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Case Report

CASE REPORT

Retinoblastoma in children: a case series from Senegal

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Abstract

Retinoblastoma is the most common tumor of the eye in Senegalese children. Diagnosis occurs often at a late stage, when enucleation is unavoidable. In this report, we describe all recorded cases occurring in Senegal over a 10-year period (2005–14). For the 106 cases clinically and radiological identified, the mean age at diagnosis was 30 months (2 months / 10 years). Leucocoria (74.6%) and exophthalmos (42.3%) were common signs of the disease. For the 67 cases identified through patient-file examination, extra-ocular forms were present in 30% of cases at the time of diagnosis and retinal detachment in 19% of cases. Chemotherapy and surgery were the only available therapeutic methods. Overall survival at 5 years was 70%. Retinoblastoma is a serious illness that threatens the lives and sight of affected children. There are about 10 cases per year in Senegal. Management can be enhanced by improving existing technical platforms and training medical staff.

INTRODUCTION

Retinoblastoma (RB) is the leading eye tumor in children across the world and has been called a 'two-speed disease' in terms of its management in high-resource versus resource-limited settings [1].

In developed countries, screening programs and health education have led to diagnosis at an early stage. In addition, the therapeutic management of RB has greatly evolved over the past decade with advances in chemotherapy but also and especially thanks to local treatments such as thermotherapy and radiation therapy. Indications for enucleation are now fewer, and preserving sight has become the main objective. Five-year survival after diagnosis is almost 100% in high-resource settings [1, 2].

In low-resource contexts, however, RB remains a fatal disease [3, 4]. Patients often present at late stages when loss of sight has already occurred in the affected eye because of low population awareness of the condition and lack of screening programs. Treatment focuses thus on the preservation of life, and surgical intervention becomes inevitable. Despite efforts in the field of chemotherapy, the rate of death and recurrence after enucleation remains high in developing countries [3, 4].

RB is a cancer with a fatal spontaneous evolution, making treatment imperative [5]. The purpose of treatment is primarily to save life and if possible to preserve the patient's sight.

In Senegal, therapeutic management of RB underwent a major change in 2005 with the introduction of chemotherapy. Preliminary results published in 2010 showed encouraging signs with a reduction of more than half the mortality rate from 94% in 1994 to 35.6% in 2010 [4]. Senegal does not yet have conservative treatments or radiation therapy for children. In addition, absence

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of screening leads to late diagnosis with the presence of high-risk histological prognostic factors at the time of enucleation. Treatment is therefore limited to chemotherapy and surgery.

In this case series, we describe the evolutionary profiles of RB in Senegal over a 10-year period subsequent to the introduction of chemotherapy.

METHODS

We carried out a multi-center retrospective analysis from November 2005 to October 2014 drawing on patient files from the Aristide le Dantec Hospital, the only pediatric eye surgery center in Senegal. Data were collected from patient medical records and pathology reports available in three public laboratories. Out of 106 patients with suspected RB, 10 died before surgery; the remaining 96 underwent an operation, but only 67 of them (68 eyes, since one case was bilateral) had tissue sent for histological analysis. The Kaplan–Meier method was used to establish the survival curve after treatment.

RESULTS

Diagnostic assessment

During our study period, for the 106 cases clinically and radiologically identified, the sex ratio was 1.03. Almost half of our patients were 3 years old or older ($n = 49$, 48.5%). Familial RB was found in three cases. A first- or second-degree consanguinity was present in 31.1% of the children. RB was bilateral in 24 cases (20.7%). The mean age at diagnosis of the bilateral form was 23.3 months (1.9 years) with extremes of 2 and 48 months. RB had reached a single eye in 82 cases at an average age of 30.6 months (2.5 years) with extremes of 2 and 120 months.

The circumstances of discovery of RB were multiple. During our study period, leucocoria and exophtalmos were found in at least one in two children in 97 cases (74.6%) and 55 cases (42.3%), respectively. Ocular pain, strabismus, adnexal inflammatory syndrome, decreased visual acuity and chemosis were present in about one in four children.

Pathologically, the tumor was intraocular in 48 cases and extra-ocular in 20 cases. The tumor was accompanied by retinal detachment in 13 cases (19.2%).

Therapeutic intervention

All patients with suspected RB received chemotherapy with vincristine-cyclophosphamide or vincristine-carboplatin-etopside. The thermotherapy and the curi-therapy do not exist now in Senegal. Ten patients died before surgery. Of the 96 operated patients, for 80 enucleation (including three bilateral enucleations) and 19 exenteration, histopathological results were found for 67 of them (one case of bilateral enucleation). Pathologically, RB was well differentiated in 11 cases (16.2%), moderately differentiated in 31 cases (45.6%) and poorly differentiated in 26 cases (38.2%).

After a mean of 57.2 months, 15 patients were lost to follow-up. Of the patients still followed, 28 (41.8%) were in remission, 10 (15%) had recurrence on the enucleated side and 14 patients (20.1%) had died. The 2-year overall survival was 84%, 5-year overall survival was 70% and 34% at 10 years (Fig. 1).

In multivariate analysis, tumor differentiation and pT1 were related to remission. Retrolaminar optic nerve invasion, massive choroidal invasion and a pT3 stage were risk factors for recurrence. Low tumor differentiation, optic nerve sectional area involvement, tumor scleral breaking and pT4 stage were associated with death.

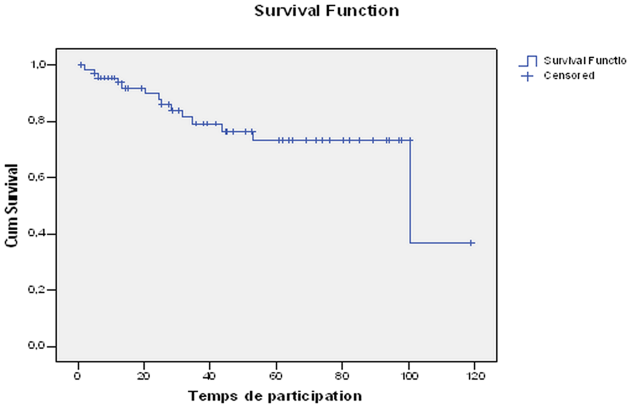

Figure 1: Overall survival curve using the Kaplan–Meier method.

DISCUSSION

This study on RB is the second and largest since the introduction of chemotherapy for this condition in Senegal [4]. It included all diagnosed cases in the country for the 10-year period we looked at. However, the total absence of histopathological analysis may have introduced selection bias with regard to histopathological factors of RB in Senegal.

Our study documents the delay in diagnosis of RB in Senegal with admission at advanced stages of the disease and consequently high death rates. Seventy-nine percent of patients treated with chemotherapy survived but at the price of blindness; 20.1% mortality contrasts with zero mortality from RB in developed countries [2]. The future challenges of RB management in Senegal are population sensitization and education, improvement of the medical equipment and decentralization of centers of care.

In conclusion, the management of RB in Senegal can be further improved to preserve the life and sight of affected children. Senegal's aim is to eliminate RB as a cause of death by 2020 as part of its national plan to fight cancer. This will require close collaboration among ophthalmologists, oncologists, pathologists and radiation therapists.

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CONFLICT OF INTEREST STATEMENT

None declared.

INFORMED CONSENT

Not necessary.

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