Intraoperative OCT Reveals Important Retinal Features of Retinoblastoma

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Retinoblastoma is a dangerous and blinding ocular malignancy of children. Management of this life-threatening condition requires detailed examination and treatment of tiny retinal tumors or seeds. General anesthesia is required at each examination and therapeutic session, particularly for infants and young children. Diagnostic imaging is performed at each examination to assess tumor and retinal status, along with an estimate of visual potential.

Optical coherence tomography (OCT) is a noninvasive in vivo imaging technique that generates high-resolution cross-sectional images using low-coherence interferometry. Since its introduction in 1991, OCT has been widely adopted in ophthalmology for diagnosis, monitoring, and clinical decision-making.[1] Traditional OCT systems require focused attention and cooperation from the patient. For these reasons, OCT has not been widely adapted for use in young children, particularly those with retinoblastoma.

The development of hand-held intraoperative OCT has resulted in clinical applications previously unforeseen in pediatric retinal disease.[2,3] We describe the utility of intraoperative OCT for tumor and retina monitoring. This tool has proven to be a valuable technique that aids decision-making regarding therapy and potential visual acuity in young preverbal children with retinoblastoma.

CASE DESCRIPTION

A 14-month-old white male with no family history of retinoblastoma displayed exotropia of the left eye (OS) for 6 months and was found to have retinoblastoma. On examination, visual acuity was fix and follow in the right eye (OD) and no fix or follow OS. There was no nystagmus, and intraocular pressures were normal.

The right fundus showed a solitary retinoblastoma measuring 12 mm in base and 6 mm in thickness. This tumor had a cavitary (low-grade) appearance and showed no subretinal fluid, subretinal seeds, or vitreous seeds. The left fundus showed a solitary macular retinoblastoma measuring 15 mm in base and 6 mm in thickness with dependent subretinal fluid and subretinal seeds extending to the inferior ora serrata (Figure, A and B). Ultrasonography of each eye disclosed an intraocular tumor with intralesional calcification, consistent with retinoblastoma.

It was decided to use three-agent intravenous chemoreduction with vincristine, etoposide, and carboplatin for a planned six cycles with supplementary foveal-sparing transpupillary thermotherapy for tumor consolidation. Both tumors showed favorable response with regression to type 3 scars after six cycles (Figure, C and D).

In each eye, the tumor involved the macula, and

At a Glance

- A 14-month-old white male with no family history of retinoblastoma displayed exotropia of the left eye for 6 months and was found to have retinoblastoma.
- Intraoperative optical coherence tomography was instrumental in identifying tumor features, estimating visual potential, and as an aid to clinical decision making.

