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RESEARCH ARTICLE

TRILATERAL RETINOBLASTOMA: A CASE REPORT

***[1]MAKITA, Chantal, [1]NGANGA NGABOU Géraud Charles Freddy, [2]NSONDE MALANDA Judith, [3]MASSAMBA Alphonse**

[1]Ophtalmology Department, Teaching University Hospital of Brazzaville, Congo
[2]Cancer and Radiotherapy Department, Teaching University Hospital of Brazzaville, Congo
[3]Biostatistics and Biosciences Department, Brazzaville Institute of Health and Sports,Marien NGOUABI University, Brazzaville-Congo

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ABSTRACT

We report the case of a 2-year-old child with no history especially access the service at the age of one month for intraocular retinoblastoma unilat right. Then eighten months later there was a location in the left eye. The patient is seen 20 months later with large bilateral tumors of both orbits extending towards the zeugmatic area on right and the front left area. Oculars structures are covered with two larges chemosis. The objective brain scan large tumor in the brain. The treatment consisted of chemotherapy with Vincristine-Adriblastine-Cyclophosphamide-Etoposide protocol. After sixth treatment we found a significant decrease in tumor volume. Then child is lost.

INTRODUCTION

Presentation

Retinoblastoma is a highly malignant tumor and the most common intraocular tumor in children (El Kettani, 2014). The diagnosis is essentially clinical, the prognosis is excellent in developed countries where the survival is of the order of 95% (Lumbroso-Le Rouic, 2008 and Balmer, 2008). In the Congo and in the developing countries it is diagnosed very late, in a highly evolved form with high mortality (NsondéMalanda, 2011 and Sow, 2014). Trilateral retinoblastoma is rare, in Burkina three cases had been reported. It is defined as an association of an intraocular neuroblastic tumor and an inherited or bilateral hereditary retinoblastoma most often, sometimes sporadic (Blach, 1999). The tumor of the brain is considered a primary tumor and not a metastasis (Zografos, 2002). We report a case.

Medical History

2-year-old male infant, with no specific history according to parents, is seen at 1 month of age in the leukocoria department, ophthalmic examination with fundus revealed unilateral retinoblastoma right intraocular stage.

***Corresponding author: MAKITA, Chantal,**
Ophtalmology Department, Teaching University Hospital of Brazzaville, Congo

The pediatric examination was normal. A B-mode echocardiography and an orbito-cerebral tomodensitometry had been proposed to supplement the clinical examination not performed by the parents for lack of financial means and even asked for the child's foolishness. Then 6 months later, according to the parents they found another location of the intraocular tumor in the left eye. The patient returned 20 months later with a voluminous bilateral tumor of both orbits, extending to the zygomatic region on the right and the frontal region on the left, thus exceeding the Reese-Elsworth stage V. The ocular structures are inaccessible and covered by two large chemosis (Figure 1). The cerebral CT scan described a large tumor of 40 X 30 mm in the brain (Figure 2).

Workup and Treatment

The therapeutic management carried out in the carcinology department consisted of chemotherapy with the Vincristine-Adriblastine-Cyclophosphamide-Etoposide protocol in 6 courses for 3 months. In the first month, the patient received three courses of treatment two weeks apart under the control of a blood count, the second month two courses spaced 3 weeks apart and the last month a single course. We have seen a clear regression of the tumor from the first treatment (Figure 3) and this gradually until the 5th cure (Figure 4). Then the child is lost sight of.

Figure 1. Bilateral retinoblastoma

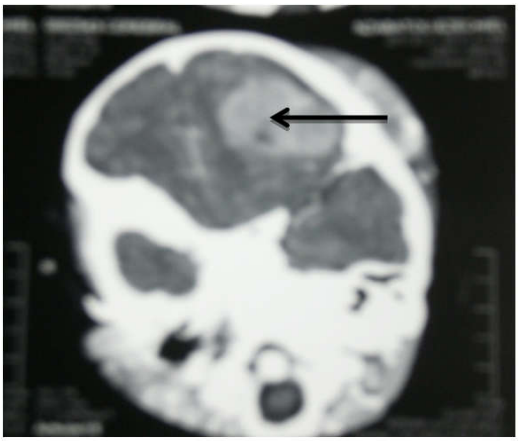

Figure 2. Cerebral computed tomography with brain tumor

Figure 3. After the 1st chemotherapy treatment

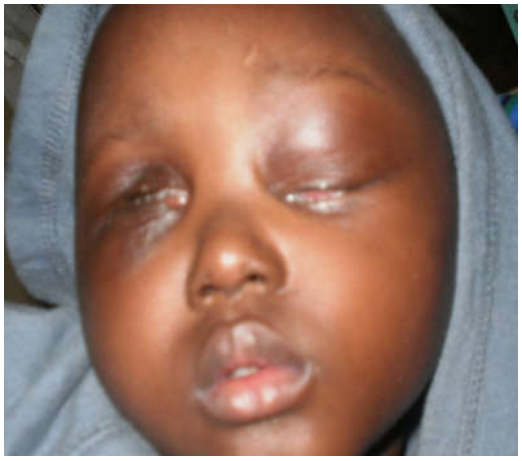

Figure 4. After the 5th chemotherapy treatment

DISCUSSION

The present clinical case shows the existence of this retinoblastoma entity in our country made of the association of the tumor of the brain that appeared 20 months after the discovery of bilateral retinoblastoma. This corroborates with data from the literature on the average time to onset of brain injury that is at least 21 months for KIVELA (Kivëla, 1999). The pathophysiology of this syndrome is not well known. It has been suggested that the intracranial tumor derives from pinealoblasts. Marcus et al. (Marcus, 1998) notes another origin that of the germinal strain of primitive neuroblasts under ependymaires not intended for a pineal differentiation. It could therefore be considered as a primary neuroectodermal tumor.

CT and MRI are the only preferred complementary examinations of retinoblastoma. They have the advantage of locating the intraocular tumoral lesions to analyze the locoregional extension and to evaluate a possible intracranial localization of the tumor. They also allow evolutionary follow-up (Nikièma, 2009). But these exams are not easy to access because of the very high cost in our countries. Therapeutic management required the intervention of oncologists where a chemotherapy treatment was instituted and very beneficial for our case. Namely that high cost of drugs and side effects are constraints to further treatment. Some authors believe that chemotherapy associated with local treatments gives good results on tumor control and ocular conservation given the sensitivity of this tumor (El Kettani, 2014; Balmer, 2008; NsondéMalanda, 2011; Sow, 2014; Blach, 1999 and Zografos, 2002). But the question remains to ask what about the cerebral involvement? We could not perform a second scan to evaluate the tumor regression at the cerebral level. According to the literature, involvement of the central nervous system suggests a fatal event (Gündüz, 2006).

Conclusion

Trilateral retinoblastoma is rare but present in our country. It is an expensive tumor of diagnosis and dark prognosis which poses a therapeutic challenge.

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