Original Article

"Double-layer sign" on spectral domain optical coherence tomography in pachychoroid spectrum disease

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Purpose: The "double-layer sign (DLS)" describes the shallow and irregular elevation of the retinal pigment epithelium from the underlying intact Bruch's membrane visualized on the spectral domain optical coherence tomography. In this study, we evaluated the frequency, characteristics of the space within the double layer and other features in the pachychoroid spectrum to aid the clinical diagnosis of these variants. **Methods:** This retrospective study evaluated the features of the DLS on multimodal imaging in consecutive patients with a clinical diagnosis of one of the four variants of pachychoroid: pachychoroid pigment epitheliopathy (PPE), pachychoroid neovasculopathy (PCN), chronic central serous chorioretinopathy (CCSCR), and polypoidal choroidal vasculopathy (PCV). The features of the DLS were graded by two masked graders. **Results:** Overall, 102 eyes of 79 consecutive patients with pachychoroid spectrum were identified for grading. Sixteen eyes with PPE did not show any evidence of DLS. The DLS was identified in 15/16 (93.75%) eyes with PCN, 11/35 (31.43%) with CCSCR, and 32/35 (91.43%) with PCV ($P < 0.001$). The space within the DLS showed moderate hyperreflectivity in all eyes with PCV and PCN, while the space in the DLS in CCSCR showed uniform hyporeflectivity in 10/11 (%) eyes. **Conclusion:** The DLS sign was most frequent in polypoidal vasculopathy and PCN. A hyporeflective gap within the DLS favored the diagnosis of CCSCR.

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Pachychoroid is defined as increased choroidal thickness owing to enlarged Haller's layer that compress the vessels in the inner choroid and is best visualized on enhanced-depth imaging optical coherence tomography (EDI-OCT). A spectrum of diseases may manifest with pachychoroid and these include pachychoroid pigment epitheliopathy (PPE), central serous chorioretinopathy (CCSCR), pachychoroid neovasculopathy (PCN), and polypoidal choroidal vasculopathy (PCV).[1,2] The pachychoroid appearance on EDI-OCT is also associated with increased choroidal hyperpermeability and dilated choroidal vessels on indocyanine green angiography (ICGA).[3,4]

Long-standing abnormal increase in the choroidal thickness may result in chronic choroidal congestion and secondary backpressure changes causing structural damage to the retinal pigment epithelium (RPE) and Bruch's membrane.[5] These changes may manifest as irregularities in the RPE, pigment epithelial detachment, subretinal fluid, and intraretinal fluid. A characteristic feature that is observed on optical coherence tomography (OCT) in some eyes with pachychoroid is the "double-layer sign" (DLS), which describes the separation of the irregularly elevated RPE from the inner layer of the Bruch's membrane.[6]

Sato *et al*. initially described DLS in as a tomographic feature of the branching vascular networking polypoidal vasculopathy.[7] It has been established as the most distinctive sign that correlates well with early hypercyanescence of the branching network on ICGA. Sato *et al*. postulated that "the space within the DLS reflects fluid accumulation between the basement membrane of the RPE and the inner boundary of the Bruch's membrane/choriocapillaris complex." This fluid is presumed to arise from the leaking vascular channels of polyps. However, DLS was also first described in eyes with chronic central serous retinopathy by Yang *et al*. indicating that DLS *per se* may not be disease specific and that there may be other features in the DLS that may help differentiate the variants within the pachychoroid spectrum.[8] The aim of our study was to comprehensively evaluate all EDI OCT images of consecutive patients with a diagnosis of a pachychoroid variant to discern various tomographic features of DLS to better characterize the pachychoroid disease spectrum.

Methods

We retrospectively evaluated the clincial case records and multimodal imaging of consecutive patients with pachychoroidopathy that presented to the vitreoretinal services at our institute between June 2015 and December 2015.

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The study was conducted in accordance to the tenets of the Declaration of Helsinki and was approved by the Institutional Review Board. Written informed consent was obtained from each patient before any imaging procedures were done as part of routine clinical care.

