Tapioca Melanoma of the Iris Without Iris Heterochromia

Austin Bach, DO, MPH, Sara McGowan, OD, Aaron S Gold, OD, Victor M Villegas, MD3, Andrea C Wildner, CRA, Fiona J Ehlies BSc(Hons), CDOS, Jordan M Thompson, MD, J Antonio Bermudez-Magner, MD, Sander R Dubovy, MD, Timothy G Murray, MBA

Introduction

Tapioca melanoma of the iris is a rare form of diffuse iris melanoma. To date, there have been a total of 20 cases of tapioca melanoma of the iris reported.1-13 It usually presents unilaterally with hyperchromic heterochromia and the patient may have glaucomatous changes secondary to obstruction of the aqueous outflow. The neoplasm is generally described as lightly pigmented with a nodular surface similar to the appearance of tapioca that is diffuse and circumferential and involving the iris, anterior chamber angle and possibly the ciliary body.14,15 It is described as a lightly pigmented, multinodular lesion.14 Making a clinical distinction between a malignant and benign lesion of the iris is very difficult. It must be confirmed immunohistochemically with a number of different stains. Melanomas of the iris are known to cause local invasion and when they become metastatic, it is usually to the liver.15 Identification of tapioca melanomas of the iris is important because its presentation and surgical management may differ from that of other malignant iris melanomas.

Case Presentation

A 14-year-old white female presented to our clinic for evaluation of a mass on the iris of her right eye. The patient had no significant medical or surgical history and her family history was non-contributory. The lesion was originally noted approximately three years prior to this exam, however, over the last four months a progressive increase in size of the lesion had been observed. The patient was being treated elsewhere with prednisolone acetate 1% four times per day. On examination, best-corrected visual acuities were 20/20 OD and OS and her pressures were 8 and 11, respectively. Slit-lamp examination revealed a distorted pupil OD; her visual fields and extraocular motility were intact and Amsler grids were stable. The patient's anterior segment examination revealed an elevated, markedly vascular, amelanotic, multifocal mass situated at the 5:30 clock hour of the iris OD, along with multiple multifocal smaller satellite lesions (Figure 1A). The patient's anterior segment examination was unremarkable and did not display iris heterochromia when compared to the fellow eye (Figure 1B). On gonioscopic evaluation of the right eye, the lesion appeared to extend along the angle from the 4:30 to 7:00 clock hours (Figure 2, partial view of mass extension along angle). High-resolution echography measured the tumor to be approximately 8 x 2.2 mm, without posterior extension (Figure 3). Dilated fundus examination was unremarkable OU.

The patient underwent a biopsy of the lesion which showed the stroma largely replaced by sheets and nests of cellular tumor. The tumor was composed of amelanotic/lightly pigmented spindle cells with bland nucleolus. Rare nucleoli were identified. The cells formed interweaving fascicles. The melan A and S-100 stains were positive and the K167 stain showed moderate positivity. The LCA stain was negative. It was determined to be a low grade spindle melanoma, consistent with clinical findings.

On November 22, 2010, the patient underwent an uneventfulenucleation of the right eye. She is presently free of metastatic disease.

Discussion

The most unique presentation of the patient in this case was the lack of iris heterochromia. It has been thought that tapioca melanoma of the iris occurs in patients with heterochromia,12,13 but, as can be clearly seen by Figure 1, our patient did not present with heterochromia. The satellite lesions covered some of our patient's iris, the majority of the tumor remained localized inferiorly, allowing excellent visualization of the iris stroma. She also had not developed intraocular pressures nor did she show any glaucomatous changes. These deviations from the norm of this rare disease may bring up the question of different genomic types of tapioca melanoma of the iris with different presentations in different patient populations.

Summary

The patient was a 14-year-old white female who presented to our clinic with a mass on her right iris. The lesion was amelanotic and multifocal. The patient underwent an uneventful enucleation of the right eye. The tumor was determined to be a low grade spindle melanoma. The lack of iris heterochromia is unique and highlights the distinct presentation of this rare disease.