# Ocular coherence tomography angiography features of congenital hypertrophy of retinal pigment epithelium

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Congenital hypertrophy of retinal pigment epithelium (CHRPE) is a benign, pigmented, flat lesion arising from the retinal pigment epithelium (RPE). In this study, we describe optical coherence tomography angiography (OCTA) features of two eyes with solitary CHRPE. We found that the retinal vasculature over CHRPE was normal in both cases. We observed that in solitary CHRPE, segmentation artifacts can interfere in the interpretation of retinal vasculature due to thinning of the outer retina. Visualization of the underlying choroidal vasculature was obscured to some extent by masking effect of the hyperpigmented RPE. The choroidal vasculature was better appreciated on en face OCTA. On OCTA, the retinal and choroidal vasculature associated with CHRPE was found to be normal in our study.

**Key words:** Congenital hypertrophy of retinal pigment epithelium, optical coherence tomography angiography, solitary, pigmentation, masking, unmasking

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Congenital hypertrophy of retinal pigment epithelium (CHRPE) is a benign, pigmented, flat lesion arising from the retinal pigment epithelium (RPE).[1] Various vascular changes over solitary CHRPE have been described using fundus fluorescein angiography (FFA).[2,3] In FFA, the visualization of the choroidal vasculature is obscured by hypertrophied RPE. Optical coherence tomography angiography (OCTA) features of various retinal and choroidal tumors have been described.[4,5] In this study, we report OCTA features of two cases of solitary CHRPE.

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## Case Report

### Case 1

A 40-year-old male presented to us for routine eye examination. On ocular examination, he had best-corrected visual acuity (BCVA) of 6/6, N6 in both eyes. Left eye anterior segment was normal. Fundus examination of the left eye revealed a well-demarcated, flat, brownish-black pigmented lesion in inferonasal quadrant measuring three disc diameters in size [Fig. 1a]. The lesion was hypo-autofluorescent on fundus autofluorescence (FAF) [Fig. 1b]. OCT over the lesion revealed thinning of the outer retinal layers [Fig. 1c]. OCTA on automatic segmentation at the level of superficial capillary slab showed a signal void area corresponding to the lesion, giving an impression of hypoperfusion [Fig. 1d]. But the vasculature was found to be normal on manual segmentation [Fig. 1e]. Deep capillary slab revealed normal vasculature after manual segmentation [Fig. 2a and b]. Choroidal slab at the level of large choroidal vessels showed a patch of heterogeneous signals corresponding to the extent of the CHRPE. The flow signals of the lesion were high where the lesion was hypopigmented superiorly. The large choroidal vessels which were seen like a ribbon till the margin of the lesion could not be traced into the lesion on OCTA. Projection artifact due to overlying vessels could be seen in deeper choroidal slabs [Fig. 2c]. The underlying choroidal vessels could be better appreciated on en face OCTA [Fig. 2d].

