggestive of a retinoblastoma, confirmed by an orbito-cerebral scan. **Conclusion:** Retinoblastoma, which is easy to diagnose in its classic presentation, should be mentioned when we are in front of any orbital and / or periorbital inflammation of the child.

**Key words:** Orbital cellulitis, Leucocoria, Retinoblastoma.

## Résumé

**Objectif:** rapporter le cas clinique d'un rétinoblastome d'une symptomatologie peu habituelle. **Observation:** Il s'agissait d'un rétinoblastome unilatéral se manifestant par les symptômes caractéristiques d'une cellulite orbitaire chez une fillette de deux ans. Une hospitalisation aux urgences pédiatriques avec prise en charge classique avait permis la régression des signes infectieux, autorisant ainsi un examen plus aisé de la patiente et qui révélait une leucocorie évocatrice d'un rétinoblastome confirmé par un scanner orbito-cérébrale. **Conclusion:** Le rétinoblastome, de diagnostic aisé dans sa présentation classique, devrait être évoqué devant toute inflammation orbitaire et /ou périorbitaire de l'enfant.

**Mots clés:** cellulite orbitaire, leucocorie, rétinoblastome.

## Introduction

Retinoblastoma is a malignant tumor developed from cone cells bloodline. It is the most common primitive intraocular malignant tumor of the child. Nevertheless it is a rare tumor whose incidence is between 1/15000 and 1/20000 of births in France [1], 1/24000 in Tunisia [2]. In sub-Saharan Africa it is the second most common tumor among childhood cancers [3, 4]. In 90% of cases, these are sporadic forms and only 10% of familial forms [1]. It is more often revealed by leucocoria or strabismus; and sometimes, it is revealed like uveitis presentation, spontaneous hyphema, neovascular glaucoma, preseptal cellulitis, proptosis, pseudohypopion or bulbar ptyse [5, 6, 7]. Orbital cellulitis is an uncommon form of revelation of retinoblastoma [8]. We report the case of unilateral retinoblastoma in a two-year-old girl with clinical signs that suggest orbital cellulitis.

## Clinical observation

A girl of 02 years is taken in consultation, in the Department Ophthalmology of Sylvanus Olympio (University Hospital Center of Lomé), by her parents for swelling and redness of the eyelids with

total closure of the right eye evolving since 03 days. She is the third child on a sibling of 03, the two oldest being apparently healthy. During pregnancy, everything was normal and she was born at term according to the mother and no past history of familial eye tumor could be found during the interview. Three days ago, the child presented in the right eye purulent secretions, swelling of the eyelids evolving to their closure, associated with fever unsuccessfully treated with paracetamol as self-medication. On examination, the child had fever and general condition altered. Ophthalmologic examination of the right eye noted a red, hot, hard and painful swelling extended to both the upper and lower eyelids and to the outer part of the orbit, with impossible palpebral opening (Figure 1).

Slit Lamp and Fundoscopic examination of this eye was impossible. In the left eye, there was good ocular tracking with a normal anterior segment; the fundoscopic examination could not be performed because of the painful context. The diagnosis of orbital cellulitis was evoked. Complementary exams requested included an orbito-cerebral computed tomography (CT), the cells blood count (CBC), C Reactive Protein (CRP), and an otolaryngologist consultation was asked for etiological check-up. The child was hospitalized in paediatric emergencies, where parenteral triple antibiotic therapy made with ceftriazone, metronidazole and gentamicin were done, and intravenous antipyretic and local care was instituted. Intravenous corticosteroid therapy was introduced 48 hours of antibiotic treatment onset.

