Journal of the Egyptian National Cancer Institute (2015) 27, 195–203

Cairo University

Journal of the Egyptian National Cancer Institute

www.elsevier.com/locate/jnci
www.sciencedirect.com

Full Length Article

Clinical presentation of intraocular retinoblastoma; 5-year hospital-based registry in Egypt

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Received 2 January 2015; revised 31 August 2015; accepted 10 September 2015
Available online 17 October 2015

KEYWORDS

Retinoblastoma;
Egypt;
Developing countries;
Incidence

Abstract *Purpose:* To study the presenting signs of Retinoblastoma in Egypt at Egypt's main pediatric oncology referral center.
Methods: This is a prospective descriptive study (hospital-based registry) conducted at Children's Cancer Hospital Egypt between July 2007 and December 2012.
Results: Out of 262 patients diagnosed with retinoblastoma, 244 were suffering from intra-ocular disease at presentation. One hundred thirty-nine (57%) patients presented with unilateral disease, while 105 (43%) suffered bilateral disease. The mean age at presentation was 20.6 ± 17 months, averaging 18.87 ± 11.76 months for bilateral and 25.72 ± 18.78 months for unilateral disease. 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. Patients with advanced disease (Group C–E) had longer duration of symptoms.

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Peer review under responsibility of The National Cancer Institute, Cairo University.

http://dx.doi.org/10.1016/j.jnci.2015.09.002

Conclusion: In Egypt, retinoblastoma patients present more frequently with advanced disease. There is an ever-increasing need to develop a national team dedicated to studying disease significance and formulating a national awareness program.

Introduction

Retinoblastoma is the most common primary ocular malignancy of childhood, with a global incidence of 11 patients per million children younger than five [1,2]. No racial predilection is known for retinoblastoma. Studies showed no significant difference in the incidence of retinoblastoma by gender for children aged between 0 and 14. The current estimated boy-to-girl ratio is 1.12:1.

Retinoblastoma could occur unilaterally or bilaterally. Leukocoria is the most common presenting sign of the disease followed by strabismus; the least frequent presenting signs being atrophia, and proptosis. Retinoblastoma is a disease usually discovered by parents. The International Intraocular Retinoblastoma Classification (IIRBC) allows better initial evaluation and follow-up of intra-ocular tumor size and location allowing for better risk assessment in the chemotherapy era compared to Reese-Ellsworth Classification [3].

Although retinoblastoma presentation and management is a real health problem of developing countries, the extent of the problem is under-studied in the international literature [4].

Our aim was to discuss the presenting symptoms and signs of retinoblastoma in patients diagnosed at Egypt's major pediatric referral center during the five years since its inauguration (2007–2012). This is the first publication to describe the disease using this approach with a large cohort of patients in such a short period in Egypt.

Patients and methods

We prospectively included all children diagnosed at the Children's Cancer Hospital – Egypt (CCHE) – and reported to its retinoblastoma registry from 2007 to 2012 – in our study. Initial data included symptoms, family history of cancer, clinical manifestations, physical examination, and radiological and laboratory studies. Clinical information and follow-up data were available for all patients who formed the basis of this study. The initial evaluation of the children included complete ophthalmological and systemic examination. The ophthalmological evaluation included (a) a dilated-fundus examination using RetCam™, with scleral indentation under general anesthesia (EUA); (b) ultrasonography of the eye; and (c) CT/MRI of orbit. In patients where extra-ocular dissemination was clinically suspected, systemic evaluations were conducted. This included a full blood count; a lumbar puncture for cerebrospinal fluid (CSF) cytological analysis; and a bone marrow aspiration looking for tumor cells and evidence of metastases. Case Report Forms (CRFs) were developed based on a review of PubMed-indexed publications. IIRBC was implemented for initial staging of intraocular disease. Details of IIRBC can be found elsewhere [3,5]. This study adhered to ethical considerations regarding research on human subjects.

Data management was done using REDCap (Research Electronic Data Capture) tool hosted at CCHE after replacement of an in-house developed software [6,7]. All statistical calculations were done using SPSS (Statistical Package for the Social Science; IBM, USA) version 20 for Microsoft Windows.

Statistical analysis

Quantitative data were described in terms of mean ± standard deviation (SD), Qualitative data were described as proportions and percentages. To compare quantitative variables between two-study groups we used the student *T*-test and Mann–Whitney *U* if data were normally or not normally distributed respectively. A one-way analysis of variance (ANOVA) and a Kruskal–Wallis one-way analysis of variance (KWOVA) were used for comparing quantitative variable among more than 2 groups if data were normally or not normally distributed respectively. Chi square (X^2) test was used for comparing categorical data, and Fisher's Exact test instead if expected frequency is less than 5.

The Pearson moment correlation equation and Spearman rank correlation equation were used to correlate various variables for linear relationships in normally distributed and non-normally distributed data respectively. A probability value (*p*-value) less than 0.05 was considered statistically significant.

Results

Demography and geographical distribution

Two hundred and sixty-two (262) patients presented to CCHE between July 2007 and December 2012, representing 4.2% of the total number of patients that were treated at the hospital during the same period. Isolated intraocular retinoblastoma was found in 244 (93%) patients while 18 (7%) patients initially presented with extra-ocular retinoblastoma. Of those patients with intraocular disease, 139 (57%) presented with unilateral disease. The other 105 (43%) were bilaterally-affected. All in all, 349 eyes were diseased. The ratio of males to females was 1.02:1.0.

The hospital received patients from different countries; 220 patients (90.2%) were Egyptian, while 9.8% were from other countries. This included 15 patients (6.1%) from Yemen, 6 (2.5%) from Sudan, 2 (0.8%) from Palestine and one patient (0.4%) from Syria.

Fig. 1 shows the rate of enrollment of patients starting from July 2007. Thirty-four patients with retinoblastoma were enrolled in 2008 versus 61 patients in 2012.