Choroidal neovascularization in an eye with foveal hypoplasia associated with albinism

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An association of foveal hemorrhage or choroidal neovascularization with oculocutaneous albinism is very rarely reported.[1] The associated features such as foveal hypoplasia, high myopia, or presence of posterior staphyloma that could lead to suboptimal vision are one of the reasons that these patients are either underdiagnosed or present at a very late stage of scarring. We report a case of choroidal neovascularization associated with albinism where multimodal imaging helped us in conforming the diagnosis.

A 54-year-old man reported with chief complaints of seeing a dark spot in central vision in the right eye for 5 days. On examination he had depigmented skin with white hair and eyebrows. He had horizontal nystagmus with 40° exotropia in the right eye. His best-corrected visual acuity was 20/600, N18 (−7.0 D) in the right eye and 20/125, N10 (−3.0D) in the left eye. Anterior segment showed depigmented irides with nuclear sclerosis grade 2 cataract in the right eye; the left eye was pseudophakic. Fundus examination revealed

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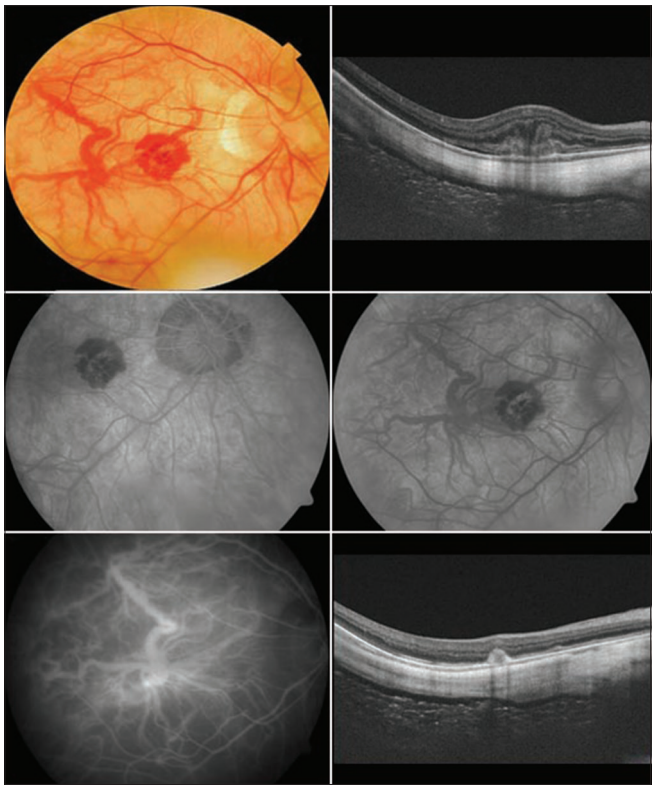

Figure 1: (Top left) Color fundus photo of the right eye. The choroidal vessels are prominent at the posterior pole with presence of subfoveal hemorrhage. (Top right) OCT at presentation shows hyperreflective lesion in subretinal space, suggestive of type 2 CNVM with generalized thinning of the choroid. (Middle) Fluorescein angiography. (Middle left) Early phase shows blocked fluorescence due to hemorrhage and (Middle right) late phase shows minimal hyperfluorescence due to CNVM. (Bottom left) ICG angiography shows dilated ampullae of vortex veins with late leakage in the area of CNVM. (Bottom right) Post intravitreal anti-VEGF injection OCT shows resolution of hemorrhage with the formation of subretinal scar

albinotic depigmented fundi in both the eyes with subretinal bleed in the macular area in the right eye [Fig. 1, top left]. Fundus fluorescein angiography (FFA) and indocyanine green angiography (ICGA) in the early phase showed block fluorescence and late phase showed minimal hyperfluorescence in the area of bleed suggestive of choroidal neovascular membrane (CNVM) [Fig. 1, middle and bottom left]. The optical coherence tomography (OCT) in the right eye revealed subretinal hyperreflective lesion with generalized thinning of the choroid and absence of foveal pit in the left eye [Fig. 1, top right]. He received one intravitreal injection of 1.25 mg bevacizumab in the right eye. One month following the injection the right eye vision improved to 20/125, N10; there was resolution of macular bleed with formation of subretinal scar with no evidence of subretinal fluid on OCT [Fig. 1, bottom right]. The total duration of follow-up was 3 months.

Discussion

Oculocutaneous albinism is a congenital abnormality due to disorder in melanin production manifesting in nystagmus, hypopigmented iris, depigmented fundi, foveal hypoplasia, strabismus, and high myopia.[2] A foveal hemorrhage in oculocutaneous albinism is relatively rare. Shinno *et al*.[1] have reported a FFA and ICGA confirmed case of CNVM. We confirmed the presence of CNVM by retinal imaging, though the etiology was uncertain. This CNVM could be either due to break in Bruch's membrane secondary to high myopia[3] or alterations in the foveal architecture secondary to hypoplasia. In our case although lacquer crack was not seen, a refractive error of −7 D as well as presence of posterior staphyloma made a strong case in favor of high myopia as the cause for CNVM. Despite myopia and myopic astigmatism accounting for 60–70% of refractive error in patients with oculocutaneous albinism,[4] and the incidence of myopic CNV is around 10% in high myopes,[5] foveal hemorrhage or CNVM in albinism has not been reported often.[3] This could be attributed to various reasons, such as presence of depigmented fundi, lack of contrast, and structural alterations in foveal anatomy, that is, hypoplasia. We noticed a generalized atrophy of choroid on OCT, which could be an early indicator for development of CNVM. FFA and OCT help in accurate diagnosis where clinical examination alone may not be enough. Similar to other causes of CNVM, anti-vascular endothelial growth factor (VEGF) injection therapy helps. Our follow-up was limited to 3 months only. While there is a possibility of recurrence, the chance in this case is relatively remote as the lesion was already a scar. All the same, a longer and periodic follow-up is mandatory.

This case report highlights the need for multimodal imaging in case of oculocutaneous albinism as CNVM due to high myopia can be missed due to late presentation in view of poor visual acuity and lack of contrast in the fundus.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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