Case definition of pachychoroid

Pachychoroid characteristics on the multimodal imaging were defined as the presence of any one of the following features: choroidal thickness greater than 270 μm on EDI-OCT and/or presence of pachyvessels.[1,2,4]

On the basis of multimodal imaging, the eyes were categorized into the four pachychoroid variants:

Pachychoroid pigment epitheliopathy

Presence of RPE irregularities with/without pigment epithelial detachment (PED) but no associated intraretinal fluid (IRF) or subretinal fluid (SRF).

Chronic central serous chorioretinopathy

Diagnosis of CCSCR was depending on the presence of features of CSCR lasting for more than 3 months associated with multimodal imaging characteristics such as serous macular detachment (SMD), IRF, PED, increased choroidal thickness, dilated choroidal vessels, and mid-phase choroidal hyperfluorescence in ICGA.[8]

Pachychoroid neovasculopathy

Type 1 neovascularization occurring secondary to CCSCR or long-standing PPE without any polyps on ICGA.

Polypoidal choroidal vasculopathy

PCV was diagnosed on ICGA as the presence of nodular hyperfluorescence appearing within the first 6 min with/without the presence of abnormal vascular network.[9]

Multimodal imaging

All patients underwent spectral domain optical coherence tomography (SD-OCT) (Spectralis HRA + OCT, Heidelberg Engineering, Heidelberg, Germany), enhanced depth imaging OCT (EDI-OCT), digital fluorescein angiography (DFA), and ICGA.

SD-OCT images were generated using the Dense Volume Scan [(20° × 20°; Scan configuration of 49 B-scan sections, 16 Frames OCT ART Mean, 120 um spacing, high speed (512 A scans/B scan)]. Each of the 49 B-scan sections were evaluated for DLS, and the longest DLS was chosen for the analysis. In addition, other features such as PED, IRF, and SRF were also recorded. Evaluation of SD-OCT for automated central macular thickness (CMT) was also recorded using the 25-line raster scan protocol. Single horizontal EDI-OCT line scan passing through the center of the fovea was used for analysis of choroidal thickness. The DFA and ICGA images were acquired using the 55º lens taken up to 10 min in DFA and up to 20 min for ICGA. At the first appearance of dye on ICGA, dynamic ICGA was performed using the video ICGA mode of spectralise for duration of 30 s. This dynamic ICGA was utilized to visualize the filling of the branching vascular network, including presence or absence of feeder vessel. Presence of this vascular network is characteristically seen in PCN and PCV cases.

Image analysis: Choroidal thickness

Choroidal thickness was measured manually with the help of built-in calipers in OCT software. Measurements were taken from the outer portion of hyperreflective line corresponding to the RPE to the inner portion of hyperreflective zone corresponding to the choroidoscleral junction by two masked observers (J. S, S. C.) [Fig. 1]. They were obtained in the subfoveal region (subfoveal choroidal thickness; SFCT). SFCT and CMT were compared between the 4 groups.

Image analysis: Double-layer sign

DLS was defined as two hyperreflective layers separated by a gap: The inner layer is the hyperreflective irregularly elevated RPE, and the outer layer is the inner layer of the Bruch's membrane [Fig. 2]. The presence or absence of DLS was scrutinized on SD-OCT for all the patients. Two masked graders (J. S., S. C) performed detailed analysis of DLS. Height and width of DLS was measured and compared between both the groups [CCSCR and PCV; Fig. 3]. The space between these two hyperreflective lines was classified as hyporeflective or hyperreflective when compared to the reflectivity of the vitreous. The presence of DLS was also correlated with SD-OCT features such as the presence of thumb-like pigment epithelial detachment and height of submacular detachment.

Statistical analysis was done using the SPSS software, version 16.0 (SPSS Inc., Chicago, IL, USA). All data were expressed in the form of mean ± standard deviation. One-way ANOVA test and Kruskal-Wallis test were used to compare the age, CFT, SFCT, DLS dimensions, and SMD height across the groups with post-hoc analysis. A *P* value of less than 0.05 was considered to be statistically significant.

Results

Overall, 102 eyes of 79 patients with a diagnosis of pachychoroid were included. These included 16 eyes with PPE, 16 eyes with PCN, 35 eyes with CSCR, and 35 eyes with PCV.

