Outcome of Unilateral Retinoblastoma: A Ten Years Experience of Children's Cancer, Hospital Egypt

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Abstract: Background: A majority of children with retinoblastoma (60%) have a disease in one eye only (unilateral disease). This is a retrospective study to evaluate two different treatment modalities in those patients for saving their lives and vision. Methods: Four hundred and four patients were diagnosed with unilateral intraocular retinoblastoma at Children's Cancer, Hospital Egypt (CCHE) through the period of July/2007 until December/2017. Management strategies included primary enucleation versus ocular salvage treatment. Results: Patients presented with mean age 24.5 months with range (1.2-154.3 months). According to the international retinoblastoma classification, Group D (n=172, 42%) was the most common, followed by group E (n=142, 35%), group C (n=63, 16%), and group B (n=27, 7%). All patients were alive at the end of the study except four patients who died, with 5-years overall survival 98.3% [CI, (96.5-100%)]. Patients presented with advanced disease and poor visual prognosis (n=241, 59.6%) underwent primary enucleation with 6 cycles adjuvant chemotherapy if they had highrisk features in the enucleated eye; only four patients out of 241 ended-up either with extraocular metastasis (n=3) or death (n=1). While systemic chemotherapy and focal therapy were the primary treatment for those who presented with favorable disease status and good visual prognosis (n=163, 40.4%); seventy-seven patients of them (47%) ended up with a pre-defined event (enucleation, EBRT, off protocol chemotherapy or 2ry malignancy). Ocular survival for patients received primary chemotherapy + focal therapy was [50.9% (CI, 43.5-59.6%)] at 3 years and [46.9% (CI,39.3-56%)] at 5 years. Comparison between upfront enucleation and primary chemotherapy for occurrence of extraocular metastasis revealed that there was no statistical difference between them except in group D (p value). While for occurrence of death, no statistical difference in all classification groups. Conclusion: In retinoblastoma, primary chemotherapy is a reasonable option and has a good probability for ocular salvage without increasing the risk of metastasis in comparison to upfront enucleation except in group D.

Keywords: CCHE, chemotherapy, enucleation, retinoblastoma

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