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 hypopigmentation 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 hypopigmentation may have triggered...
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 3: Fundus autofluorescence images of the right (a) and left (b) eyes reveal normal autofluorescence distribution but vitreous floaters are detected.

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.

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. Our patient may have pigmented features similar to that of Caucasians; therefore, hyperpigmentation and hypopigmentation could coexist.

Moreover, choroidal melanocytosis may be a risk factor for malignancy, 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.

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The unique association of two lesser known entities like choroidal osteoma (CO) and IgG4 inflammatory disease is highlighted in patients presenting with recurrent posterior scleritis.

IgG4-related ocular inflammation is characterized by IgG4 plasma cells and IgG4-associated disease with recurrent posterior scleritis is a rare entity. Choroidal osteoma (CO) may be secondary to inflammatory etiologies like idiopathic orbital inflammation and sclero-uveitis.

Citation:

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