

# Peripheral Exudative Hemorrhagic Chorioretinopathy Simulating Choroidal Melanoma

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Peripheral exudative hemorrhagic chorioretinopathy (PEHCR) is a degenerative condition of the peripheral retina that tends to occur as a bilateral, often symmetric process in elderly patients.<sup>1,2</sup> There are only a few reports on PEHCR in the literature.<sup>1-4</sup> Some authorities consider PEHCR as the peripheral version of age-related macular degeneration while others believe PEHCR is a specific variant of polypoidal choroidal vasculopathy (PCV).<sup>4</sup>

The clinical features of PEHCR include retinal pigment epithelial (RPE) mottling or atrophy in the periphery, RPE detachment (serous or hemorrhagic), subretinal fluid or hemorrhage, subretinal exudation, and, rarely, vitreous hemorrhage.<sup>1</sup> The spectrum of these features ranges from mild RPE abnormalities to extensive exudative retinopathy threatening the fovea from the periphery.

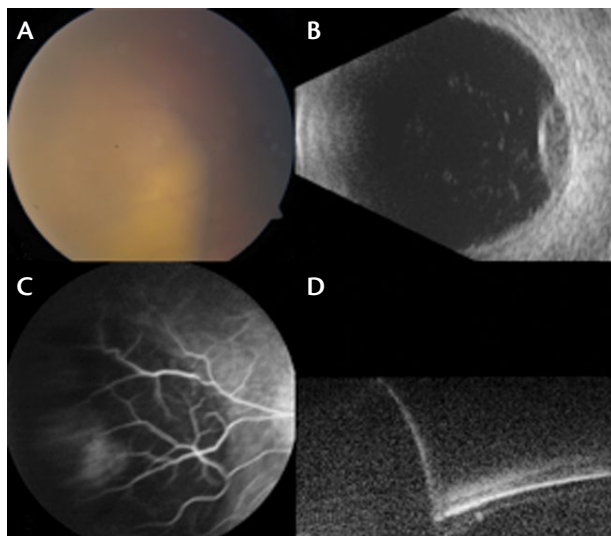
Another major concern is that PEHCR has clinical similarities to uveal melanoma. Most often, the hemorrhagic pigment epithelium detachment (PED) appears as a dome-shaped mass of similar size, location, and configuration as a peripheral choroidal melanoma. With time, the PED demonstrates slow involution, confirming the diagnosis of PEHCR and not melanoma. However, at the time of diagnosis, the pseudomelanoma picture can lead to diagnostic confusion. In fact, in a report on uveal melanoma in 12 000 patients, it was found that 1739 (14%) of patients referred for melanoma were actually found to have pseudomelanoma.<sup>5</sup>

The 5 leading choroidal pseudomelanomas include choroidal nevus (49%), PEHCR (8%), congenital hypertrophy of the RPE (6%), hemorrhagic detachment of the retina or pigment epithelium (5%), and circumscribed choroidal hemangioma (5%).<sup>5</sup> Herein, we describe a patient referred for melanoma management in his only eye, who was found on examination to have a pseudomelanoma, namely PEHCR. In this report, we highlight the features that distinguish these 2 clinical entities.

## CASE REPORT

A 78-year-old white man with a history of enucleation of the left eye (OS) for glaucoma from epithelial downgrowth following cataract surgery was found on routine examination to have a choroidal mass in his right eye (OD). His retina consultant advised evaluation by the Ocular Oncology Service at Wills Eye Hospital for suspected choroidal melanoma.