The average age of the cohort was 60.90 ± 10.61 years. Patients with CCSCR were significantly younger compared to other pachychoroid variants, namely PCN and PCV (CCSCR: 55.66 ± 10.89 years; PCV: 62.89 ± 8.64 years; PCN: 69.56 ± 7.77 years; $P < 0.001$). However, there was no significant difference between the mean age of PCN and PCV groups. Patients with PCN and PCV showed equal gender

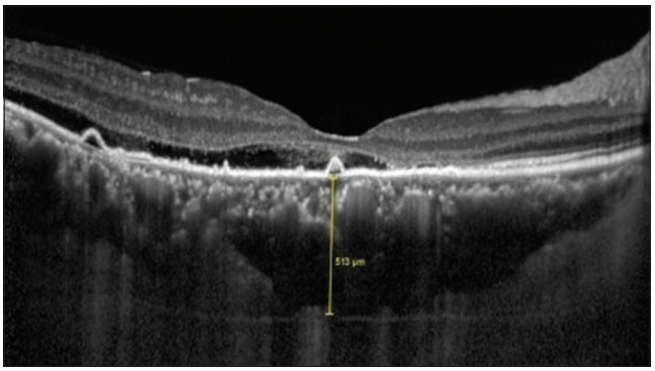

Figure 1: Optical coherence tomography enhanced depth imaging (horizontal scan) through the fovea demonstrating subfoveal choroidal thickness measurement from outer portion of hyperreflective line corresponding to the retinal pigment epithelium to the inner portion of hyperreflective zone corresponding to the choroidoscleral junction (yellow vertical line)

distribution, but there were significantly more male patients in the PPE and CCSCR group ($P = 0.005$).

The SFCT was significantly more in the CCSCR group as compared with the PPE, PCV, and the PCN groups (CCSCR: 467.23 ± 97.13 µm; PPE: 341.12 ± 67.02 µm; PCV: 297.11 ± 82.47 µm; PCN: 375.5 ± 94.47 µm; $P < 0.001$). The SFCT in PCN eyes was also significantly more than PCV eyes ($P = 0.006$) [Table 1].

There were no cases of DLS in the PPE group. Fifteen eyes with PCN had DLS (93.75%). In total, 32 of the 35 PCV eyes (91.43%) showed the presence of DLS, whereas DLS was seen in only 11 of the 35 eyes of CCSCR (31.43%). DLS was significantly associated with PCV and PCN as compared to CCSCR.

We also observed that all 32 eyes of PCV and all 15 eyes of PCN with DLS had a characteristic moderate hyperreflectivity in the space between the undulated RPE and Bruch's membrane, respectively [Fig. 4a]. In contrast, DLS seen in CCSCR showed uniform hyporeflectivity [Fig. 4b] in all of the 11 eyes, except one that showed moderate hyperreflectivity. The presence of hyperreflectivity in DLS was significantly associated with PCV and PCN ($P < 0.001$) [Table 1].

DLS height was significantly more in the PCV and PCN groups as compared with the CCSCR group (PCN: 73.19 ± 32.95 µm; PCV: 90.69 ± 34.93 µm; CCSCR: 43.18 ± 16.4 µm; $P = 0.032$). DLS width was also significantly more in the PCN and PCV groups as compared with the CCSCR group (PCN: 1716.44 ± 857.44 µm; PCV: 2382.06 ± 1143.45 µm; CCSCR: 962 ± 210.53 µm; $P < 0.001$). There was no significant difference in DLS dimensions between PCN and PCV eyes. There was no significant difference in the SMD height between all the groups (PCN: 96.38 ± 72.86 µm; PCV: 155.65 ± 73.76 µm; CCSCR: 139.39 ± 116.93 µm; $P = 0.07$). Similarly, there was no significant difference in CMT among all the groups (PPE: 248.81 ± 39.89 µm; PCN: 310.38 ± 76.97 µm; PCV: 304.89 ± 92.79 µm; CCSCR: 292.94 ± 148 µm; $P = 0.33$) [Table 1].

