Bilateral isolated choroidal melanocytosis with hypopigmented posterior pole

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Key words: Choroidal melanocytosis, hyperpigmentation, hypopigmentation

The presence of choroidal melanocytic hyperpigmentation without any associated scleral or skin pigmentation was first defined by Ausburger *et al*.[1] as "isolated choroidal melanocytosis" in 11 Caucasian individuals. Isolated choroidal melanocytosis must be evaluated closely; the differential diagnoses include ocular or oculodermal melanocytosis, choroidal nevus, melanoma, bilateral diffuse uveal melanocytic proliferation, and systemic conditions, such as Waardenburg syndrome. Only a few cases have been reported since,[2,3] and most reports focused mainly on choroidal hyperpigmentation itself, concentrating on the range, shape, and bilaterality of pigmentation. Herein, we present a unique case of bilateral isolated choroidal melanocytosis with hypopigmented posterior pole.

Ultrawide-field color photographs [Fig. 1] showed central hypopigmentation with peripheral choroidal hyperpigmentation around the 360° field in both eyes. Due to the depigmentation, both posterior fundi appeared yellow-orange as in Vogt-Koyanagi-Harada disease. Fluorescein angiography [Fig. 2] and fundus autofluorescence [Fig. 3] were normal and no hyperfluorescence was found in pigmented lesions. Optical coherence tomography [Fig. 4] showed normal structures of the retinal pigment epithelial layer, choroid, and retina. Given the rarity of hypopigmentation among Asians, our case is unique, as peripheral choroid hyperpigmentation and central hypopigmentation existed simultaneously.

Discussion

Two possible hypothetical explanations can be proposed for our patient's funduscopic findings. Autoimmunity offers one explanation. Similar to what is seen in Vogt-Koyanagi-Harada disease, choroidal hyperpigmentation may have triggered

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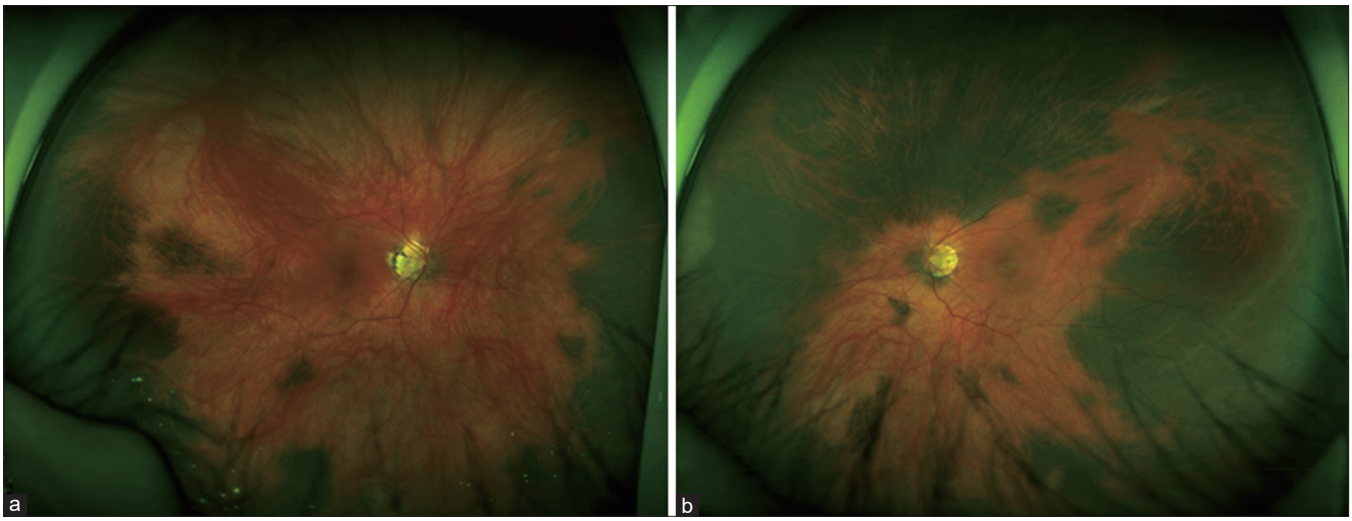

Figure 1: Ultrawide-field color photographs of the right (a) and left (b) eyes show peripheral, flat, and diffuse choroidal hyperpigmentation around the 360° field

