

Case Report: Chemoreduction and Foveal-sparing TTT for Macular Retinoblastoma

Combined therapies allow globe retention and limited visual preservation.

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The management of retinoblastoma has had a remarkable evolution in the past century, from enucleation to methods of radiotherapy to current strategies of chemotherapy.¹ Over the past decade, new techniques of chemotherapy (chemoreduction [CRD]) using one, two, and three agents combined with adjuvant thermotherapy or cryotherapy have evolved. Our understanding of the value and limitations of chemoreduction and adjuvant methods for retinoblastoma has improved, and this method has replaced enucleation and external beam radiotherapy in many cases.^{2,3}

There is special consideration when using this technique for macular retinoblastoma, as thermotherapy or cryotherapy can profoundly reduce macular vision. Our preference is to use foveal-sparing thermotherapy in an effort to treat the tumor but preserve some central visual acuity.² Herein, we illustrate a case of macular retinoblastoma treated with chemoreduction and foveal-sparing thermotherapy.

CASE REPORT

A 9-month-old boy presented with leukocoria and esotropia in the left eye for 3 months. He had no family history of retinoblastoma. He was the product of in vitro fertilization, born at 36 weeks gestation by cesarean section. The right eye was normal with fix-and-follow vision. The left eye had no fix nor follow vision and

there was a large macular retinoblastoma measuring 18x18 mm in diameter and 8.7 mm in thickness (Figure A). There was extensive subretinal fluid inferiorly with numerous visible subretinal seeds. The eye was classified as group D according to the International Classification of Retinoblastoma (ICRB).¹ The final diagnosis was unilateral sporadic retinoblastoma, and CRD with adjuvant consolidation therapy was advised.

Six cycles of CRD using vincristine, etoposide, and carboplatin were delivered. Chemotherapy-induction cryotherapy was provided at the first examination to induce a high concentration of chemotherapy into the vitreous cavity.

Following tumor reduction after the first chemotherapy cycle, transpupillary thermotherapy (TTT) was delivered to the regressed retinoblastoma for approximately 6 minutes, with a power of 500 mW and a spot size of 1.2 mm with little visible uptake in the areas of calcification and slight gray-white uptake in the noncalcified portion. A foveal-sparing approach was used so that the central 1.5-mm fovea and papillomacular bundle were not treated.² Three months following completion of chemoreduction there were recurrent subretinal seeds, which were controlled with several sessions of cryotherapy. The eye remains stable with no further recurrence at 25 months follow up (Figures B-D).

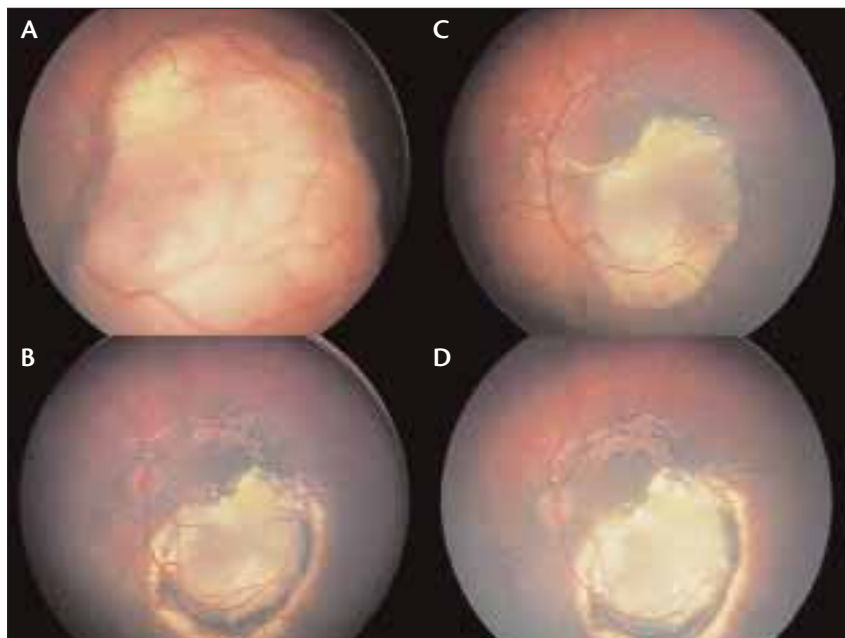


Figure. A 9-month-old boy with macular retinoblastoma in the left eye. (A) Fundus view of retinoblastoma in the macula before treatment. Subretinal fluid and subretinal seeds are barely visible. (B) After first chemotherapy. Type 3 regression with 30% calcium content. Note the subtle residual subretinal fluid nasally and inferiorly near the tumor margin, the calcified subretinal seeds superior to the optic disc and superior to the tumor. (C) After third cycle of chemotherapy. Type 3 regression with 30% calcium content. The subretinal fluid is reabsorbed and foveal-sparing thermotherapy has left an atrophic chorioretinal scar around the tumor base. (D) One year after chemoreduction. Type 3 regression with 40% calcium content is noted, and the tumor remnant remains stable.

DISCUSSION

The treatment of macular retinoblastoma is particularly challenging due to the balance of tumor control with the potential for treatment-related visual loss.² Chemotherapy has been shown to provide excellent control for retinoblastoma classified as groups A, B, and C with more than 90% success.⁴ Group D eyes show less success, as only approximately 50% show lasting control with chemoreduction. Newer approaches with the addition of subconjunctival carboplatin have improved tumor control. Chemoreduction is a favorable treatment for macular retinoblastoma as the tumor regresses, and often there is anatomically intact macular tissue that could provide some remnant of visual acuity. It has been shown that treatment of macular retinoblastoma with foveal-sparing thermotherapy in addition to chemoreduction provides more complete tumor control. Shields and associates evaluated 68 macular retinoblastomas treated with chemoreduction.² Those treated with chemoreduction and foveal-sparing thermotherapy showed lasting control in 83% of tumors at 4 years follow-up compared with 65% in those treated with chemoreduction alone.²

An analysis of visual results following successful chemoreduction for retinoblastoma revealed that 50% of eyes showed visual acuity of 20/40 or better at 5 years follow-up.⁵ Those with macular retinoblastoma displayed final visual acuity of 20/40 in 24% whereas those with extramacular retinoblastoma showed similar vision in 90%.⁵ In eyes with macular retinoblastoma, foveal-sparing TTT was important in preservation of visual acuity.

In summary, the treatment of macular retinoblastoma is particularly important for the patient's lifelong safety as well as the patient's ultimate visual acuity. The decision for tumor consolidation technique can profoundly affect a child's long-term visual acuity. The 5 minutes that it takes to deliver foveal thermotherapy could lead to 50 years of blindness. Hence, our preference is to spare the fovea of thermotherapy with the intention of protecting the child's visual acuity, particularly if this is the only remaining eye. ■

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