On assessing the DLS-associated features with PCV, 30 of the 32 eyes had thumb-like PED associated with DLS (93.8%) of which 2 eyes had PED on either side of the DLS [Fig. 5]. On ICGA, 30 of the 32 eyes had associated polyps with Branched Vascular Network (BVN) (93.8%),[Fig. 6a] whereas the remaining 2 eyes had only BVN. These findings were confirmed on multimodal image by simultaneous analysis of ICGA and SD-OCT [Fig. 6c]. In addition, 81.8% of the eyes in the CCSCR group had associated SMD, whereas 75% of eyes in the PCV group had associated SMD. At the site of DLS on ICGA, 5 eyes showed mid-phase hyperfluorescence (45.5%) and 4 eyes had dilated choroidal vessels (36.4%) in the CCSCR group [Fig. 7a and b], which was confirmed on SD-OCT [Fig. 7c], whereas 15 eyes had late geographic hyperfluorescence (46.9%) [Fig. 6b], and 9 of them had dilated choroidal vessels (28.1%) in the PCV group [Fig. 6a]. In PCN group, 9 of the 15 eyes had associated dilated choroidal vessels (60%) [Fig. 8c], while a fine abnormal network of vessels was visualized in 4 of these eyes (26.77%) [Fig. 8a and b]. Moreover, on DFA, leakage at the site of DLS was noted in 15.6% of the eyes (5 eyes) in the PCV group [Fig. 6d and e], 53.33% of eyes in PCN group (8 eyes), [Fig. 8d and e] and 45.5% of the eyes (5 eyes) in CCSCR group [Fig. 7d and e].

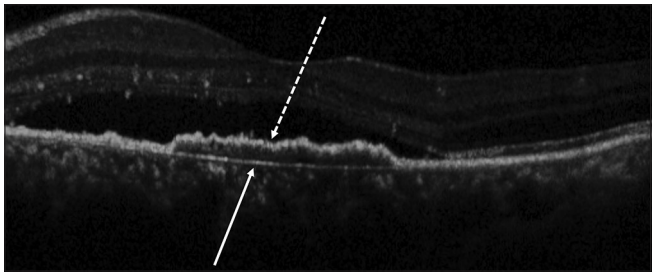

Figure 2: Spectral domain optical coherence tomography (horizontal scan) showing the "Double-Layer Sign." The inner layer is the hyperreflective irregularly elevated retinal pigment epithelium (dashed arrow), and the outer layer is the inner layer of the Bruch's membrane (solid arrow)

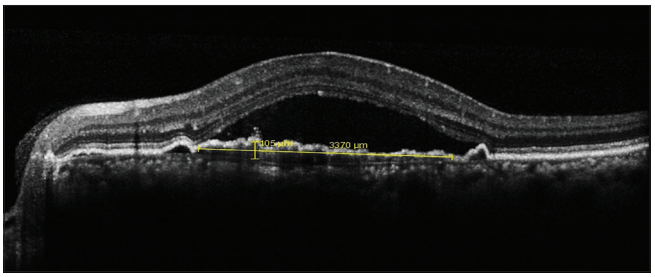

Figure 3: Spectral domain optical coherence tomography (horizontal scan) showing measurements of dimensions of double layer sign. Height of double-layer sign was defined as the greatest vertical distance measured from Bruch's membrane to the inner layer of retinal pigment epithelium (vertical yellow line, 105 µ), and the width was the widest horizontal dimension of the double-layer sign (horizontal yellow line, 3370 µ)

Table 1: Demographic and morphological features of pachychoroid spectrum of disease

	PPE	CCSCR	PCN	PCV	*P*
Age (years)	60.50±8.610	55.14±11.265	69.56±7.771	62.89±8.642	<0.001
Sex ratio (male:female)	12:4	29:6	8:7	18:17	
SFCT (µm)	341.12±67.01	467.23±97.13	375.50±94.47	297.11±82.47	<0.001
SMD height (µm)	0	139.39±116.93	96.38±72.86	155.65±73.76	0.066
CMT (µm)	248.81±39.89	292.94±148	310.38±76.97	304.89±92.79	0.194
Number of eyes DLS present	0/15	11/35	15/15	32/35	<0.001
DLS height (µm)	0	43.18±16.4	73.19±32.95	90.69±34.93	0.032
DLS width (µm)	0	962±210.53	1716.44±857.44	2382.06±1143.45	<0.001

