Coexistence of retinal angiomatous proliferation and pachychoroid phenotype

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Retinal angiomatous proliferation (RAP) is usually seen in the eye with thin choroid and drusen.\cite{1,2} Pachychoroid is the morphological description of the thick choroid.\cite{3} We herein report two cases of RAP noted in eyes with pachychoroid.

Case 1

Fundus photograph of the right eye of a 48-years-old gentleman with 6/18, N8 vision showed RPE alterations [Fig. 1a and b]. Spectralis SDOCT line scan showed intraretinal edema, hyperreflective spot, subfoveal choroidal thickness (CT) of 436 µ and dilated large choroidal vessels. Line scan through the area of thickening showed serous RPE detachment and edema [Fig. 1c and d]. Early phase fluorescein angiogram (FA) showed hyperfluorescence at fovea [Fig. 2a]. Indocyanine angiogram (ICGA) showed hot spot [Fig. 2b]. Late phase FA [Fig. 2c] showed a leak whereas ICGA [Fig. 2d] showed hypercyanescence at fovea amidst diffuse hypercyanescence due to choroidal hyperpermeability. A diagnosis of pachychoroid phenotype with RAP was made.

Case 2

Fundus photography of left eye of a 62 years old gentleman with 6/18, N10 vision revealed RPE alteration at fovea [Fig. 3a and b]. SDOCT scan through fovea showed hyperreflective spots, subretinal fluid, and CT of 336 µ. Line scan above fovea showed prominent hyperreflective spot with dilated large choroidal vessel layer [Fig. 3c and d]. Early FA image showed hyperfluorescence corresponding to hyperreflective spot and leak in late phase [Fig. 4a and b] suggestive of RAP in an eye with pachychoroid phenotype.

Discussion

Thickening of RPE and drusen in eyes with RAP hamper the diffusion of VEGF from RPE to choroid, which disturbs choroidal autoregulation leading to attenuation of choriocapillaris.\cite{2,4} Thin choroid reduces the transport of metabolites back to retina, accentuating the retinal ischemia, and leading to RAP.\cite{21}

Figure 1: (a) Color fundus photograph of the right eye shows retinal pigment epithelium (RPE) alterations (black arrow). (b) Confocal scanning laser ophthalmoscopy based multicolor image shows greenish hue (yellow arrow) owing to thickening. RPE atrophy is highlighted as reddish hue (black arrow). (c) Spectral domain optical coherence tomography (SDOCT) line scan through the fovea shows intraretinal thickening (yellow arrow) and hyperreflective spot (red arrow). Subfoveal choroidal thickness is 436 µ. Dilated Haller’s layer vessels (yellow stars) are seen. (d) SDOCT line scan slightly above fovea shows serous RPE detachment (red arrow) and intraretinal edema (yellow arrow)

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Attenuation of choriocapillaris is sine qua non of pachychoroid phenotype.[5] The pachychoroid neovasculopathy (PNV) in these eyes is a result of ischemic environment in sub-RPE space owing to attenuation of choriocapillaris.[3] Attenuation of choriocapillaris is common pathology between RAP and pachychoroid. It may be possible for eyes with pachychoroid to develop type 3 neovascularization or RAP in the manner in which they develop PNV. This plausible explanation for RAP in pachychoroid eyes needs to be substantiated in a larger case series.

Figure 2: (a) Early phase fluorescein angiography (FA) image of right eye shows hyperfluorescence at fovea. (b): Early phase indocyanine angiography (ICGA) shows hot spot at fovea. (c): Late phase FA image shows hyperfluorescence increasing in area and intensity suggestive of leak from retinal angiomatous proliferation (RAP). (d): Late phase ICGA image shows diffuse hypercyanescence at macula due to choroidal hyperpermeability in pachychoroid eyes. RAP is seen as focal increased hypercyanescence at fovea

Figure 3: (a) Color fundus photograph of left eye shows RPE alterations (black arrow). (b) Multicolor image shows greenish hue nasal to fovea owing to thickening (yellow arrow). (c): SDOCT line scan through fovea shows hyperreflective spot (yellow arrow) at the level of outer plexiform layer. There is small subretinal fluid (red arrow). Subfoveal choroidal thickness is 336 µ. (d) SDOCT line scan superior to fovea shows well defined hyperreflective spot (yellow arrow) and shallow RPE detachment along with dilated Haller’s layer vessels (yellow star)

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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