Asymptomatic Temporal Mass with Submacular Fluid in an 86-Year-Old Male

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Introduction:

An 86-year-old male was referred from his optometrist with report of peripheral retinal mass in the right eye. The patient had a history of Alzheimer's and reported no symptoms.

Figure 1: Temporal subretinal fibrosis and hemorrhage of the right eye.

Exam:

Visual acuity was 20/30 in the right eye and 20/40 in the left. Anterior segment examination was unremarkable with an intraocular pressure of 15 in both eyes.

Post segment examination was notable for fine macular drusen bilaterally. In the temporal macula of the right eye there was shallow subretinal fluid, tracking from an area of subretinal fibrosis and hemorrhage in the temporal periphery (Figure 1). The peripheral exams of the left eye and remainder of the right eye were unremarkable.

OCT imaging of the right macula confirmed the presence of subretinal fluid extending to the fovea (Figure 2). Fluorescein angiography demonstrated blockage in the area of the retinal hemorrhages and hyperfluorescence suggestive of a neovascular membrane adjacent to the areas of fibrosis (Figure 3).

Management:

Our patient was diagnosed with peripheral exudative hemorrhage chorioretinopathy (PEHCR). Given the presence of subfoveal fluid the patient was treated with intravitreal bevacizumab . At 6-week follow-up the submacular fluid had improved and additional bevacizumab was administered. At 3-month follow-up the macular fluid was resolved and remained so at 6-month follow-up without additional treatment (Figure 4).

Discussion:

Known also as eccentric disciform, extramacular disciform, and hemorrhagic peripheral pigment epithelium disease, PEHCR received its name in a 1980 case series documenting 32 lesions in 27 eyes.[1] In that series PEHCR lesions were characterized by the presence of peripheral blood in the subretinal and/or sub-RPE space with or without exudates.

PEHCR is a condition of elderly, Caucasian patients with a preponderance of female presentations. Shields CL et al, in reporting the largest PEHCR case series with 173 lesions, noted a majority of lesions appeared temporally between the equator and ora serrata.[2] Mean lesion diameter was 10mm and mean height was 3mm. Bilateral lesions were present in one-third of patients. Appearance and coloration of the lesions varied widely with the chronicity of hemorrhage and with the

presence or absence of exudates and fibrosis.

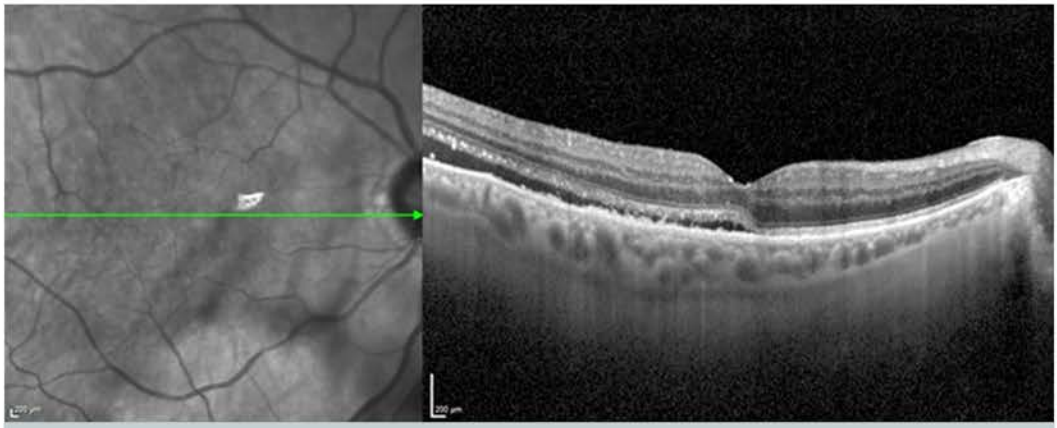

Figure 2: Shallow subretinal fluid emanated from the temporal lesion into the macula of the right eye.

Despite often culminating in disciform lesions reminiscent of age-related macular degeneration (AMD), PEHCR lesions were not always found in conjunction with macular AMD pathology. Approximately half of all presentations demonstrated drusen, RPE pigmentary changes, or choroidal neovascular membranes in the ipsilateral or contralateral macula. Peripheral drusen and RPE changes were found more frequently, seen in approximately two-thirds of ipsilateral eyes with PEHCR lesions.[2]

Often PEHCR lesions are referred to an ocular oncologist given concern for intraocular malignancy, particularly choroidal melanoma. Shields JA et al, in reporting on over 12,000 referrals for choroidal melanoma, reported 1,739 cases (14%) to be mimicking conditions, termed pseudomelanomas.[3] Of these pseudomelanomas PEHCR represented 13% of cases, second only to choroidal nevi (49%).

In distinguishing PEHCR lesions from melanomas the presence and extent of subretinal and sub-RPE hemorrhage favors a diagnosis of PEHCR. Location is helpful as well as PEHCR lesions rarely present posterior to the equator and do not advance anterior to the ora serrata. Transillumination can be a useful tool to identify a melanoma advancing into the pars plana; such extension is not seen in PEHCR lesions.[2]

Ultrasonography is a particularly useful adjunct. Most PEHCR lesions demonstrate echogenicity on B-scan and high internal reflectivity on A-scan, in contrast to the echolucency and low internal reflectivity seen in choroidal melanomas.[2,3] Fluorescein angiography can be useful in identifying the intrinsic vessels of a neoplasm in contrast to the hemorrhage-related blockage or, more rarely, choroidal neovascular membrane of a PEHCR lesion.

