Vitreous hemorrhage as the presenting feature of peripheral exudative hemorrhagic chorioretinopathy in Indian eyes

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Five Indian patients presenting with unilateral vitreous hemorrhage (VH) underwent pars plana vitrectomy (PPV) and were found to have lesions suggestive of peripheral exudative hemorrhagic chorioretinopathy (PEHCR). All eyes had extensive sub-retinal hemorrhage; three also showed an elevated mass lesion. The temporal retina anterior to the equator was the most commonly affected site. Exudative manifestations were absent in all. No recurrences occurred over a mean follow-up of 17.5 months. Although PEHCR is reported to be rare in Asians, this series demonstrates that it can present as VH in Indians as well. Unless the macula has irreversible damage, a favorable outcome can be obtained with PPV alone.

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Peripheral exudative hemorrhagic chorioretinopathy (PEHCR) is a rare peripheral chorioretinal degenerative disorder characterized by sub-retinal hemorrhage (SRH) and/or sub-retinal pigment epithelium (RPE) hemorrhage that may be accompanied by exudation.[1] Although frequently asymptomatic and associated with a benign outcome, macular involvement or breakthrough vitreous hemorrhage (VH) can impair vision. It is predominantly seen in elderly, Caucasian women, particularly with hypertension or atherosclerotic artery disease.[1] PEHCR has been described rarely in the Asian subcontinent. This report describes five Indian patients who presented with VH and were found to have features of PEHCR.

Case Reports

The demographic data, clinical features, including systemic history, management, and outcomes of the five patients presenting with unilateral VH who underwent pars plana vitrectomy (PPV) are elaborated in Table 1. Fig. 1 depicts case 1 and 2, while Figs. 2 and 3 illustrate case 4 and 5 respectively.

Discussion

PEHCR may be an underdiagnosed entity owing to a frequently asymptomatic clinical course and peripheral location of lesions. The largest series by Shields *et al*. reported 173 eyes with PEHCR and concluded that it can be typically mistaken for choroidal melanoma.[1] Among the patients 99% were Caucasian and a predilection for white race has been confirmed by two other large series.[2,3] PEHCR is extremely rare in Asians and only three isolated cases have been reported from the Indian subcontinent.[4-6] This series elaborates five Indian patients who underwent PPV for VH and were found to have features suggestive of PEHCR. The criteria for diagnosis of PEHCR included peripheral subretinal or sub-RPE blood or exudation or peripheral choroidal neovascularization (CNV). The peripheral fundus was defined as the portion beyond the macula (>3 mm from the foveola).[1]

