Major Article

Strabismus in retinoblastoma survivors with long-term follow-up

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Because retinoblastoma (Rb) is today considered to be a curable cancer, with a survival rate of nearly 100% in developed countries and a high eye salvage rate, it is appropriate to focus attention on quality-of-life measures in Rb survivors. The purpose of this noncomparative interventional study was to report the long-term strabismus rate in salvaged Rb patients and to investigate possible risk factors leading to strabismus. The medical records of patients with Rb treated at the Royal London Hospital over a 9-year period were reviewed retrospectively. Of 149 Rb patients treated during the study period, 42 were included: 22 with bilateral and 20 with unilateral disease, all of which eyes were salvaged at final follow-up (ie, 64 eyes). Patients presented at a mean age of 8.2 months (range, 0.3-58.3 months). According to the International Intraocular Retinoblastoma Classification (IIRC) the worse eye at presentation was group A (1 eye), B (16 eyes), C (12 eyes), D (11 eyes), or no Rb (2 eyes). Fifteen patients (36%) were initially referred for screening due to family history of Rb. Overall treatments included intravenous chemotherapy (62 eyes), intraophthalmic artery chemotherapy (10 eyes), brachytherapy (11 eyes), transpupillary thermotherapy (22 eyes), cryotherapy (47 eyes), and external beam radiotherapy (4 eyes). At final follow-up (mean, 93.7 months), 69% of patients had strabismus, with exotropia being the most common type (18 patients), followed by esotropia (8 patients) and alternate exotropia/esotropia (3 patients). On univariate analysis, worse eye group IIRC and cTNMH, sporadic cases, strabismus and foveal tumor at presentation were found to be significant factors leading to strabismus at final visit ($P \leq 0.043$). On multivariate analysis, however, only foveal involvement, defined as tumors within 1.5 mm from the foveola, was found to be significant ($P < 0.001$). With the increasing use of conservative modalities and improved eye salvage rate, the findings from this study represent important additional information for Rb patients, their families and their physicians.

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PURPOSE To report the long-term strabismus rate in salvaged retinoblastoma (Rb) patients and investigate possible risk factors leading to strabismus.

METHODS The medical records of patients with Rb presenting at a single institution over a 9-year period were reviewed retrospectively with regard to ocular alignment outcomes after long-term follow-up.

RESULTS A total of 64 eyes of 42 patients (22 bilateral cases [52%]) were included, presenting with International Intraocular Retinoblastoma Classification (IIRC) in the worse eye as follows: group A (n = 1), B (n = 16), C (n = 12), D (n = 11), no Rb (n = 2). Fifteen patients (36%) were initially referred because of family history of Rb. Mean age at presentation was 8.2 months (range, 0.3-58.3 months). Overall treatments included intravenous chemotherapy (62 eyes), intraophthalmic artery chemotherapy (10 eyes), brachytherapy (11 eyes), transpupillary thermotherapy (22 eyes), cryotherapy (47 eyes), and external beam radiotherapy (4 eyes). At final follow-up (mean, 93.7 months), 69% of patients had strabismus, with exotropia being the most common type (n = 18), followed by esotropia (n = 8), and alternate exotropia/esotropia (n = 3). On univariate analysis, the worse eye group IIRC and cTNMH, sporadic cases, strabismus, and foveal tumor at presentation were found to be significantly associated with strabismus at final follow-up ($P \leq 0.043$). On multivariate analysis, only foveal involvement was found to be significant ($P < 0.001$).

CONCLUSIONS Strabismus, exotropia in particular, is a common adverse sequela following successful conservative treatment for Rb, with 69% of the present cohort having some type of deviation after long-term follow-up, for which foveal tumor at presentation was found to be a significant risk factor. (J AAPOS 2018;■:1.e1–1.e7)

Retinoblastoma (Rb), the most common primary intraocular cancer of childhood,[1] was commonly managed by enucleation. With recent advances in Rb management, including the use of intravenous chemotherapy (IVC), development of screening programs for at-risk patients, and case referral to specialized centers, Rb is considered a curable cancer, with a survival rate of nearly 100% and high eye salvage rates in developed countries.[2] The recent introduction and use of targeted chemotherapy[3,4] has allowed salvage of even advanced cases. With greater success in globe salvage, more attention is being paid to quality-of-life measures in Rb survivors. To our knowledge, the literature includes only a single report on long-term orthoptic outcomes, including strabismus, in Rb patients.[5] The present study aimed to report the rate of strabismus on long-term follow-up and to investigate potential risk factors associated with strabismus in salvaged Rb patients from all International Intraocular Retinoblastoma Classification (IIRC) groups.[6]

Subjects and Methods

This study was approved by the Barts Health NHS Trust Institutional Review Board and followed the tenets of the Declaration of Helsinki. The medical records of consecutive treatment-naive Rb patients presenting to the London Retinoblastoma Service from August 1, 2004 to August 15, 2012, were analyzed retrospectively.

The following data were retrieved from the record: patient age, sex, family history of Rb, presenting signs, clinical variables at presentation, and results of genetic testing. All eyes were classified

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