PERIPHERAL EXUDATIVE HEMORRHAGIC CHORIORETINOPATHY WITH POLYPS

Lesions that mimic choroidal melanoma may actually belong to a spectrum of peripheral neovasculopathy.

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Peripheral exudative hemorrhagic chorioretinopathy (PEHCR) is a degenerative condition of the peripheral fundus seen in elderly patients.[1-4] PEHCR is characterized by variable amounts of subretinal or sub–retinal pigment epithelium (sub-RPE) hemorrhage and fluid admixed with subretinal exudation, most commonly located in the temporal periphery.[1] The sub-RPE hemorrhage can appear as a dome-shaped mass mimicking the features of choroidal melanoma. In fact, PEHCR is second only to choroidal nevus among lesions that simulate choroidal melanoma.[5] While the exact etiology of PEHCR is unclear, the condition has been described as a peripheral variant of age-related macular degeneration (AMD) and, more recently, as a peripheral manifestation of idiopathic polypoidal choroidal vasculopathy (PCV).[4,6-8]

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AT A GLANCE

- Characterized by subretinal or sub-RPE hemorrhage and fluid admixed with subretinal exudation, PEHCR has been described as a peripheral variant of AMD and as a peripheral manifestation of idiopathic PCV.
- In a patient referred for possible choroidal melanoma, clinical findings were consistent with PEHCR with polyps and no evidence of melanoma.
- After observation for 4 months, the lesion demonstrated marked improvement with resolution of RPE detachment and hemorrhage.

Mantel et al described similarities between PEHCR and PCV in 2009 and followed with a case series of PEHCR in 2012, describing polyp-like changes seen on indocyanine green angiography (ICGA).[3,4] Others have similarly reported polyp-like changes in PEHCR on ICGA and have suggested that PEHCR represents a spectrum of peripheral neovasculopathy that includes lesions with and without polyps.[7,8] Herein, we report a case of PEHCR *with* polyps, seen on ICGA, in a patient referred for suspected choroidal melanoma.

CASE REPORT

A 76-year-old African-American woman was referred for possible choroidal melanoma in her right eye (OD). The

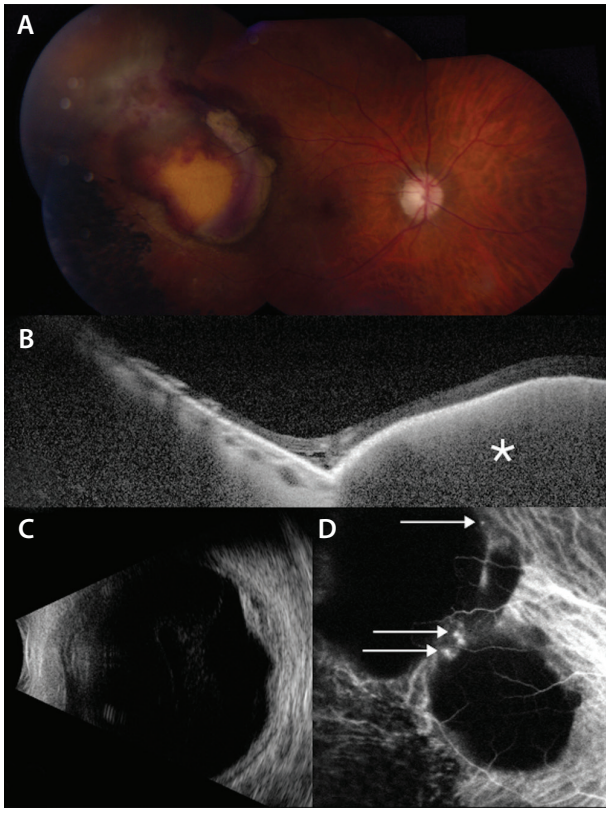

Figure 1. Abruptly elevated hemorrhagic lesion located in temporal quadrant with inferior pigmentary changes. The red, yellow, and gray areas represent different stages of subretinal and sub-RPE hemorrhage (A). OCT demonstrated the abruptly elevated RPE detachment, indicated with an asterisk (B). B-scan ultrasonography OD showed an elevated lesion measuring 3.3 mm with heterogeneous acoustic reflectivity (C). ICGA demonstrated multiple polyp-like hypercyanescent foci (arrows) adjacent to two areas of blocked cyanescence representing hemorrhage (D).

patient reported chronic floaters in each eye (OU) and long-term blurred vision in her left eye (OS), attributed to previous branch retinal vein occlusion. Her medical history included congestive heart failure and systemic hypertension. Her family history included sarcoidosis in a nephew.