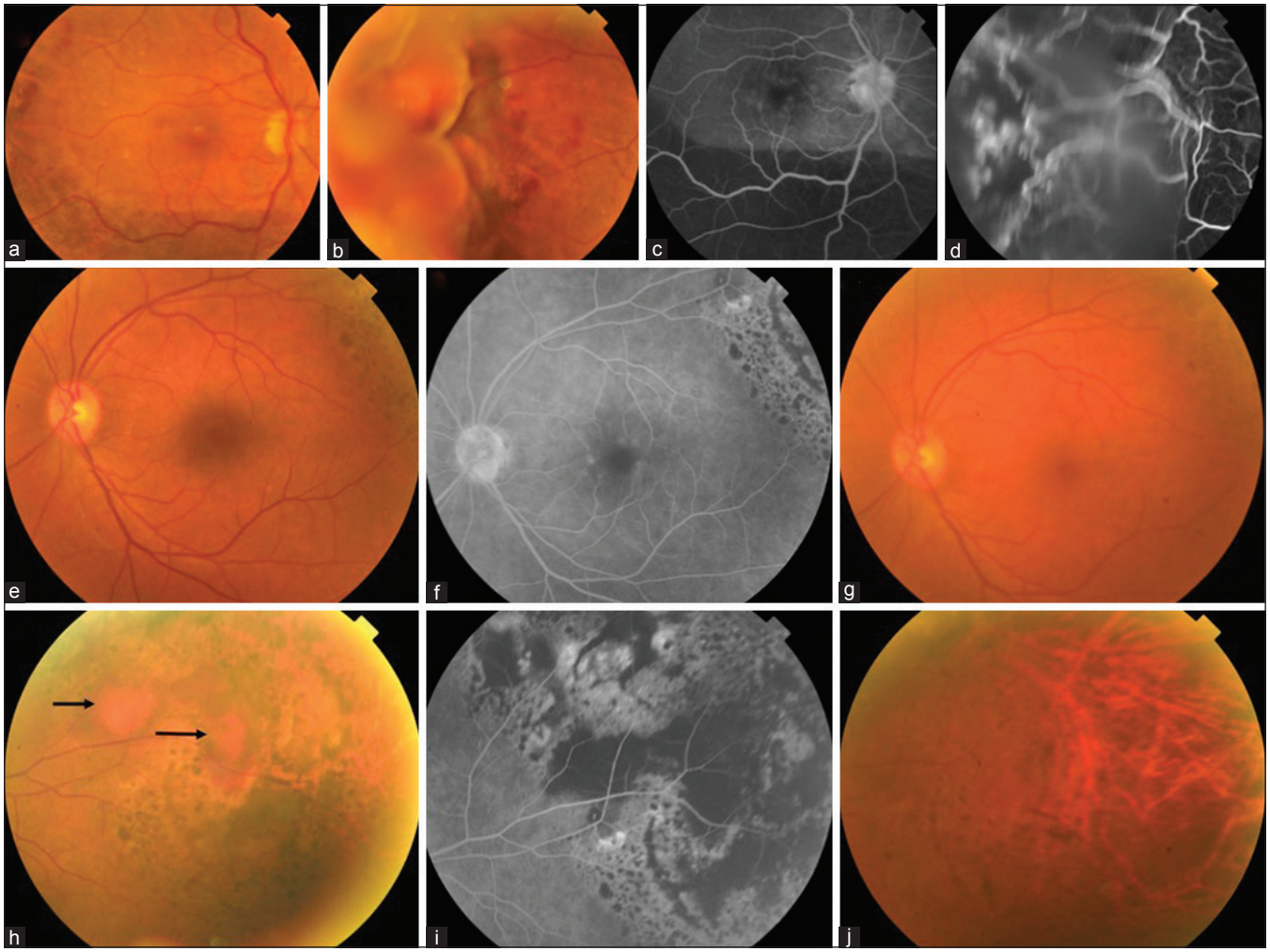

Figure 1: Case 1: (a) Right eye fundus 1 week postoperatively showing altered SRH inferior to the macula. (b) The temporal periphery had elevated mounds of SRH. (c) FA showed blocked fluorescence in the area of SRH with ill-defined leaks near the fovea and temporally. (d) Case 2. (e) Left eye fundus 2 weeks postoperatively showing pigmentation superotemporally with two orange lesions (*black arrows*). (h) FA showed blocked fluorescence (pigment) with areas of staining (f), (i) and no active leaks. Three years follow-up showed marked decrease in pigment with resolution of the orange lesions (g), (j)

Table 1: Demographic data, clinical features, management, and outcomes of the five cases in the study

Age (years)	Sex	Presenting complaints	Systemic history	BCVA (initial)	Clinical features	USG B scan	Fellow eye	Intervention	Intraoperative features	Postoperative imaging	BCVA (final)	Follow -up (months)
82	M	DOV OD × 6 weeks	Nil	HM+	NS grade 2, VH	VH, PVD, SRM (medium to high internal reflectivity) temporally, 7.8 mm diameter, 3.6 mm thickness	OS 6/6, NAD	OD Phaco + IOL + 23G PPV	SRH nasally, inferiorly, temporal elevated mounds of altered SRH	FA-blocked fluorescence (SRH), ill-defined leaks temporally and around fovea	6/9	2
60	M	DOV OS × 6 months	CAD × 2 years, on AG	HM+	Clear lens, VH	VH, PVD, attached retina	OD 6/6, NAD	OS 23G PPV	RPE changes, orange lesions (SRH) ST to macula	FA - blocked fluorescence (SRH + pigment), staining	6/6	36
60	M	DOV OD × 4 months	CAD, on AG	PL+, PR +	VH	VH, PVD	OS 6/6, NAD	OD 23G PPV	SRH superiorly, inferiorly, at the macula, submacular fibrosis	-	CF half meter	1
67	F	DOV OS × 1 month	Hypertension × 3 years	PL+, PR+	Cataract, VH	VH, PVD, SRM (medium internal reflectivity) temporally, 3.1 mm diameter, 2.8 mm thickness	OD 6/18, cataract, drusen	OS Phaco + IOL + 23G PPV	SRH inferiorly, temporal elevated mounds of SRH, RPE alterations at macula	FA-blocked fluorescence (SRH), temporal staining SD-OCT - drusen, scarring	6/24	8
48	F	DOV OD × 6 weeks	Nil	HM+	VH	VH, PVD, SRM (medium to high internal reflectivity) temporally, 5.8 mm diameter, 3.1 mm thickness	OS 6/6, NAD	OD 23G PPV	SRH nasally, inferiorly, temporally, at the macula, SRM along IT arcade	FA-blocked fluorescence (SRH), ill-defined leaks temporally SD-OCT - SRH at the macula just sparing the fovea, hemorrhagic PED along IT arcade	6/9	24