### **Clinical evolution**

The clinical course was marked by the regression of the infectious and inflammatory signs thus making ophthalmological examination easier, allowing us to find: a residual soft palpebral edema with good palpebral opening, a mydriasis, a leucocoria, a non-pulsatile, non axile reducible exophthalmia. We then evoked a retinoblastoma. This diagnosis was later confirmed by CT findings presenting: heterogeneous intraocular formation of the right containing calcifications, infiltration of fat, thickening of the external rectus muscle and narrowing of the optic channel. The left eye was without abnormality (Figure 2). The results of biological analysis were: hypochromic microcytic anemia at 10.3 g/dl of hemoglobine, leukocytosis with 12,000 cells /mm<sup>3</sup> at the CBC; CRP elevated to 28.65 mg/l. Thus, the diagnosis of retinoblastoma, group E according to the IRC classification (International Retinoblastoma Classification) had been made and the patient referred for adequate management in the Ophtho- paediatric and Onco-paediatric Units.

### **Discussion**

#### **The Age of Discovery**

Earlier is the age of discovery; better is the vital and visual prognosis [2]. The median age of discovery is 2 years for unilateral forms and 1 year for bilateral forms [1]. This age in our case was 24 months or 2 years and the clinical stage was advanced. Some authors found an age lower to our [5, 6, 9]. Indeed Sachdeva *et al.*, 2011 [6] reported a case of unilateral retinoblastoma revealed by orbital cellulitis' symptomatology in a child of 13 months, at undetectable stage on CT but visible on the ocular ultrasound mode B. In the same way, Das *et al.*, 2016 [9] in India reported a case of retinoblastoma in 18 month's boy with the same clinical picture and an advanced retinoblastoma like our revealed by CT. This difference in age of discovery could be explained by the ability of parents to consult fast in an appropriate service and the disparities in the medicalization of populations from one country to another. On the other hand, other authors found an age higher than our [2, 10] like Chebbi *et al.*, 2014 [2] on his study on the clinical profile of retinoblastoma in Tunisia. They found an average age of 34.2 months for unilateral forms. The fact that this study involved a large sample (200 children) over a period of 06 years could justify the age of discovery superior to ours. Nalci *et al.*, 2017 [10] reported two cases of unilateral retinoblastoma with orbital cellulitis' symptomatology in two children aged respectively 3 and 4 years with an advanced clinical stage. It is established that in well-developed countries with good sanitary system, the age of discovery is earlier than that of under-medicalized developing countries. In addition, there is a routine screening for familial forms, but there is no yet a consensus on what to do in sporadic forms like our case for early diagnosis.

### **Discovery Circumstances**

Retinoblastoma is more often revealed by leucocoria (> 50%) or strabismus (16%) [1, 2, 7]. Although orbital cellulitis is a rare mode of discovery in developed countries, it's still common in developing countries because of late diagnosis [11], with prevalence oscillating between 1.3% and 4.8% [12, 13]. Menon *et al.*, 2009 [7], when they study the late-onset retinoblastoma presentations in Malaysia, found orbital cellulitis as a discovery mode's in 3% of cases. This study enrolled 105 children. This result is lower than those of Walinkjar *et al.*, 2013 [5] in India and Chebbi *et al.*, 2014 [2] in Tunisia, which found respectively 5.4% and 4.5%. These small differences could be explained by the disparity in sample sizes. The high incidence of retinoblastoma manifesting as orbital cellulitis in developing countries is linked to the fact that the parents don't bring the child fast to health services and / or they refuse to adhere to prescribe treatments [7, 10], or because of under-medicalization of the country [2].

Orbital cellulitis as a circumstance of discovery is often associated with advanced tumor. This is an immune response induced by tumor necrosis [5]. In our report case, orbital cellulitis was the mode of revelation and associated with advanced stage of the tumor according to the results of the scan. Similarly, Walinkjar *et al.*, 2013 [5] in their study in 2013 found 14 cases with this mode of discovery and all at advanced stages with invasion of 80 - 100% of the intraocular space. Das *et al.*, 2016 [9] reported the case of an 18-month-old boy presenting with orbital cellulitis associated with an intraocular calcified mass in the posterior segment and nerve thickening in favour of advanced retinoblastoma in the CT scan. However, Chebbi *et al.*, 2014 [2] in their study, conducted during 16 years on patients followed and treated for retinoblastoma, they had found 9 cases with orbital cellulitis' symptomatology including of 2 extra ocular advanced forms [2].