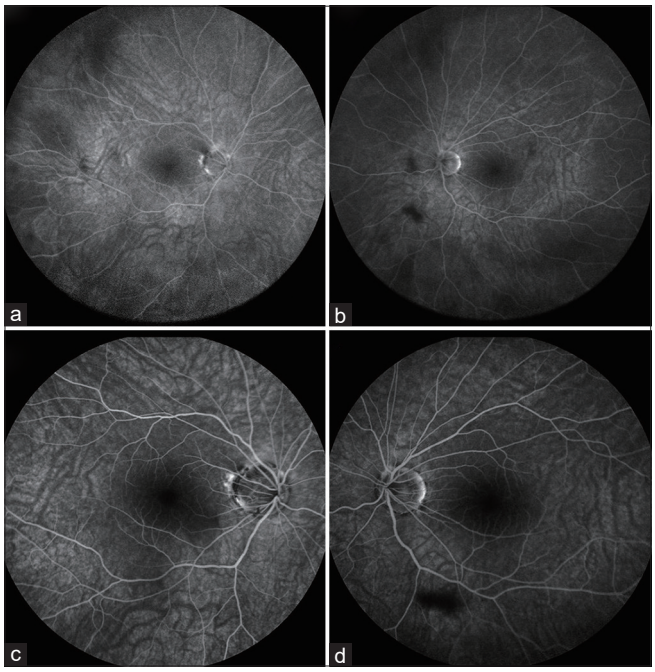

Figure 2: Fundus fluorescein angiography of the 102°-field (a: right eye, b: left eye) and 55°-field (c: right eye, d: left eye) showing a normal angiographic pattern

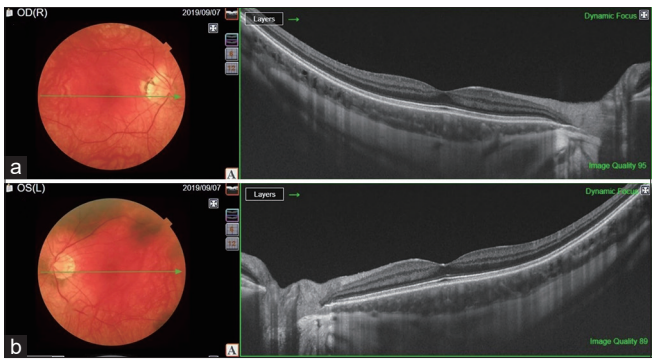

Figure 4: Color fundus photographs and optical coherence tomography images of the right (a) and left (b) eyes demonstrate hypopigmented fundus with normal retinal and choroidal structures

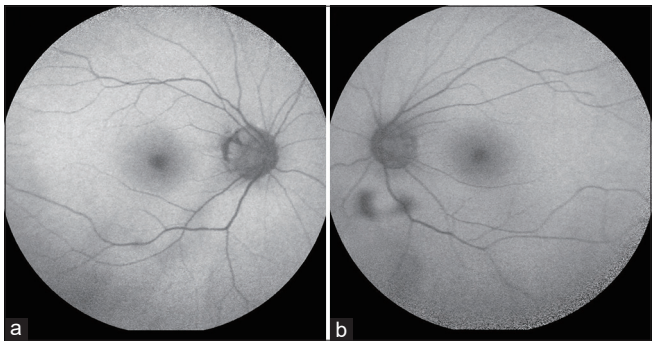

Figure 3: Fundus autofluorescence images of the right (a) and left (b) eyes reveal normal autofluorescence distribution but vitreous floaters are detected

an autoimmune response to melanocytes, leading to hypopigmentation at the posterior pole. Another possibility is racial differences in choroidal pigmentation and melanoma incidence. Hypopigmented fundus and choroidal melanoma are more prevalent among Caucasians than Asians.[4] Our patient may have pigmentary features similar to that of Caucasians; therefore, hyperpigmentation and hypopigmentation could coexist.

Moreover, choroidal melanocytosis may be a risk factor for malignancy,[5] similar to ocular or oculodermal melanocytosis. Our case highlights the need for further research to better understand this condition.

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Conflicts of interest

There are no conflicts of interest.

References

1. Augsburger JJ, Trichopoulos N, Correa ZM, Hershberger V. Isolated choroidal melanocytosis: A distinct clinical entity? Graefes Arch Clin Exp Ophthalmol 2006;244:1522-7.
2. Fine HF, Brue C, Eandi C, Jacobs MI, Pulitzer M, Yannuzzi LA. Bilateral isolated choroidal melanocytosis. Retin Cases Brief Rep 2009;3:272-4.

3. Heinz KD, Demirci H, Elner VM. Bilateral annular isolated choroidal melanocytosis. Ophthalmic Surg Lasers Imaging Retina 2019;50:e74-e76.
4. Kaliki S, Shields CL, Shields JA. Uveal melanoma: Estimating prognosis. Indian J Ophthalmol 2015;63:93-102.
5. Shields CL, Kaliki S, Livesey M, Walker B, Garoon R, Bucci M, *et al*. Association of ocular and oculodermal melanocytosis with the rate of uveal melanoma metastasis: Analysis of 7872 consecutive eyes. JAMA Ophthalmol 2013;131:993-1103.