BCVA=Best corrected visual acuity, USG=Ultrasound, M=Male, F=Female, DOV=Diminution of vision, OD=Right eye, OS=Left eye, HM=Hand motions, NS=Nuclear sclerosis, VH=Vitreous hemorrhage, PVD=Posterior vitreous detachment, SRM=Subretinal mass, NAD=No abnormality detected, Phaco=Phacoemulsification, IOL=Intraocular lens, 23G PPV=23 gauge pars plana vitrectomy, SRH=Subretinal hemorrhage, FA=Fluorescein angiography, ST=Superotemporal, IT=Inferotemporal, CAD=Coronary artery disease, AG=Anti-coagulants, RPE=Retinal pigment epithelium, PL=Perception of light, PR=Projection of rays, CF=Counting fingers, PED=Pigment epithelial detachment

Advanced age and female preponderance have been noted to be a typical feature of PEHCR.[1-3] Mean age in this series was 63.4 years (range 48–82 years), lower than that in the literature (range 70–80 years)[1] and males and females were almost equally distributed. Ethnic differences might account for this difference. Two patients were on anticoagulant therapy, a risk factor for massive SRH. Hence normalization of blood pressure and discontinuation or reduction of anticoagulants if medically acceptable is advisable.

All five patients presented with unilateral poor best corrected visual acuity (BCVA) owing to dense VH. Decreased vision from VH caused by PEHCR has been reported to range from 8.9[2] to 14%[1] at baseline, and PPV may be required in up to 7%.[1] Only a single case report of VH as the presenting feature of PEHCR has been reported from the Indian subcontinent.[5] Preoperative ultrasound B scan picked up a mass lesion in three eyes, in addition to VH. Ultrasound findings of solid or hollow acoustic quality have been shown to correspond

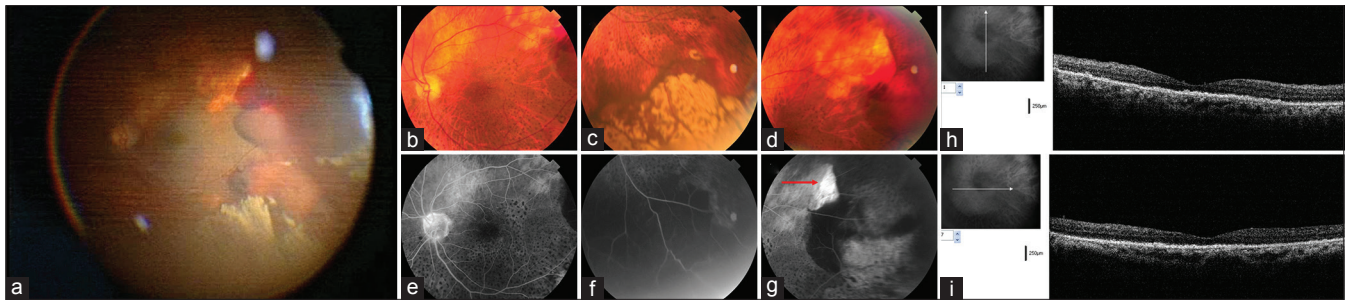

Figure 2: Case 4: (a) Intraoperative photograph of the left eye after vitrectomy showing temporal mounds of SRH and altered SRH inferiorly. Fundus photograph 6 weeks postoperatively showed pigmentary alterations at the macula (b), inferior altered SRH (c), and resolving SRH temporally (d). FA showed blocked fluorescence due to the pigment at the macula (e) and inferiorly (f). A well-defined area of staining was observed temporally (*red arrow*) (g). Vertical (h) and horizontal (i) SD-OCT scans through the macula showed presence of drusen and scarring