This mode of revelation is therefore associated with a poor visual prognosis because the globe can no longer be conserved [5, 9, 10]. Thus, orbital cellulitis revealing a retinoblastoma is most often correlated to an advanced tumor with a poor vital and visual prognosis.

### **Regarding laterality**

The tumor is unilateral in 60% of cases and non-hereditary and sporadic; in 40% of cases it is bilateral and always hereditary [1, 2]. Walinkjar and al., [5] in a retrospective study including 260 children with retinoblastoma were interested in those with orbital cellulitis as mode of disclosure. They had identified 14 of which 10 were the subject of their study. Of these 10 cases, they noted 6 cases of bilateral form and 4 cases of unilateral form (all interesting the right eyes). In our case, retinoblastoma was unilateral and involved the right eye too. This is similar to the case of unilateral retinoblastoma in the right eye reported by Das *et al.*, 2016 [9]. Nalci *et al.*, 2017 [10] reported two cases of unilateral retinoblastoma involving the left eye in one and the right to the other. There is no predilection for the laterality right or left in the occurrence of retinoblastoma, or impact on the evolution of the affected eye. Sporadic unilateral forms are associated with higher severity and poor visual prognosis compared to familial bilateral forms.

### **Sporadic or hereditary case?**

We haven't found a family history of ocular tumor, and in front of the unilateral character, we could evoked a sporadic form; however, only long-term follow-up and a genetic study could confirm this. In fact, Chebbi *et al.*, 2014 [2] found bilateralization in 3.2% of cases after an average follow-up of 16.2 months. This allows us to suggest a long-term follow-up in unilateral cases as in our young patient.

### **Associated signs**

Infectious signs (fever) and local inflammation, confirmed by a blood test were associated to tumor in our patient case. It was an orbital cellulitis. This symptomatology had been reported by several authors [5, 6, 9], this demonstrates that we are not in front of an isolated case. Thus, orbital

inflammation can be the mode of revelation of a retinoblastoma and it is therefore necessary to know how to evoke it.

### **The prognosis**

Leucocoria, exophthalmia and orbital cellulitis associated with retinoblastoma are correlated with severe advanced forms and poor visual prognosis because enucleation has to be done [2]. Life-threatening are function of early diagnosis, rapid and adequate treatment [14]. Without treatment, the evolution is towards complications and death due to metastasis [2]. Some authors have observed a good vital prognosis, without recurrence or new tumor, for cases diagnosed at an advanced stage but whose appropriate management had been well conducted: 01 year for Das *et al.*, 2016 [9], 16 years for Nalci *et al.*, 2017 [10]. Our child had a poor visual prognosis based on the stage at the time of diagnosis, because the globe can no longer be preserved.

### **Conclusion**

Easily diagnosed in its classic presentation, retinoblastoma should be evoked in presence of any orbital and/or periorbital inflammation signs of the child. Although ultrasound and computed tomography contribute a lot to diagnosis; however, a good clinical exam remains the standard.



**Figure 1. Peri-orbital and orbital inflammatory swelling of superior and inferior eyelid: orbital cellulitis masking retinoblastoma in two-years-old girl**



**Figure 2 a). CT scan without contrast injection**



**Figure 2 b). CT scan with contrast injection: Heterogenous formation with calcifications within it (blue arrow) in the right intra-ocular space, fat infiltration and thickening of the external right muscle, retraction of optical channel: first evoking right eye retinoblastoma**

**Conflict of Interest:** The authors declare no conflict of interest.

**Contributions of the authors:** All cited authors participated in the writing and/or the correction of this manuscript of which they have approved the final version.