PPE: Pachychoroid pigment epitheliopathy, CCSCR: Chronic central serous chorioretinopathy, PCN: Pachychoroid neovasculopathy, PCV: Polypoidal choroidal vasculopathy, SFCT: Subfoveal choroidal thickness, SMD: Serous macular detachment, CMT: Central macular thickness, DLS: Double layer sign

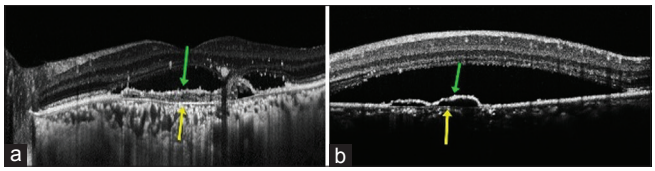

Figure 4: Spectral domain optical coherence tomography images representing the "Double Layer Sign" in both the disease entities. (a) In polypoidal choroidal vasculopathy eyes, the double-layer sign had a characteristic moderate hyperreflectivity in the space between the undulated retinal pigment epithelium (green arrow) and Bruch's membrane (yellow arrow). (b) In contrast, double-layer sign seen in chronic central serous chorioretinopathy showed uniform hyporeflectivity between the retinal pigment epithelium (green arrow) and Bruch's membrane (yellow arrow)

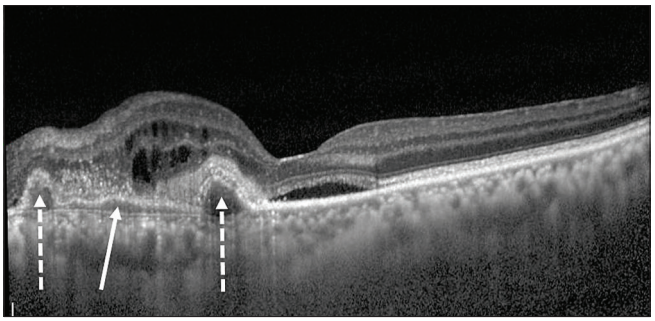

Figure 5: Spectral domain optical coherence tomography image of a hyper reflective double layer sign (bold white arrow) flanked on either side by a thumb-like pigment epithelium detachment (dashed white arrows)

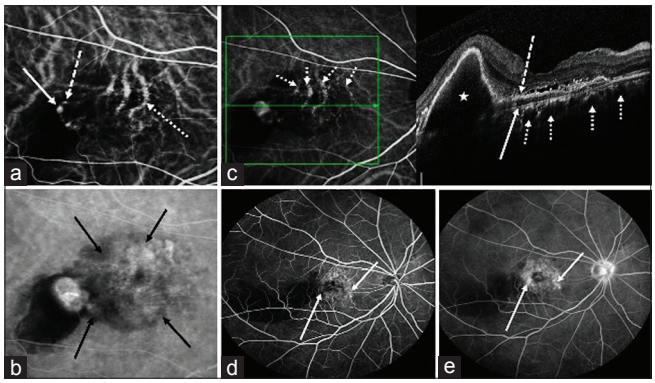

Figure 6: Multimodal imaging showing various features associated with the Double Layer Sign in polypoidal choroidal vasculopathy. (a) Indocyanine green angiography image demonstrating nodular hyperfluorescence suggestive of polyp (bold white arrow) along with abnormal vascular network with feeder vessel suggestive of Branched Vascular Network dashed arrow). Dilated choroidal vessels in close proximity to the vascular network (dotted white arrow) are also seen. (b) Late phase indocyanine green angiography image showing area of late geographic hyperfluorescence (bold black arrows) at the site of Branched Vascular Network. (c) Indocyanine green angiography image with simultaneous spectral domain optical coherence tomography scan through the polyp and Branched Vascular Network confirming pigment epithelial detachment at the site of polyp (white star) with a hyperreflective double-layer sign in continuity with it (dashed and bold long white arrows). Underlying the double-layer sign, multiple large hyporeflective lumina are seen in the choroid (dotted small white arrows) suggestive of pachyvessels which corresponding to the site of dilated vessels on indocyanine green angiography. (d and e) Early and late fluorescein angiography images showing leakage at the site of double-layer sign (bold white arrows)