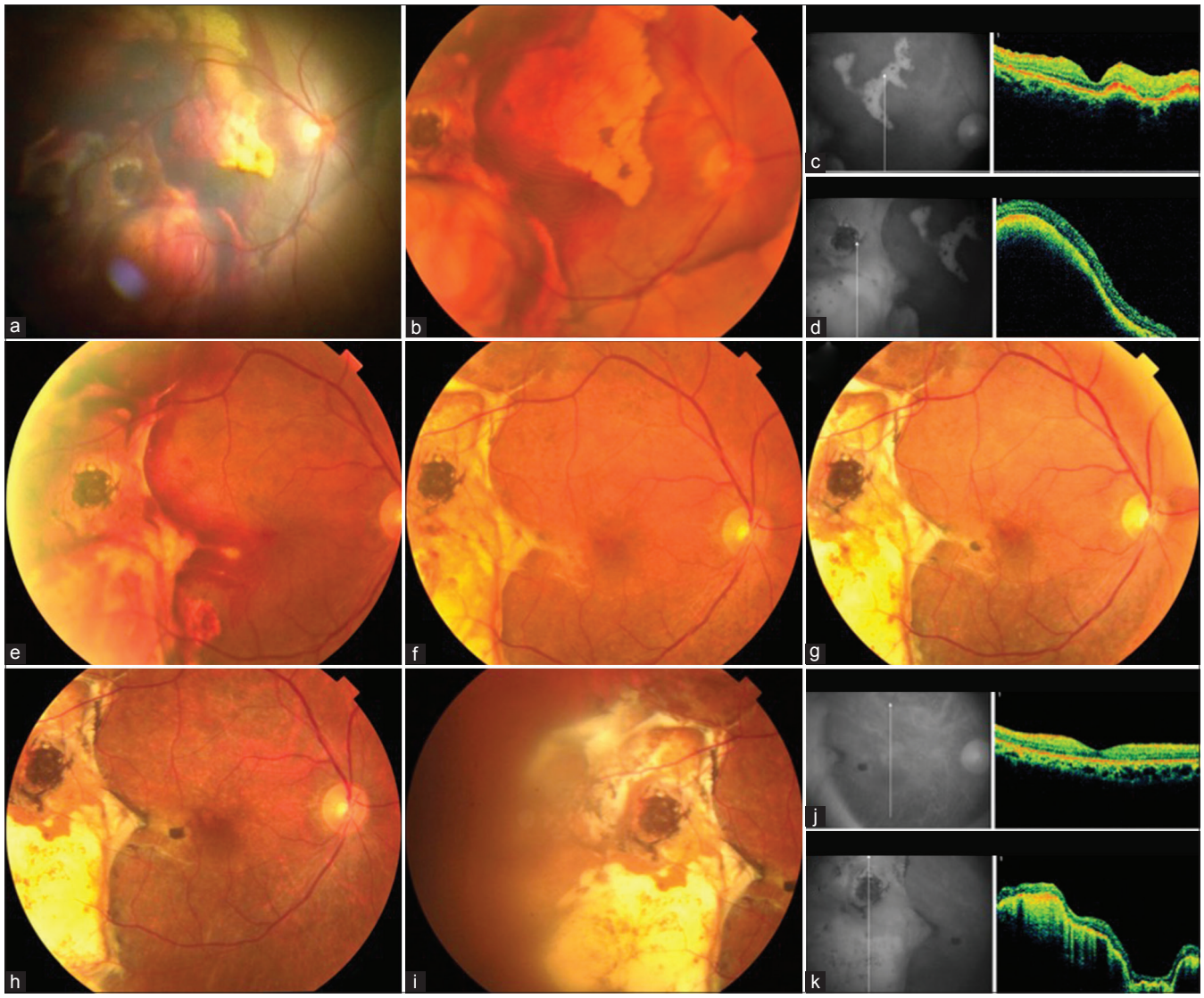

Figure 3: Case 5: (a) Right eye intraoperative photograph showing extensive SRH, an elevated mass inferotemporally and hyperpigmentation temporally. (b) SRH decreased at 1 week follow-up. (c) SD-OCT at 2 weeks showed SRH sparing the fovea. (d) SD-OCT scan through the mass lesion confirmed that it was sub-RPE hemorrhage. Serial fundus photographs at 6 weeks (e), 6 months (f), 1 year (g), and 2 years (h) showing gradual resolution of the SRH and organization of the hemorrhage temporally (i). (j) SD-OCT scan through the macula was normal. (k) SD-OCT scan through the mass lesion showed decrease in height of the hemorrhagic PED

with the clinical features of hemorrhagic and serous pigment epithelial detachment (PED).[2]

