A young woman with no acknowledged white non-Hispanic ancestry received a rare diagnosis.

BY KAREEM SIOUFI, MD; RENELLE POINTDUJOUR-LIM, MD; AND CAROL L. SHIELDS, MD

UVEAL MELANOMA IN AN AFRICAN AMERICAN PATIENT

The incidence of uveal or choroidal melanoma in the African American population remains exceptionally low when compared with the incidence in the white population. Data from the National Cancer Institute’s Surveillance, Epidemiology, and End Results (SEER) database shows that 97.8% of patients with choroidal melanoma are of white non-Hispanic ancestry, whereas only 0.5% are of African heritage.

In a comprehensive retrospective case series of 8100 patients with uveal melanoma, Shields et al concluded that nonwhite non-Hispanics demonstrated larger melanoma, more often located peripherally, and with intraslesional hemorrhage, compared with white non-Hispanics who had melanoma. However, Kaplan-Meier analysis of that cohort demonstrated similar risk for metastatic disease in all races. Herein we report a case of choroidal melanoma in a relatively young African American woman.

CASE STUDY

A 30-year-old African American woman noted glare in her left eye (OS) for 2 months. She was found to have a retinal detachment with an underlying mass and was referred for opinion and management. She denied knowledge of white non-Hispanic ancestry and was otherwise healthy.

On examination, she demonstrated Fitzpatrick skin type 6 and showed no evidence of oculodermal melanocytosis (Figure, A). Her visual acuity was 20/25 in each eye (OU), and intraocular pressures were 12 mm Hg OU. The anterior segment revealed mild complexion-associated melanosis in the limbal region OU.

Fundus evaluation of the right eye was normal. The fundus OS displayed a pigmented choroidal mass in the inferonasal juxtapapillary region, measuring 14 mm in basal dimension and 8.1 mm in thickness on ultrasonography. Surrounding subretinal fluid was noted (Figure, B). The mass demonstrated mild acoustic hollowness, particularly in the basal area on B-scan ultrasonography (Figure, C) and low internal reflectivity on A-scan ultrasonography. Enhanced-depth imaging optical coherence tomography confirmed the presence of subretinal fluid nasal to the optic disc (Figure, D) and documented an intact macula (Figure, E). These features are consistent with choroidal melanoma in an African American woman.

Options for management included enucleation or plaque radiotherapy. After appropriate informed consent, the patient opted for treatment with plaque radiotherapy using iodine-125 radioisotope on an 18-mm notched, custom-designed plaque applicator. The patient refused prognostic genetic testing.

DISCUSSION

Although uveal melanoma is the most common primary intraocular neoplasm in adults, it is uncommon for uveal melanoma to occur in African American individuals. In fact, the incidence in this population is estimated to be between 0.3% and 0.5%. Kivela estimated that as many as 7095 patients worldwide are affected annually with uveal melanoma, with 4747 cases in white non-Hispanics, 1286 in Asians, 738 in Hispanics, and 316 in Africans.

- Uveal melanoma is the most common primary intraocular neoplasm in adults.
- The incidence of uveal melanoma in African Americans individuals is quite low.
- When it does occur in African Americans individuals, uveal melanoma tends to be larger and have more invasive features yet with a prognosis similar to that in white non-Hispanics.

AT A GLANCE
that report, the incidence per million was 6.0 for white non-Hispanics, 0.3 for Asians, 1.6 for Hispanics, and 0.3 for Africans.

Singh et al reviewed the US SEER database over a 25-year period and determined the overall age-adjusted incidence of uveal melanoma to be 4.3 per million, with 98% of cases occurring in the white non-Hispanic population. They noted that the rates of uveal melanoma remained stable over this 25-year period and that there was no variance based on geographic location in the United States.

In the 2015 Doyne Lecture, Shields et al described a comprehensive overview of 8100 patients with uveal melanoma over a 35-year period based on race. They observed racial composition of white non-Hispanic (n = 7918, 98%), Hispanic (n = 105, 1%), Asian (n = 44, <1%), and African American (n = 33 <1%) persons. Based on race (white non-Hispanic, Hispanic, Asian, and African American), they found that several factors were significantly different in these groups, including, respectively, mean age at presentation (58, 48, 44, and 52 years; P < .001); distance of posterior tumor margin to foveola (5, 5, 6, and 4 mm; P < .001); distance from posterior tumor margin to optic disc (5, 5, 6, and 4 mm; P < .001); tumor basal diameter (11, 12, 12, and 13 mm; P < .001); tumor thickness (5.4, 7.1, 6.5, and 7.5 mm; P < .001); intraocular hemorrhage (in 10, 14, 11, and 24%; P = .02); and rupture of Bruch membrane (in 20, 27, 39, and 36%; P = .001). Further multivariate analysis of this large cohort revealed increasing risk for metastasis with increasing age (P < .001), ciliary body location (P < .001), increasing tumor base (P < .001), pigmented tumor (P < .001), subretinal fluid (P < .001), intraocular hemorrhage (P = .045), and extraocular extension (P = .036).

Importantly, the risk for metastasis did not change based on race, and Kaplan-Meier analyses for metastasis at 5 and 10 years were 15% and 25% for the entire cohort. Compared with white non-Hispanic, despite a relative risk for metastasis of 0.73 for Hispanics, 1.42 for Asians, and 0.31 for African Americans, there was no statistical difference in metastasis or death from uveal melanoma based on race.

In comparison, cutaneous melanoma in nonwhite non-Hispanics demonstrates worse prognosis. In a cohort of 41,072 patients with cutaneous melanoma, Hu et al noted racial disparity, with incidences of 96.6% in white non-Hispanics, 2.7% in Hispanics, and <1% in non-Hispanic blacks. In contrast to uveal melanoma, race was an important factor for metastasis in cutaneous melanoma. Furthermore, Cormier et al reviewed the SEER database from 1992 to 2002 and found that nonwhite non-Hispanics were two to four times more likely to present with advanced stage 4 cutaneous melanoma, compared with white non-Hispanics. After adjustment for age, sex, and region, they found that nonwhite non-Hispanics demonstrated a two- to threefold greater risk for cutaneous melanoma—specific mortality compared with white non-Hispanics. The increased mortality risk is believed to be related to more advanced disease, especially in patients in whom cutaneous melanoma is not suspected.
CONCLUSION

Although the vast majority of choroidal melanoma occurs in white non-Hispanics, ophthalmologists should be aware of the possibility of uveal melanoma in African American individuals. Similar to cutaneous melanoma in African Americans, uveal melanoma tends to be larger and to have more aggressive features. Despite these characteristics, the overall prognosis is similar to that in white non-Hispanics. It is speculated that this protective effect could be related to background choroidal and/or cutaneous pigmentation, less threatening genetic alterations, or unexplained immunologic factors.


Renelle Pointdujour-Lim, MD
ocular oncology clinical fellow at Wills Eye Hospital, clinical instructor of ophthalmology at Thomas Jefferson University Hospital, both in Philadelphia, Pa.
renellelim@gmail.com

Carol L. Shields, MD
co-director of the Ocular Oncology Service, Wills Eye Hospital, Thomas Jefferson University in Philadelphia, Pa.
member of the Retina Today editorial advisory board
carolshields@gmail.com

Kareem Sioufi, MD
research intern at Wills Eye Hospital in Philadelphia, Pa.
kareem.sioufi@gmail.com

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