Small Choroidal Melanoma Arising in Diffuse Choroidal Melanocytosis

A man with a history of congenital oculodermal melanocytosis developed a small choroidal melanoma.

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The etiology of uveal melanoma remains poorly understood.[1-4] Factors related to the development of melanoma include both host and environmental sources. Host factors that lead to risk for melanoma include lighter colored eyes, lighter skin color, and ability to suntan when exposed to solar radiation.[4] Environmental factors for development of melanoma include prolonged ultraviolet light exposure, as in arc welding.[2,3,5] In a meta-analysis by Shah and associates,[5] occupational sunlight exposure was a borderline significant risk factor for uveal melanoma. Some studies have found sunlight exposure to be a risk, while others have not.[2,3,5] One important host factor that has been shown to be an important risk factor for uveal melanoma is the presence of ocular melanocytosis.[6,7] Ocular melanocytosis is a congenital condition characterized by hyperpigmentation of the periocular skin, episclera, uvea, and orbit, affecting 0.04% of Caucasians.[8] Herein we report a patient with longstanding congenital oculodermal melanocytosis who developed choroidal melanoma in his eighth decade of life.

CASE REPORT

A 72-year-old white man with known birthmark on the right eye was discovered on routine examination to have a pigmented choroidal mass. The patient was unaware of the risks of the birthmark for melanoma and maintained routine eye care every few years. On current examination, visual acuity was 20/20 in each eye. Intraocular pressure was 19 mm Hg in each eye. The left eye was unremarkable. External examination revealed cutaneous melanocytosis of the right periocular region including scalp, temple area, and upper and lower eyelids. The right eye displayed diffuse melanocytosis of the episclera, iris, and choroid. There was a choroidal mass superior to the macular region measuring 8 mm in basal diameter and 2.5 mm in thickness by ultrasound. Overlying orange pigment was seen clinically and confirmed on autofluorescence. Subretinal fluid was observed clinically and confirmed on optical coherence tomography (Figure 1). Despite the small size of the mass, a diagnosis of choroidal melanoma arising from diffuse choroidal melanocytosis was rendered. Treatment with plaque radiotherapy and thermotherapy consolidation was provided, and the tumor responded with complete regression to a flat chorioretinal scar.

COMMENT

Early detection of melanoma is a key component in minimizing risk for metastasis and ensuring overall better prognosis for the patient.[9] In many instances, it is challenging to differentiate a choroidal nevus from a small choroidal melanoma as each can show similar size, color, and related features. In an analysis of 2514 patients, risk factors have been identified to assist in distinguishing benign choroidal nevus from small malignant melanoma.[9] These risk factors have proved critical in the early diagnosis of melanoma and apply only to small choroidal lesions of 3 mm or less in thickness, as in the case described here. These factors include thickness

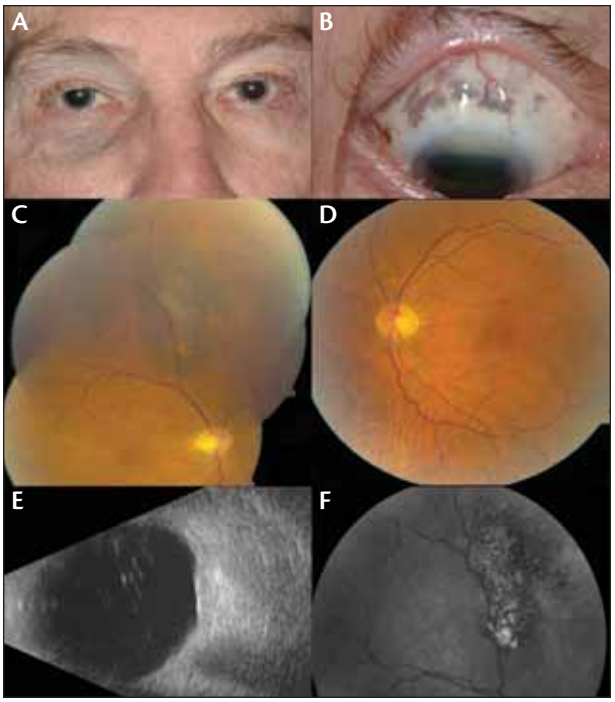

Figure 1. A 72-year-old man with oculodermal melanocytosis showing skin (A) and scleral (B) pigmentation. There was diffuse choroidal melanocytosis in the right eye (C) in comparison with the left (D), and a small choroidal melanoma in the right eye (C) of 2.5 mm thickness (E) was noted within the bed of melanocytosis. There was autofluorescence evidence of overlying lipofuscin orange pigment (F).

greater than 2 mm, related subretinal fluid, symptoms, overlying orange pigment (lipofuscin), tumor margin within 3 mm of the optic disc, ultrasonographic hollowness, absence of surrounding halo, and absence of overlying chronic drusen.[9] Our patient displayed 6 of 8 risk factors, implying high likelihood for further growth of the lesion into a larger melanoma.

Several studies have explored the relationship of ocular melanocytosis to the development of uveal melanoma in Caucasians.[6,7] Patients with melanocytosis are at greater risk for development of uveal melanoma than unaffected Caucasians. It has been estimated that 1 in 400 whites with ocular melanocytosis will develop uveal melanoma during their lifetime, and that development of melanoma is at slightly younger age than unaffected whites.[7]

Furthermore, ocular melanocytosis can promote risk for multifocal uveal melanoma, mostly 2 tumors and rarely 3 tumors in 1 eye.[10] There is a subset of melanocytosis that involves only a geographic portion of the eye, termed partial melanocytosis. The involved portion still carries the risk for melanoma, even though the entire eye is not affected. In an evaluation of 89 eyes with partial ocular melanocytosis, uveal melanoma was noted in 7 cases.[11] In each case, the melanoma originated within the region of ocular melanocytosis.[11] This further supports the link between melanocytosis and uveal melanoma.

It is advised that patients with oculodermal melanocytosis be monitored closely (twice yearly) for development of uveal melanoma. As in our case, many patients with a birthmark are unaware of its significance and hence unaware of the need for monitoring. Additionally, many patients remain asymptomatic despite the presence of a malignancy, as seen in our case, so the role of routine screening by the ophthalmologist is critical. ■

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