Intraoperatively, altered SRH involving two or more quadrants, commonly inferior and temporal, was seen in four

eyes (80%). The temporal retina anterior to the equator is more commonly affected by PEHCR and usually one or two quadrants are involved.[1,2] Hemorrhagic manifestations were the most prominent feature in all eyes, and interestingly lipid exudation was absent in all. Chronic RPE changes (hyperpigmentation and atrophy) have been found in the vicinity of PEHCR[2] and were observed in cases 2 and 3. Macular abnormalities such as drusen, RPE alterations, and CNV might accompany the disease[2] and were encountered in three eyes. In addition, spectral domain optical coherence tomography (SD-OCT) of the macula can reveal subretinal fluid or SRH or sub-RPE hemorrhage. PEHCR may be bilateral in 25% of the cases and may include peripheral RPE changes and/or macular lesions.[1,2] Only one case demonstrated drusen in the fellow eye in this series.

Fluorescein angiography (FA) demonstrates irregular peripheral late hyperfluorescence without evidence of neovascularization with or without persistent hypofluorescence from masking hemorrhagic lesions,[2] as was seen in four eyes in our series, and maybe diagnostic in 83% cases.[7] Ultrawide field imaging has been shown to be useful to diagnose and objectively follow-up patients with PEHCR since the pathology lies anterior to the equator.[6]

It has been speculated that PEHCR may be a peripheral subtype of polypoidal choroidal vasculopathy owing to clinical similarities such as serous and hemorrhagic PED, SRH, and lipid exudation, a remitting–relapsing course with recurrent hemorrhagic episodes, but without polyps as demonstrated by indocyanine green angiography (ICGA).[2] However, dynamic ultrawide field ICGA has identified peripheral polyp-like choroidal telangiectasia and abnormal choroidal vascular networks in PEHCR.[3] Thus, a new classification of PEHCR has been proposed, which includes both eyes with and without polyps.[8] Unfortunately, ICGA could not be performed in the patients in this study.

Majority of PEHCR lesions remain stable or regress spontaneously, leaving RPE atrophy, hyperplasia, and fibrosis.[1] Close observation is recommended for peripheral lesions, as visual prognosis may be poor once central vision is impaired. Treatment has been indicated where the macula appears threatened or involved by fluid, exudation, or hemorrhage. A favorable response has been observed with intravitreal antivascular endothelial growth factor (VEGF) therapy, although this may also reflect the natural course of the disease.[8-10] Laser photocoagulation,[4,6] cryoretinopexy, or photodynamic therapy (PDT) may also be applied in cases with peripheral polyps. However, laser and cryotherapy might trigger massive SRH[10] and PDT may enlarge chorioretinal atrophy after treatment.

Less commonly, an aggressive course may ensue with massive submacular and/or VH, necessitating surgical intervention.[5,8,9] All five eyes in this series required PPV for dense VH. No additional treatment by anti-VEGF injection or laser or cryotherapy was performed intraoperatively or postoperatively. Three eyes achieved a final BCVA of 6/9 and 1 of 6/24 after a mean follow-up of 17.5 months (range 2–36 months). Minimal visual improvement was observed in one eye with a macular disciform scar. None of the eyes had any recurrences. Laser has been performed to prevent the spread of fresh subretinal bleeds to the macula,[5] but none of our cases showed fresh hemorrhage. Anti-VEGF therapy has been used along with PPV or post-PPV to prevent the spread or recurrence of SRH in eyes with VH from PEHCR.[9] However, the presumed beneficial effect of anti-VEGF injections needs to be weighed against the natural course of the disease. Use of silicon oil tamponade has been described in two eyes with peripheral hemorrhage found intraoperatively; however, the rationale for this is unclear.[9]

Conclusion

To conclude, there is a paucity of reports about PEHCR leading to VH in non-Caucasians. This series shows that it can occur in Indians as well. Rather than a low incidence, it may be underdiagnosed and a high index of suspicion might lead to increased recognition of these cases. Optimal visual and anatomic results can be obtained with PPV to clear the hemorrhage and additional treatment may not be required, even on long-term follow-up. However, permanent loss of vision can be encountered in cases with irreversible macular pathology.

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Conflicts of interest

There are no conflicts of interest.

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