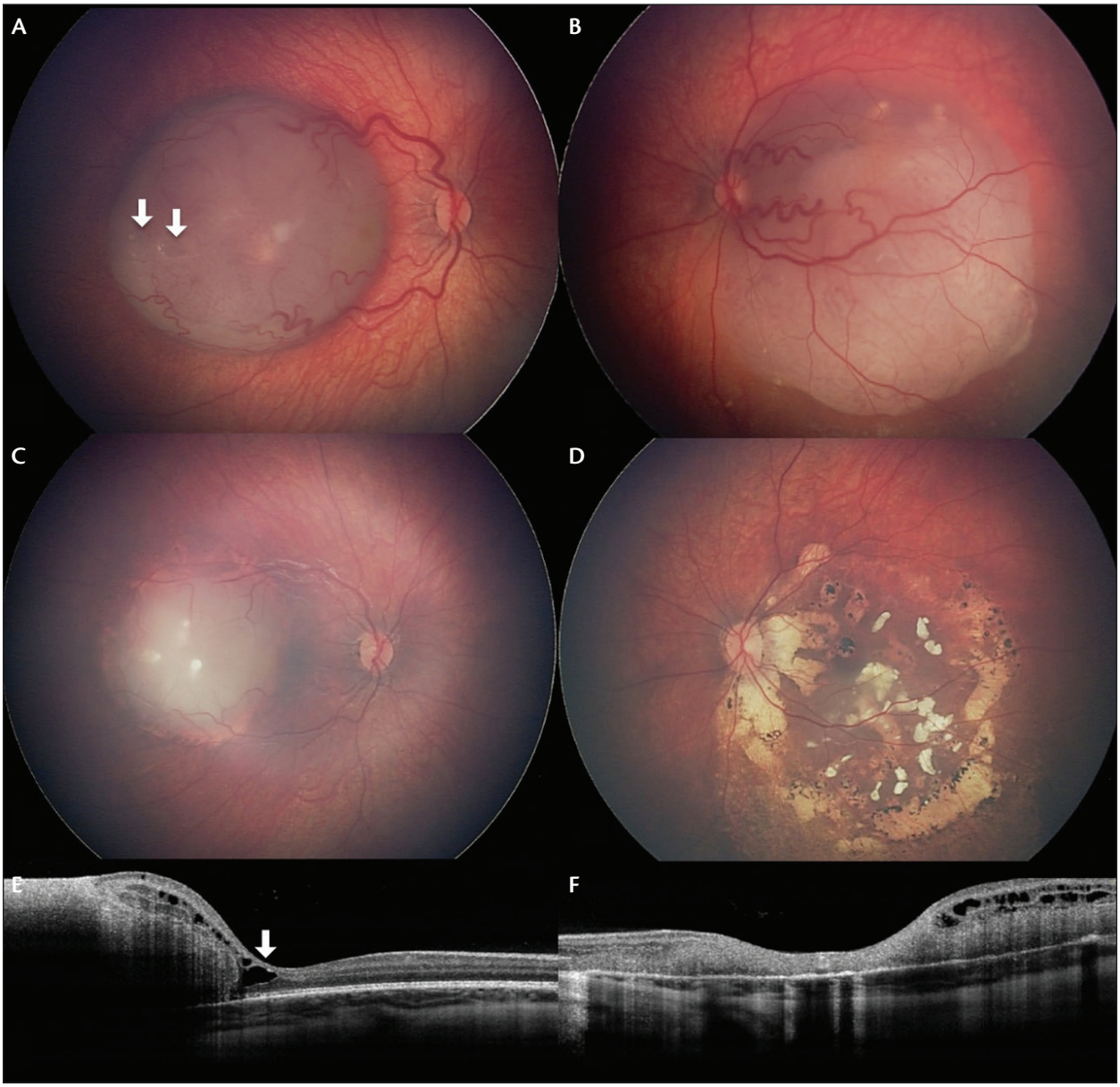

Figure: A 14-month-old boy with bilateral macular retinoblastoma. At presentation, the right eye (A) had a macular tumor with cavitary changes (arrows) and dilated feeding vessels, and the left eye (B) had a larger tumor touching the optic nerve with dilated feeding vessels and focal areas of calcification. Following six cycles of chemoreduction, there was mild regression of the solid tumor in the right eye (C) and more extensive regression in the left eye (D). In both eyes, the residual tumor appeared to be in the fovea; however, handheld intraoperative optical coherence tomography showed the tumor splitting the foveal region with cystoid macular edema (E, arrow). The most important finding was that the foveola was structurally intact in the right eye, except for outer retinal edema. On the other hand, there was loss of normal retinal architecture in the left eye (F).

there was concern regarding future visual potential. Intraoperative OCT (iVue, Optovue) revealed a regressed retinoblastoma scar in the outer retina OD with the central foveola visible. There was cystoid edema and no subretinal fluid. The foveola was draped over the medial margin of the regressed tumor scar.

OCT of the left eye displayed disorganized foveal retina with thinning, no foveal contour, and no subretinal fluid (Figure, E and F). Judging from the findings on macular OCT in this preverbal 20-month-old child, we predicted that the visual potential could be favorable OD and would likely be poor OS, with additional risk of

amblyopia. Short-term daily patching was prescribed to minimize vision loss OS.

DISCUSSION

Visual impairment from macular retinoblastoma can be caused by direct damage of the tumor to foveal structures, longstanding subfoveal fluid, subretinal or vitreous seeds, and/or necessary treatment to this region for tumor control. The management strategy for retinoblastoma involves a delicate balance of tumor control and globe salvage against visual outcome. This is especially relevant with macular retinoblastoma, where a single misplaced laser spot could be visually devastating.

OCT is remarkably valuable in these circumstances for estimating the ultimate visual potential based on foveal anatomy. As illustrated in our case, despite the facts that the tumor involved the foveola and that later regression of the tumor split the foveola, OCT revealed a somewhat intact foveola draped over the margin of the tumor. This was not clinically visible, as the foveal reflex was not apparent during examination. We have found OCT useful in locating an eccentric foveola following settling of extensive subretinal fluid.

Because it allows monitoring of vitreous and subretinal seeds, OCT can also contribute to decision-making regarding further therapies. Intraoperative OCT utilizes high-resolution spectral domain technology and has the ability to perform enhanced depth imaging of the choroid. This feature can be useful in assessing choroidal thickness following intraarterial chemotherapy, as that treatment modality has been shown to cause choroidal thinning.[4]

There are few reports in the literature on the use of OCT for children with retinoblastoma. In 2004, our team reviewed the application of time-domain OCT in children with intraocular tumors in an office setting and realized that children as young as 4 years old would generally cooperate. Using OCT, we found that pediatric retinoblastoma demonstrated retinal disorganization, cavitary changes, and posterior shadowing from tumors.[5] Later, Rootman and associates reported their experience with handheld spectral domain OCT for retinoblastoma.[6] They evaluated 22 tumors in 16 patients whose mean age was 1.9 years, all of whom were imaged under general anesthesia. Small active noncalcified retinoblastomas appeared as thickening within the middle retinal layers, causing a smooth nodular elevation and draping of the nerve fiber layer and internal limiting membrane. Vitreous seeding appeared as spherical lesions superficial to the retina, causing posterior shadowing when it was of substantial size. These authors proposed that the clinical applications of intraoperative OCT included detecting small retinoblastomas, monitoring treatment responses, and detecting subtle areas of recurrence. We describe additional uses of intraoperative OCT, including identifying normal or draped foveola, assisting in prediction of visual function, and, ultimately, allowing earlier visual acuity rehabilitation.

CONCLUSION

OCT is a safe and reliable tool for monitoring response following treatment for retinoblastoma. We believe that this modality will be important in predicting visual outcome in patients with retinoblastoma. Information from OCT may be useful in planning amblyopia treatment and maximizing the visual potential of each patient. ■

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