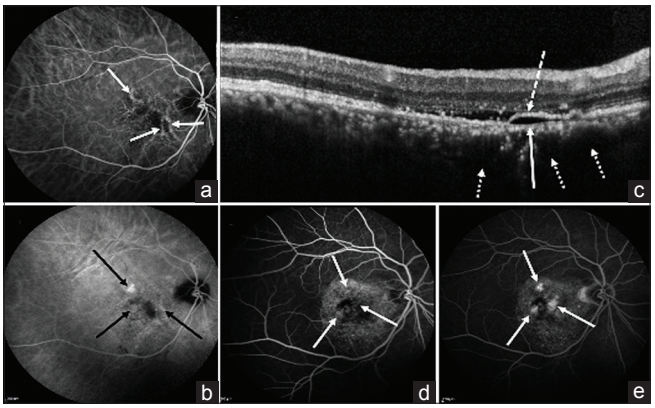

Figure 7: Multimodal image showing various features associated with the Double Layer Sign in chronic central serous chorioretinopathy. (a) Indocyanine green angiography image demonstrating the dilated choroidal vessels at the macula (bold white arrows). (b) Mid phase indocyanine green angiography image showing mid phase hyperfluorescence at the site of double-layer sign (bold black arrows). (c) Spectral domain optical coherence tomography scan at the level of Branched Vascular Network in indocyanine green angiography showing hyporeflective double-layer sign corresponding to Branched Vascular Network (dashed and bold long white arrows). Presence of pachyvessels noted beneath the double-layer sign depicted by hyporeflective large lumina in the choroid (dotted small white arrows). (d and e) Early and late fluorescein angiography images showing leakage at the site of double-layer sign (bold white arrows)

Discussion

In this study, we confirmed that the reflectivity of the longest DLS is an important diagnostic feature that can be used to differentiate the pachychoroid variants. We found that all 32 eyes of PCV with DLS had characteristic moderate hyperreflectivity in the space between the undulated RPE and Bruch's membrane. Similarly, all 15 eyes of PCN demonstrated hyperreflective DLS. In contrast, DLS seen in CCSCR showed uniform hyporeflectivity in all of the 11 eyes, except one that showed moderate hyperreflectivity. Liu *et al*. performed a comparative study to distinguish between PCV and AMD in 188 eyes on SD-OCT. SD-OCT parameters used to distinguish between the two included presence of two of the following three features: PED, DLS, and TLP.[10] From these parameters, they found SD-OCT to have a sensitivity of 89.4% and a specificity of 85.3% in detecting PCV. De Salvo *et al*. conducted an analogous study to differentiate between PCV and occult Choroidal neovascular membrane (CNVM) on SD-OCT.[11] They showed a sensitivity of 94.6%, specificity of 92.9%, PPV of 97.2%, and NPV of 86.7% to identify PCV. However, in this study, the diagnostic criteria used on SD-OCT were (any three of the four) as follows: sharp PED peak, notched PED, multiple PEDs, and hyporeflective lumen within hyperreflective lesion adherent to RPE, and DLS was not included. Yang *et al*. were the first to study DLS as an important OCT feature of both acute and CCSCR.[8] They found that DLS (seen in 51 eyes [75%]) was seen significantly ($P = 0.025$) more in the CCSCR group (29/33; 87%) as compared to the acute CSCR group (22/35; 63%). The DLS in CCSCR was more prevalent in their study population (87%)

