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Service (sector) Tumor and Pathology - Ocular Oncology N° CEP

Reese- Ellsworth I- IV retinoblastoma eyes: Clinical features, treatment outcome and factors predictive for enucleation

Authors: Torres V; Freeman G; Macedo C; Ballalai P; Santos D; Teixeira LF; Martins C; Petrilli S; Erwenne C **Purpose:** To report clinical features and treatment results of RE I-IV retinoblastoma eyes and analyze the associated factors for eventual treatment with enucleation **Methods:** The data from RE I-IV retinoblastoma eyes regarding their clinical and therapeutic aspects were studied, between December 2000 and August 2005. Such data were further analyzed for their impact on the need for enucleation using univariate analyze (Exact Fisher Test) **Results:** Forty-one (43 eyes / 85 tumors) out of 98 retinoblastoma cases were selected for this study. Twenty-four cases (58,4%) were female and seventeen (41,6%) were male. There were 15 (36,5%) unilateral cases and 26 (63,4%) bilateral cases, with median age of 20,6 months and 17,3 months at diagnosis, respectively. In the unilateral cases the first sign of the disease was strabismus (53,3%) and in the bilateral cases was leucokoria (80,7%) . The 15 eyes from unilateral cases were classified according to the Reese-Ellsworth staging as: 1 (6,6%)RE I; 1 (6,6%)RE II; 12 (80%)RE III and 1(6,6%) RE IV. The 28 eyes from bilateral cases were classified as: 6 (21,4%)RE I; 10 (35,%)RE II; 8(28,5%) RE III and 4(14,2%) RE IV. In bilateral group the stage of the contralateral eye was RE V in 24 cases (92%). The eyes have received the following initial treatment: thermochemotherapy (90.3 %), primary brachytherapy (7,3%) and thermotherapy (2,4%). Such initial therapy was effective in 80,4% of the cases. Eight cases (19,5%) developed vitreous seeds in a median follow-up of treatment of 4,2 months, requiring brachytherapy (4 cases) and external beam radiation (4 cases) with tumor control in seven cases and no response in one case that requires enucleation. Failure of conservative treatment and need for treatment with enucleation occurred in eight cases (19,5%), seven cases secondary to recurrence of the disease and one case after no response to external beam radiation therapy, in a median follow- up of 6,8 months after initial therapy. Univariate analyze showed that pole posterior localization of the lesion and previous treatment with chemothermotherapy were associated to enucleation. **Conclusions:** The majority of the low-stage retinoblastoma eyes were found when the contralateral eye presented with advanced disease. Most of the RE I-IV retinoblastoma eyes (80,4%) showed a satisfactory tumor control with a conservative approach. Nine percent of the cases required external beam radiation therapy for eye salvage and 19,5% of the cases required enucleation. Pole posterior localization of the lesion and previous treatment with chemothermotherapy were the factors associated to ultimate enucleation.