**Ethics:** parents have given an oral consent for using information's about their girl for scientific purposes.

## References

1. Zucker, J.M., Desjardins, L., Stoppa-Lyonnet, D. and Doz, F. 2005. Rétinoblastome. EMC-Pédiatrie, 2(4): 322-331.
2. Chebbi, A., Bouguila, H., Boussaid, S., Ben, N.A., Zgholi, H., Malek, I., ... & Nacef, L. 2014. Clinical features of retinoblastoma in Tunisia. Journal francais d'ophtalmologie, 37(6): 442-448.
3. Togo, B., Traore, F., Togo, A.P., Togo, P., Diakite, A.A., Traore, B., ... & Sylla, F. 2014. Epidemiology and prognosis of childhood cancers at Gabriel-Toure Teaching Hospital (Bamako, Mali). Medecine et sante tropicales, 24(1): 68-72.
4. Darre, T., Kpatcha, T.M., Bagny, A., Maneh, N., Gnandi-Piou, F., Tchangai, B., ... & Napo-Koura, G. 2017. Descriptive Epidemiology of Cancers in Togo from 2009 to 2016. Asian Pacific Journal of Cancer Prevention: APJCP, 18(12): 3407-3411.
5. Walinjkar, J., Krishnakumar, S., Gopal, L., Ramesh, A. and Khetan, V. 2013. Retinoblastoma presenting with orbital cellulitis. Journal of American Association for Pediatric Ophthalmology and Strabismus, 17(3): 282-286.
6. Sachdeva, R., Schoenfield, L., Marcotty, A. and Singh, A.D. 2011. Retinoblastoma with autoinfarction presenting as orbital cellulitis. Journal of American Association for Pediatric Ophthalmology and Strabismus, 15(3): 302-304.
7. Menon, B.S., Alagaratnam, J., Juraida, E., Mohamed, M., Ibrahim, H. and Naing, N.N. 2009. Late presentation of retinoblastoma in Malaysia. Pediatric Blood and Cancer, 52(2): 215-217.
8. Ellsworth, R.M. 1967. The practical management of retinoblastoma. Transactions of the American Ophthalmological Society, 67: 462-534.
9. Das, A., Singh, U. and Bansal, D. 2016. Retinoblastoma mimicking orbital cellulitis. Indian Pediatr, 53(11): 1037.

10. Nalci, H., Gündüz, K. and Erden, E. 2018. Necrotic intraocular retinoblastoma associated with orbital cellulitis. *Survey of Ophthalmology*, 63(1): 114-118.
11. Zomahoun B. 1992. Contribution à l'Etude du rétinoblastome dans les services d'Ophthalmologie du C.H.U De cocody (à Propos de 19 cas). *Mém. CES Ophthalmology*, 70p.
12. Shields, J.A., Shields, C.L., Suvarnamani, C., Schroeder, R.P. and DePotter, P. 1991. Retinoblastoma manifesting as orbital cellulitis. *American Journal of Ophthalmology*, 112(4): 442-449.
13. Mullaney, P.B., Karciglu, Z.A., Huaman, A.M. and Al-Mesfer, S. 1998. Retinoblastoma associated orbital cellulitis. *British Journal of Ophthalmology*, 82(5): 517-521.
14. Goddard, A.G., Kingston, J.E. and Hungerford, J.L. 1999. Delay in diagnosis of retinoblastoma: risk factors and treatment outcome. *British Journal of Ophthalmology*, 83(12): 1320-1323.

**Citation:** Kassoula Batomaguella Nonon Saa, Fokam Olive Kamgno, Dadjo Amouzou, Nidain Maneh, Diori Adam Nouhou, Kodjo Gamele, Koffi Didier Ayena and Komi Balo. 2019. Retinoblastoma Revealed by the Symptomatology of Orbital Cellulitis in a Little Girl of 02 Years at the University Hospital Center of Lomé. *International Journal of Recent Innovations in Medicine and Clinical Research*, 1(2): 1-6.

**Copyright:** This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited. **Copyright©2019;** Kassoula Batomaguella Nonon Saa *et al.*, (2019).