Once identified PEHCR lesions are often observed. Shields CL et al noted no progression in 89% of their cases, with the majority regressing to fibrosis and RPE atrophy or hyperplasia. In the minority of PEHCR lesions that progress, causes of reduced vision include breakthrough vitreous hemorrhage and subretinal fluid, such as in our patient. Recent case reports highlight the efficacy of intravitreal anti-VEGF agents in treating these PEHCR complications, as was seen in our patient.[4]

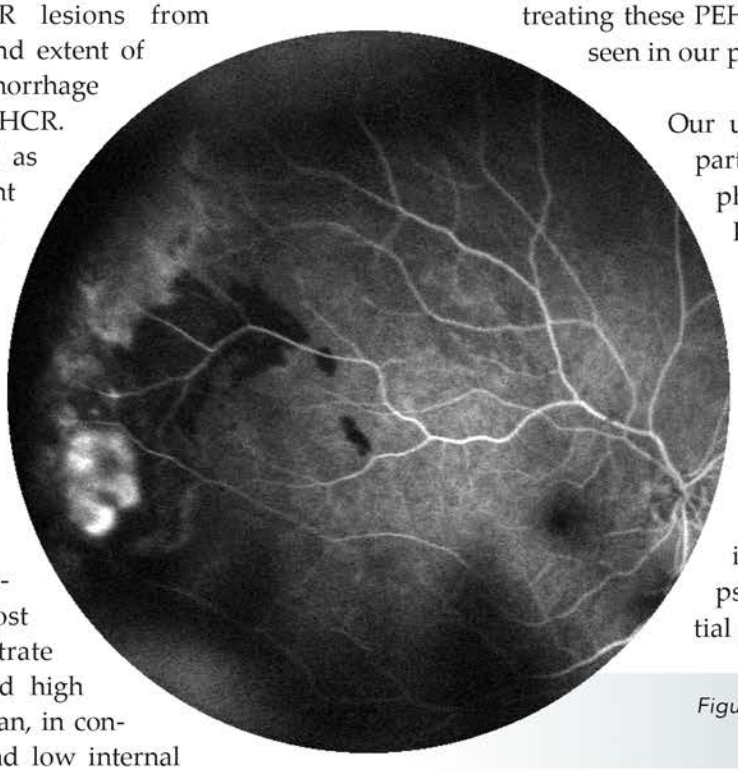

Figure 3: Hyperfluoresence of the lesion on FA.

Our understanding of PEHCR, particularly regarding pathophysiology, is evolving. Recent work has revealed a subtype of PEHCR with polyps on indocyanine green angiography representing a peripheral manifestation of polypoidal choroidal vasculopathy.[5] A complete understanding of this important member of the pseudomelanoma differential diagnosis awaits.

References:

1. Annesley WH. Peripheral Exudative Hemorrhagic Chorioretinopathy. Tr Am Ophth Soc. 1980;78:321-364.

2. Shields CL, Salazar PF, Mashayekhi A, Shields JA. Peripheral Exudative Hemorrhagic Chorioretinopathy Simulating Choroidal Melanoma in 173 Eyes. Ophthalmology 2009;113:529-535.

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4. Sax J, Karpa M, Reddie I. Response to Intravitreal Aflibercept in a Patient with Peripheral Exudative Hemorrhagic Chorioretinopathy. Retin Cases Brief Rep. 2018;0:1-3.

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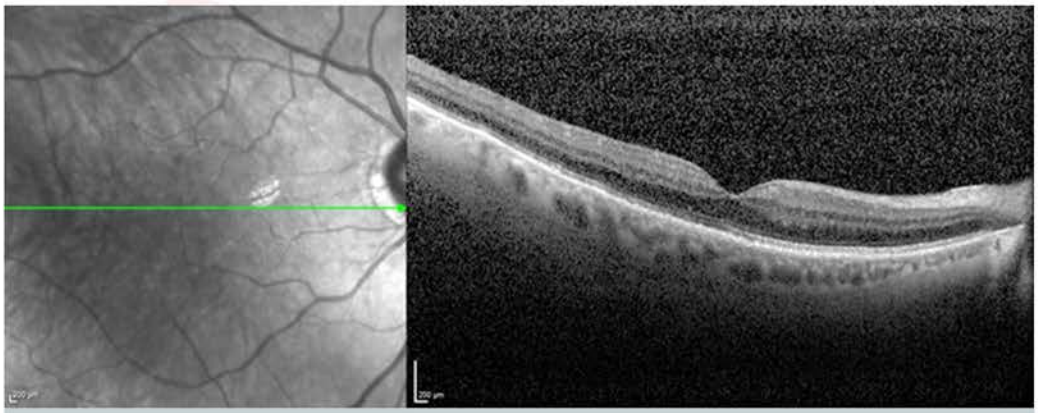

Figure 4: At 6-month follow up, status-post intravitreal bevacizumab, the macula was dry.

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