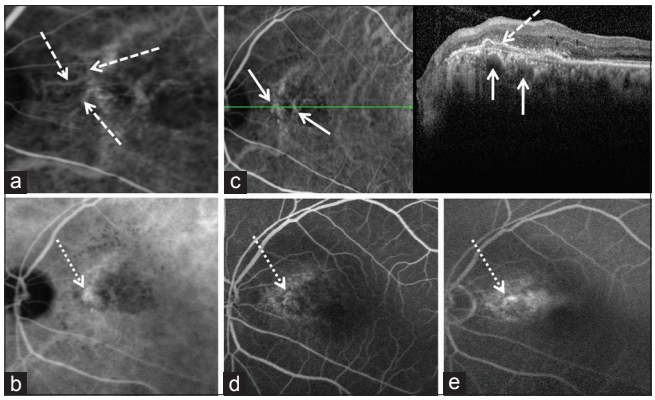

Figure 8: Multimodal image showing various features associated with the Double Layer Sign in Pachychoroid neovasculopathy. (a) Indocyanine green angiography image demonstrating abnormal vascular network suggestive of Branched Vascular Network (dashed arrow) that leaks minimally in late phases of indocyanine green angiography (dotted white arrow, b) (c) indocyanine green angiography image with simultaneous spectral domain optical coherence tomography scan illustrating a hyperreflective double-layer sign (dashed white arrow). Underlying the double-layer sign, multiple large hyporeflective lumina are seen in the choroid (bold white arrows) suggestive of pachyvessels that are corresponding to the site of dilated vessels on indocyanine green angiography (bold white arrows). (a and e) Early and late fluorescein angiography images showing leakage at the site of double-layer sign (dotted white arrows)

compared to the current study (11/35: 31.43%). Yang *et al*. studied the SD-OCT features of patients with CSCR.[8] In the eyes with DLS, the space between the undulated RPE line and the straight Bruch's membrane line appeared hyporeflective in 41 eyes (80%). In the remaining five eyes (10%) with DLS, the space was hyperreflective, whereas in the next five eyes (10%), the reflectivity in the space between the undulated RPE line and Bruch's membrane line could not be determined on the images. In this study, hyporeflective DLS was seen in around 90.9% of the eyes with CCSCR. This is slightly higher than what was noted by Yang *et al*., where it was found to be seen in 80% of the cases.

Ojima *et al*. evaluated the OCT features of PCV. They noted a hyperreflective material between the two layers of DLS in all the eyes.[12] They considered it as a sign of polypoidal lesions and BVNs.

The difference in the reflectivity between the RPE and Bruch's membrane may be because of the difference in the disease spectrum. Because PCV and PCN are a form of type 1 CNVM, the hyperreflectivity can be regarded as a neovascular complex, specifically the BVN, as described by various researchers.[13] Similarly, the PEDs seen in choroidal neovascular disorders such as wet AMD and PCV have moderate hyperreflectivity signifying fibrovascular PED.[10] Another possibility is that it could be the collected serosanguineous fluid leaking from the neovascular tissue, which gives the hyperreflectivity on SD-OCT. However, the hyporeflectivity between the RPE and Bruch's membrane may be suggestive of the presence of serous fluid. This is akin to the serous PEDs that we see in CSCR.

On quantitative analysis of DLS, we found that DLS height was significantly more in the PCV and PCN group as compared with the CCSCR group (PCN: 73.19 ± 32.95 μm; PCV: 90.69 ± 34.93 μm; CCSCR: 43.18 ± 16.4 μm; $P = 0.032$). Similarly, DLS width was significantly more in the PCV and PCN group as compared with the CCSCR group (PCN: 1716.44 ± 857.44 μm; PCV: 2382.06 ± 1143.45 μm; CCSCR: 962 ± 210.53 μm; $P < 0.001$). In addition, the dimensions of DLS in the PCV group were more as compared to the PCN group, although not significantly. This study is the first to perform such a detailed quantitative assessment of DLS. Larger dimensions of DLS in PCN and PCV is suggestive of extensive neovascular tissue with/without leaked exudation, which secondarily leads to significant separation of RPE from the Bruch's membrane leading to increased horizontal extent along with augmented RPE elevation leading to increased vertical extent. In contrast, the smaller extent of DLS in CCSCR is indicative of less significant RPE–Bruch's membrane complex impairment. Because CCSCR, PCN, and PCV form a part of the pachychoroid spectrum, with CCSCR preceding the development of PCN and PCV, the increase in the dimension of DLS in PCN and PCV as compared with CCSCR can be conjectured to be due to the progressive damage of RPE–Bruch's membrane complex as the spectrum advances. This can also explain the presence of overlying SMD associated with DLS in a large proportion of eyes in both the groups (CCSCR: 81.8%; PCV: 75%). Concomitant SMD is itself indicative of chronicity of the damage of the underlying RPE–Bruch's membrane leading to the development of DLS.