On examination, the patient's visual acuity was 20/40 OD and 20/100 OS with symmetric pupillary responses OU and normal intraocular pressure OU. External examination and ocular motility were normal. Anterior segment examination was unremarkable apart from bilateral pseudophakia. Dilated fundus examination OS revealed mild optic nerve pallor, epiretinal membrane, and a small flat choroidal freckle at the inferior optic disc margin. Fundus evaluation OD showed an elevated bilobular lesion in the temporal periphery with a mixed coloration of central

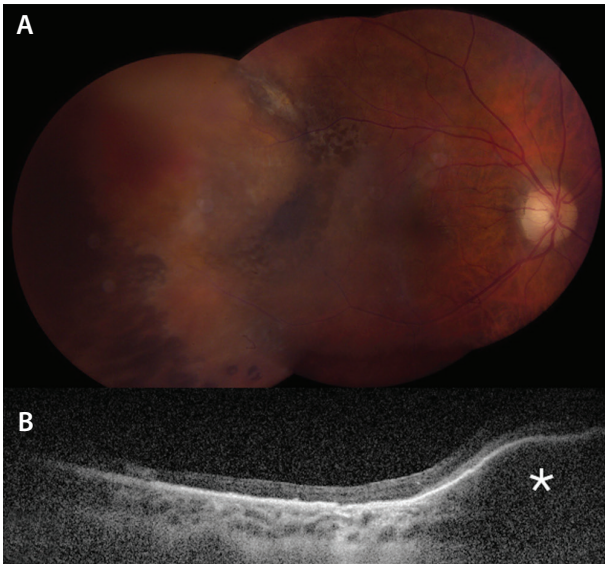

Figure 2. At 4-month follow-up, the mass has partially resolved (A) and the RPE detachment, indicated with an asterisk, has decreased in thickness (B).

orange hue and circumferential gray rim with overlying red hemorrhage. The mass measured 12 mm x 8 mm in base and 3.3 mm in thickness (Figure 1). The borders were geographic and abrupt, lined with pigment presumably of RPE origin or from an old hemorrhage. The lesion demonstrated fresh and old hemorrhage in both the subretinal and sub-RPE layers. There was diffuse peripheral RPE mottling inferiorly OU.

B-scan ultrasonography showed a bilobed, mostly echodense intraocular mass OD with heterogeneous acoustic reflectivity. Optical coherence tomography (OCT) of the mass showed abrupt elevation of the RPE, suggestive of RPE detachment. On fluorescein angiography (FA), the mass revealed nearly absolute hypofluorescence, consistent with blockage from hemorrhage, and there was no double circulation of melanoma. On ICGA, there were two large hypocyanescent areas bordered by several hypercyanescent choroidal polyps.

These findings were consistent with PEHCR with polyps, with no evidence of melanoma. Observation was advised. At 4-month follow-up, the lesion demonstrated marked improvement, with resolution of RPE detachment and hemorrhage measuring only 1.9 mm on ultrasonography (Figure 2).

DISCUSSION

In a large cohort of 173 eyes with PEHCR referred to an ocular oncology center for possible choroidal melanoma, the mean patient age was 80 years, and 99% of patients were Caucasian.[2] The most common symptoms were decreased

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visual acuity (37%) and flashes or floaters (20%). The lesions were located most often in the temporal quadrant (77%), with average measurements of 10 mm in base and 3 mm in thickness. Fundus features included subretinal hemorrhage (78%), subretinal exudation (21%), serous RPE detachment (28%), hemorrhagic RPE detachment (26%), and vitreous hemorrhage (24%). FA performed in 61 eyes revealed patchy blockage of choroidal fluorescence related to subretinal hemorrhage, sub-RPE hemorrhage, or RPE hyperplasia. B-scan ultrasonography typically demonstrated a dome- or plateau-shaped lesion with variable intrinsic acoustic quality. ICGA findings were not reported in this cohort.[2]

There has been a debate over recent years regarding the etiology of PEHCR. Some have proposed that PEHCR represents a peripheral version of classic AMD, while others assert that PEHCR is a peripheral manifestation of idiopathic PCV.[4,6-8] In the largest reported cohort of PEHCR to undergo FA, only 3% of eyes were noted to have peripheral choroidal neovascular membrane (CNVM); however, there may have been masking of the CNVM by hemorrhage.[2] In that cohort, 8% to 27% of patients had macular changes consistent with AMD (ie, drusen, RPE alterations, CNVM). In another large cohort of patients with PEHCR, possible peripheral choroidal neovascularization was seen on FA in 65% of cases, and 69% of those patients demonstrated macular changes consistent with AMD.[3] While the suggestion that PEHCR represents a peripheral variant of AMD seems intuitive based on clinical features, there are other hypotheses.

In 2009, Mantel et al suggested that PEHCR exhibited features similar to idiopathic PCV, specifically, irregular choroidal vascular networks.[3] In 2012, Mantel et al reported a consecutive series of 40 patients with PEHCR who were imaged with ICGA. Of those patients, 69% showed polyp-like changes and 50% showed irregular choroidal vascular networks.[4] Others have confirmed polyp-like changes on ICGA in PEHCR.[7,8] In a report of 10 patients with PEHCR, Goldman et al confirmed polyp-like changes on ICGA and suggested, based on an evolving understanding of PCV, that the polyps found in PEHCR are caused by type 1 neovascularization (sub-RPE CNVM).[8] They suggested that PEHCR exists as a spectrum of peripheral neovasculopathy that includes disease with and without polyps.[8]

CONCLUSION

The patient described herein demonstrated a bilobed form of PEHCR, simulating multilobulated choroidal melanoma. This hypofluorescent and hypocyanescent hemorrhagic mass exhibited tiny polyps on ICGA and demonstrated spontaneous involution without treatment. PEHCR can closely mimic choroidal melanoma in location, configuration, and color. Conversely, it should be noted that some choroidal melanomas, especially those with breaks in Bruch membrane, can display extensive subretinal hemorrhage, hiding an underlying melanoma and resembling PEHCR. Based on published reports, PEHCR most likely represents a spectrum of peripheral neovasculopathy demonstrating similarities to both AMD and PCV. ■

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