On examination, best corrected visual acuity (BVCA) was 20/30 OD. The left socket was clean and the prosthesis was well-fitted. The findings were limited to OD. The anterior segment revealed pseudophakia following cataract surgery. Fundus examination disclosed few macular drusen. In the temporal periphery was an abruptly elevated yellow homogeneous subretinal mass measuring 12 mm in basal diameter (Figure 1A). Subretinal hemorrhage was noted on the anterior margin. Transillumination failed to cast a shadow. B-scan



**Figure 1.** Abruptly elevated mass located in the temporal periphery of the right eye (A). B-scan ultrasonography of right eye showing 3.4 mm thick acoustically hollow mass (B). Fluorescein angiogram of right eye demonstrating hypofluorescence of the mass with mild central hyperfluorescence (C). Optical coherence tomography of right eye showing abrupt elevation of the retinal pigment epithelium (to the left) (D).

ultrasonography disclosed an echolucent mass 3.4 mm in thickness (Figure 1B). Fluorescein angiography (FA) demonstrated hypofluorescence of the mass with a focus of central hyperfluorescence, suggestive of choroidal neovascularization (CNV; Figure 1C). Optical coherence tomography showed an abrupt elevation consistent with RPE detachment (Figure 1D). These features were consistent with PEHCR with related hemorrhagic PED rather than choroidal melanoma. Observation was advised, and the PED showed slow involution.

## DISCUSSION

PEHCR is not well-recognized by the practicing ophthalmologist and persists as a major simulator of choroidal melanoma. The similarities between these 2 conditions relates to the clinical features of serous or hemorrhagic PED with PEHCR and the circumscribed melanocytic features of melanoma. Both conditions can show a smooth dome-shaped configuration, dark brown pigmentation, and surrounding serous retinal detachment.

Shields et al<sup>2</sup> described a large cohort of 146 patients with PEHCR who were referred to an ocular oncology center with possible choroidal melanoma. In that series, the mean patient age was 80 years and 99% of patients were white. The “tumor” was located most often in the temporal quadrant (77%) and measured 10.1 mm in

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In this monocular patient, the pigment epithelium detachment slowly resolved over time, requiring no intervention.

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base and 3 mm in thickness. The authors describe clinical features that differentiate these 2 entities. PEHCR was often located peripherally, between the equator and the ora serrata (89%).<sup>2</sup> In contrast, choroidal melanoma was most commonly found between the macula and the equator (78%).<sup>6</sup> PEHCR often involved 1 or more quadrants, whereas most melanomas affected less than 1 quadrant.<sup>2</sup> Macular or extramacular degeneration was typically seen in the eye affected with PEHCR or the contralateral eye, with features of drusen, RPE alterations, or CNV.<sup>2</sup> PEHCR-related serous or hemorrhagic PED or retinal detachment occasionally occurred bilaterally, implying age-related degeneration. Choroidal melanoma is nearly always unilateral. Finally, transillumination of the globe in PEHCR can show a shadow, but it generally abruptly ends at the ora serrata whereas melanoma can show shadow that extends into the ciliary body or iris.

Mantel and associates described clinical and angiographic features of 56 eyes in 45 patients diagnosed with PEHCR, most of whom were elderly at mean age of 77 years.<sup>3</sup> They noted a predominance of white (100%) and female (69%) patients. In that series, the most common reason for referral was suspected choroidal neoplasm (87%).<sup>3</sup> These results were consistent the earlier observations by Shields and associates.<sup>2</sup>

PEHCR classically manifests with symptoms of decreased visual acuity (37%) and flashes/floaters (20%).<sup>2</sup> Related features include subretinal hemorrhage (78%), subretinal exudation (21%), serous PED (28%), sub-RPE hemorrhage (26%), and vitreous hemorrhage (24%).<sup>2</sup> B-scan ultrasonography typically demonstrates a dome- or plateau-shaped “mass” with hollow acoustic quality. Intravenous FA shows patchy blockage of choroidal fluorescence at the site of hemorrhage, diffuse filling in serous PED, and possible PCV.

In our case, the similarity of PEHCR with choroidal melanoma was evident. The findings on diagnostic testing strongly suggested PED rather than melanoma. In this monocular patient, the PED slowly resolved over time, requiring no intervention. ■

Support provided by Eye Tumor Research Foundation, Philadelphia, PA (CLS). The funders had no role in the design and conduct of the study, in the collection, analysis and interpretation of the data, and in the preparation, review or approval of the manuscript. Carol L. Shields, MD, has had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

No conflicting relationship exists for any author. ■

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