DIFFUSE PERIPAPILLARY CHOROIDAL MELANOMA THAT EVOLVED FROM A SMALL PRESUMED CHOROIDAL NEVUS SUCCESSFULLY TREATED WITH GAMMA KNIFE RADIOTHERAPY

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Purpose: To report a case of diffuse peripapillary choroidal melanoma which began as a small choroidal nevus and was successfully treated with gamma knife radiotherapy.

Methods: Observational case report.

Results: A 31-year-old visually asymptomatic man presented for a routine eye examination and was noted to have a small choroidal nevus. Six years later, the lesion had become a diffuse juxtapapillary choroidal melanoma with a thickness of 1.9 mm. Given the peripapillary location of the tumor with involvement of about 6 clock hours of the disk, Leksell Gamma Knife radiotherapy was performed. At 5.5 years after radiation therapy, visual acuity remained 20/20. The lesion thickness had decreased to 1.5 mm, and there was no interval growth of any margins.

Conclusion: Small choroidal nevi carry low malignant potential but still deserve photographic documentation when possible with regular follow intervals. Gamma knife radiotherapy can be considered for tumors abutting the optic nerve, especially when plaque radiotherapy may be technically difficult.

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Case Report

A 31-year-old visually asymptomatic man presented for a routine eye examination and was noted to have a small choroidal nevus along the inferior arcade in his right eye (Figure 1). His visual acuity was 20/20.

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It was recommended that he returns in one year for reexamination, but did not seek further examinations until he moved to a new state 6 years later. The lesion has clearly become a diffuse juxtapapillary choroidal melanoma (Figure 2). The B-scan ultrasonography showed a 1.9 mm tumor determined with high internal reflectivity. His visual acuity remained 20/20.

Given the peripapillary location of the tumor with involvement of approximately 6 clock hours of the disk, radiation therapy in the form of gamma knife (GK) was recommended. The lesion was treated with the Leksell GK 4C. The tumor was contoured on axial magnetic resonance images, and GammaPlan software (Elekta Instruments) was used to calculate the radiation doses within the contoured structures. A dose of 20 Gy was delivered to the 50% isodose line through 1 isocenter of 8-mm collimation and 1 isocenter of 14-mm collimation. Custom collimator plugging was used to limit the dose to the lens and optic nerve.

At 5.5 years after radiation therapy, visual acuity remained 20/20. The lesion thickness had decreased to 1.5 mm, and there was no interval growth of any margins. Mild radiation retinopathy was noted, consisting of a few retinal hemorrhages over the lesion (Figure 3).

Fig. 1. Color fundus photograph of the right eye at presentation. Note small (1.0×0.5 mm), minimally pigmented choroidal nevus along inferior arcade with fibrillated margins.

Discussion

Differentiating between choroidal nevus and a small or early choroidal melanoma can be difficult, which has made treatment decisions of small suspicious lesions sometimes challenging. It is estimated that 6% of the white population has a choroidal nevus and that only 1 in 8,845 of these nevi become melanoma.^{1,2} There are several features of nevi that have been shown to predict transformation into melanoma. In an analysis of 2,514 choroidal nevi, factors predictive of growth into melanoma included tumor thickness greater than 2 mm, subretinal fluid, visual

Fig. 2. Appearance of the lesion 5 years later. A peripapillary diffuse choroidal melanoma is present with orange pigment and a thickness of 1.9 mm on B-scan ultrasonography.

Fig. 3. Lesion 5.5 years after treatment.

symptoms, orange pigment, tumor margins within 3 mm of the optic disk, ultrasonographic hollowness, halo absence, and drusen absence.³ The median hazard ratio for those with 1–2 risk factors was 3; for 3 or 4 factors, 5; for 5 to 6 factors, 9; and for all 7 factors, 21.⁴ At initial presentation, the patient reported herein had none of the risk factors. In fact, the lesion resembled that of a choroidal freckle, an entity that is believed to have no malignant potential.

Therapies for uveal melanoma include observation, enucleation, resection, plaque brachytherapy, proton beam therapy, GK therapy, and combined therapy with photocoagulation or transpupillary thermotherapy. Because disease progression is variable, there is debate over the best treatment paradigm. Currently, plaque brachytherapy and proton beam therapy are the most widely used and extensively studied forms of radiation therapy for uveal melanoma.

We use GK to treat some choroidal melanomas and solitary metastasis when plaque brachytherapy are relatively contraindicated because of large tumor size and/or proximity to the optic nerve, or because of patient's general health or choice. The GK requires a retrobulbar block to create global akinesia and attachment of a stereotactic head frame to the skull with 4 pins. Unlike plaque brachytherapy and proton beam therapy, no incisional surgery is required. In GK therapy, the patient undergoes magnetic resonance imaging localization and then radiation treatment in a single session lasting less than a half a day, which has been shown to be as effective as fractionated doses.⁵ Proton beam therapy requires an expensive

particle accelerator that is currently available at only 14 treatment centers, whereas there are approximately 125 GK facilities in the United States.^{6,7} Ocular adverse effects of GK include radiation retinopathy, cataract, optic neuropathy, and secondary neovascular glaucoma.⁸

In small series, for large or juxtapapillary tumors, GK therapy has been shown to have similar tumor and ocular outcomes when compared with alternative therapies. Multiple studies have reported local tumor control rates from 91 to 98% in patient populations with large or juxtapapillary tumors.^{5,8,11} In addition, five-year survival rates between GK treatment and enucleation are similar.¹² There are no studies comparing the visual acuity outcomes among the radiation therapies, and there is limited data on GK therapy's effect on visual acuity. According to 1 study, in patients with thick tumors or tumors proximal to the optic nerve that were treated with GK, 35% of patients had visual acuity equal or better than 20/200 at mean clinical follow-up of 44 months.¹⁰

In conclusion, this case illustrates that choroidal freckles, while carrying low malignant potential, still deserve photographic documentation when possible with regular follow intervals. In addition, GK radiotherapy can be considered for tumors abutting the optic nerve, in which plaque radiotherapy might be technically difficult. Gamma knife is a good alternative to other radiation-based therapies and enucleation for choroidal melanomas that are large or juxtapapillary. Local tumor control and mortality seem similar to other therapies. Gamma knife therapy may not be as

effective at maintaining visual acuity as the other radiation-based therapies, but further research is required to show this with certainty.

Key words: uveal melanoma, choroidal nevus, choroidal freckle, gamma knife therapy.

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