Choroidal melanoma overlying scleral buckle evading detection treated with proton beam radiotherapy

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A B S T R A C T

Purpose: to present a case of a choroidal melanoma located along a scleral buckle contour and was successfully treated with proton beam radiotherapy, without disturbing the scleral buckle.

Observations: A 60-year-old woman presented with photopsias and history of retinal detachment repair with scleral buckle 36 years prior. She had annual dilated exams with her ophthalmologist over the last several years. While initially diagnosis of choroidal detachment, hemorrhage and intraocular buckle intrusion were suspected, the patient was referred to an ophthalmic oncology center and choroidal melanoma was confirmed. The patient underwent tantalum marker placement, trans-retinal biopsy and proton beam radiotherapy. Despite no evidence of metastatic disease on systemic imaging, prognostic genetic testing of the tumor revealed high-risk alterations for future metastasis, and the patient enrolled in an adjuvant clinical trial.

Conclusions: The location of this choroidal melanoma along a scleral buckle contour presented a unique therapeutic challenge. We successfully treated the tumor with proton beam radiotherapy without disturbing the scleral buckle.

Importance: chorioretinal elevations adjacent a scleral buckle may be presumed to be due to the buckle element; ultrasound can be helpful in distinguishing an intraocular tumor. Proton beam radiotherapy has the advantage over plaque brachytherapy in this setting as removing the scleral buckle is not needed and placing a plaque over the scleral buckle risks altering the effective radiotherapy dose.

1. Introduction

Uveal melanoma usually presents in White adults, almost 50% of whom will die of this disease.1,2 Clinical diagnosis is based on ophthalmoscopy and diagnostic imaging, including fundus autofluorescence, optical coherence tomography, ultrasonography and fluorescein angiography.

1.1. Case report

A 60-year-old woman with no significant medical history presented to her general ophthalmologist with progressive “shimmery lights in the top part of my vision” for 2 months. She had a prior history of macula-on rhegmatogenous retinal detachment in the right eye 36 years prior, repaired with cryopexy and encircling scleral buckle. She reported annual or biannual visits with an ophthalmologist since her retinal detachment repair and was last seen within one year. Visual acuity with correction was 20/60 in the right eye and 20/30 in the left eye without pinhole improvement. Pupils, motility and intraocular pressures were normal. Slit-lamp examination was notable for 2+ nuclear sclerotic cataracts in both eyes. Dilated examination of the right eye revealed an encircling scleral buckle, with an elevated, darkly colored area along the eye inferiorly. The differential diagnosis included choroidal detachment, intraocular buckle intrusion and hemorrhage, and choroidal melanoma.

The patient was referred to an ophthalmic oncology center, and a pigmented, inferior choroidal tumor was seen overlying and adjacent the buckle contour (Fig. 1A), with fundus hypo-autofluorescence overlying the lesion. Fluorescein angiogram showed double circulation with increased hyper-fluorescence in late frames. Ultrasound showed a highly elevated, dome-shaped inferior choroidal mass, with medium internal reflectivity with heterogeneous appearance and adjacent exudative retinal detachment. The scleral buckle was seen indenting the eye anterior to the tumor, which measured 14.9 mm × 14.1 mm with 7.1 mm thickness (Fig. 1B). The diagnosis of choroidal melanoma was made.
The patient elected for trans-retinal biopsy with a 27-gauge vitrector, and proton beam radiotherapy (56 GyE, fractionated over 4 days). Tantalum markers for radiotherapy planning and treatment were placed at the time of biopsy. After a limited conjunctival peritomy and isolation of the inferior rectus muscle, transillumination was performed and the tumor borders marked on the sclera. Two markers were sutured anterior to the scleral buckle and two markers sutured posterior to the scleral buckle. Marker location was confirmed using scleral depression at the site of each marker with indirect ophthalmoscopy and later by the radiation oncologist with X-ray. Ultrasound measurements and biometry were taken into account in the radiotherapy plan, with axial length of 27.5 mm in the tumor eye due to axial elongation from the scleral buckle, compared to axial length of 26.1 mm in the fellow eye.

Cytology confirmed melanoma with spindle and epithelioid cells. Next-generation sequencing with the UCSF500 pan-cancer assay showed monosomy 3 and BAP1 mutation, which indicate a high risk of metastatic disease. This poor prognosis was confirmed by gene expression profiling (Castle Biosciences, Friendswood, TX), which showed a Class 2 molecular signature profile.

The patient received 6 months of systemic Sunitinib in a randomized trial comparing this agent with Valproic Acid. Abdominal MRIs every 3 months have revealed no metastatic disease, to date.

Four years after radiotherapy, the patient maintained 20/80 vision with epiretinal membrane and nuclear sclerotic cataract in the right eye. The tumor appeared regressed on exam (Fig. 1C), with dimensions on ultrasound of 13.9 mm × 12.2 mm, with 6.5 mm thickness.

2. Discussion

The location of a choroidal melanoma over a circumferential scleral buckle presented a number of challenges, which are discussed in this report.

Motiani et al. treated a similar tumor with an iodine plaque, which was placed over the buckle. Although they did not describe their radiation dosimetry, they did mention the risk of inadequate radiation dose to the tumor because the buckle increased the distance between the plaque and the tumor and also because of the possible radiation-attenuating effect of silicone. In our case, these factors were obviated by the location of the buckle distal to the tumor so that it did not interfere with the proton beam. When placing the tantalum markers, it was not necessary to disturb the scleral buckle, so that any risk of causing re-detachment of the retina was avoided. Interestingly, our patient retained vision of 20/80 in the treated eye four years after the proton beam radiotherapy whereas the case reported by Motiani et al. was complicated by radiation retinopathy. Such morbidity may have been aggravated if the radiation dose had been increased to adjust for any interference by the buckle and if such a precaution had not been necessary.

Although the scleral buckle distorted the globe, as evidenced by the disparity in axial length between the two eyes, this did not interfere with the computerized modeling of the eye when planning the proton beam radiotherapy. The radiographic confirmation of tumor location and tantalum markers ensured accuracy.

Biopsy of anterior choroidal tumors tends to be performed trans-sclerally, but the scleral buckle may have made this approach more difficult. The buckle did not interfere with our technique for performing biopsy trans-retinally.

3. Conclusions

The location of this choroidal melanoma overlying a scleral buckle created therapeutic challenges, which were overcome with proton beam radiotherapy. Trans-retinal biopsy was not compromised by the presence of the scleral buckle. Genetic analysis of the tumor sample with next-generation sequencing, gene-expression profiling and multivariable prognostication provided enough evidence of tumor lethality to enable the patient to be entered into a clinical trial of systemic adjuvant therapy.
Patient consent

The patient provided written consent for use of her ophthalmic images in this study.

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Declaration of competing interest

The authors have no conflicts of interest or financial disclosures to report.

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