On ICGA, dilated choroidal vessels were seen in 36.4% of the eyes with CCSCR, 28.1% of the eyes with PCV, and 60% of eyes with PCN . These dilated choroidal vessels may play a role in the development of DLS, SMD, and even neovascularization formation. One conceivable reason may be the occurrence of long-standing compression and microtrauma to the overlying RPE–Bruch's membrane complex. This may be true more so for PCV because the vessels also show pulsatile flow that may augment the microtrauma.[14] Studies have also demonstrated the loss of inner choroidal volume and decreased choriocapillaries blood flow overlying pachyvessels. This may lead to focal ischemia and promote neovascularization.[15]

The presence of leakage on DFA was present in 45.5% of the eyes with CCSCR, 53% of eyes with PCN, and 15.6% of the eyes with PCV. In the CCSCR eyes, leakage at the site of DLS in almost half of the eyes reinforces the existence of underlying prolonged abnormality of RPE. It is possible that this will lead to micro-RPE rip, which can be seen as the leakage point on DFA.[8] However, leakage at the site of DLS was seen less frequently in the eyes with PCV. One possibility for this occurrence could be that in PCV, DLS potentially represents the BVN, which may not necessarily leak. At the terminal ends of these abnormal vessels, we see nodular hyperfluorescentlesions that represent the polyps, which may be the actual point of leakage. To underline this histopathological association of BVN and terminal polyps, we performed a detailed analysis on multimodal imaging. On SD-OCT, we found the presence of thumb-like PED thumb-like pigment epithelial detachment (TLP), illustrating polyp, in 30 of the 32 eyes with DLS in the PCV group, of which 2 eyes had TLP on either side of DLS. On ICGA, polyps were seen in 30 of the 32 eyes with DLS, while the remaining 2 eyes had only DLS.

The strengths of our study include its adequate sample size and detailed evaluation of "DLS" in an evolving spectrum of disease, the pachychoroidopathy. Moreover, this

study incorporated all pachychoroid variants to perform a qualitative and quantitative assessment of DLS on multimodal imaging. The changes in the dimensions of DLS from one stage of pachychoroid spectrum (i.e. CCSCR) to the terminal stages (i.e., PCN and finally PCV) represent progressive worsening of RPE–Bruch's membrane complex impairment. This provides supportive imaging evidence to the evolution and progression of the spectrum. We propose long-term follow-up of these patients on multimodal imaging to evaluate the changes across the entire gamut of pachychoroid diseases.

Our study has few limitations. First, it is primarily a retrospective review study of multimodal imaging. In addition, we could not correlate between the occurrence of DLS and the underlying pachyvessels. This is because, in our study population, the EDI-OCT horizontal line scan was performed only in the foveal region. However, in many of our cases, DLS was located outside the EDI-OCT scanned area, which made it unfeasible to find an association between them.

Conclusion

Our study demonstrates that in pachychoroid disorders, presence of DLS on SD-OCT is significant feature associated with PCN and PCV as against CCSCR. Quantitatively, DLS seen in PCV and PCN is more broad-based with significantly greater height as compared to CCSCR. In addition, the presence of hyporeflectivity between the undulating RPE and underlying Bruch's membrane is a crucial marker of CCSCR, whereas hyperreflectivity distinctly correlated with PCN and PCV, indicative of underlying neovascular tissue complex. Recognition of DLS on SD-OCT is an important indicator of pachychoroid disease spectrum and is a guide to perform the additional multimodal imaging in the form of DFA and ICGA, including dynamic ICGA.

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Conflicts of interest

There are no conflicts of interest.

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