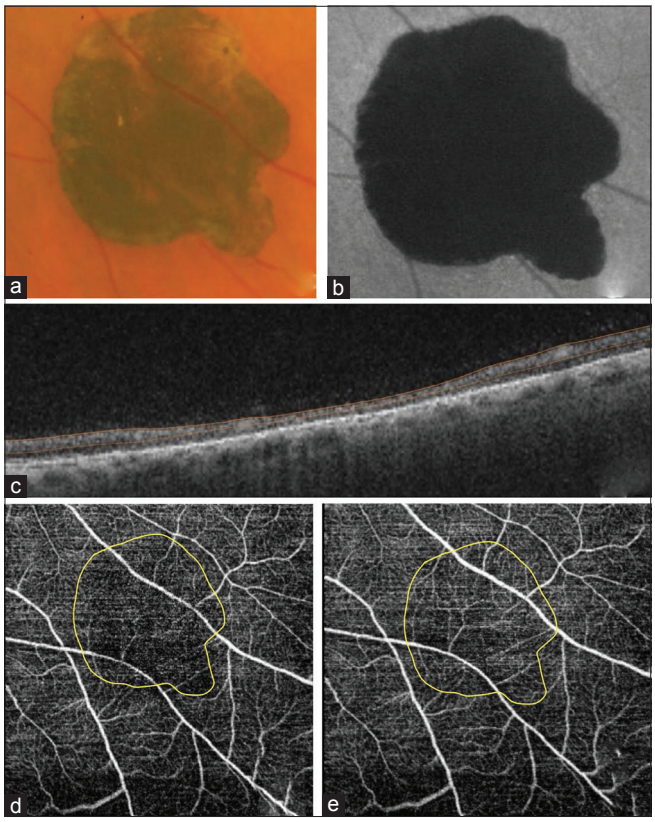


**Figure 1:** (a) Color photograph showing a well-demarcated, flat, blackish-brown pigmented lesion. (b) Uniformly hypo-autofluorescent on fundus autofluorescence with well-demarcated margin. (c) Optical coherence tomography showing normal inner retina with thinning of the outer retina with hyperreflectivity of RPE when compared with the adjacent RPE. (d) OCTA 6 × 6 mm superficial slab after automated segmentation showing a signal void area corresponding to the lesion giving an impression hypoperfusion (yellow line). (e) OCTA superficial slab after manual segmentation showing normal vasculature (yellow line)

### Case 2

A 35-year-old male presented to us for diminution of vision in both eyes. On examination, BCVA was 6/6, N6 in both eyes. Right eye examination was unremarkable. Left eye fundus examination revealed a pigmented well-demarcated brown lesion in the inferotemporal quadrant with lacunae and surrounding halo [Fig. 3a]. Lesion was hypoautofluorescent on FAF except at the lacunae where it was hyperautofluorescent [Fig. 3b]. OCTA of the lesion showed blink and motion artifacts. On manual segmentation, superficial vasculature was normal. Interestingly in the superficial slab, branching of the major blood vessel was noted which was not seen clinically [Fig. 3c]. Choroidal slab at the level of large choroidal vessels showed signal void corresponding to the lesion. A high reflectivity signal was noted at the lacunae [Fig. 3d].

## Discussion

CHRPE is a benign pigmented fundus lesion which has an indolent course. Few vascular changes associated with CHRPE are capillary nonperfusion, microanuerysms, and chorioretinal anastomosis.[2,3] All these vascular changes were described using FFA.

In both the cases in our study, the retinal vasculature over the lesion was normal. Due to thinning of the outer retina, automatic segmentation resulted in segmentation artifact. Similar thinning of outer retina was reported by Fung *et al.*, who proposed that thinning might be due to loss of photoreceptors as a result of poor phagocytic activity of RPE.[6] Hence, while analyzing the retinal vasculature over CHRPE, one should analyze the slabs after manual segmentation, as signal void areas on automated segmentation can be mistaken for hypoperfusion.

In our cases on OCTA, the hyperpigmented RPE caused masking artifacts resulting in signal void and areas of relatively hypopigmented areas (lacunae) over the CHRPE resulted in high flow signal due to unmasking. Parsons *et al.* and Lloyd WC *et al.* in their microscopic observation of CHRPE noted that RPE cells are approximately twice as tall as normal cells, with intense, uniformly distributed round pigment granules. Lacunae are associated with RPE atrophy and loss.[7,8] This report correlates OCTA findings with known histological facts about CHRPE.

En face OCTA in case 1, when compared with OCTA, had better visualization of the large choroidal vessels underneath CHRPE. Parsons *et al.* reviewed histopathological features of both solitary and grouped CHRPE.[7] Of the 10 studies reviewed, only 1 study by Shields and Tso has commented on the choroidal vasculature. Shields and Tso reported that choroid was normal on gross examination of an enucleated eye with grouped CHRPE.[9] Fung *et al.* in their study on enhanced depth imaging of CHRPE reported that the underlying choroid was identical in thickness and vascular appearance when compared with the tissue immediately outside the CHRPE margin. Based on the normal choroidal findings underlying CHRPE, it has been suspected that CHRPE is unrelated to the choroid.[6]

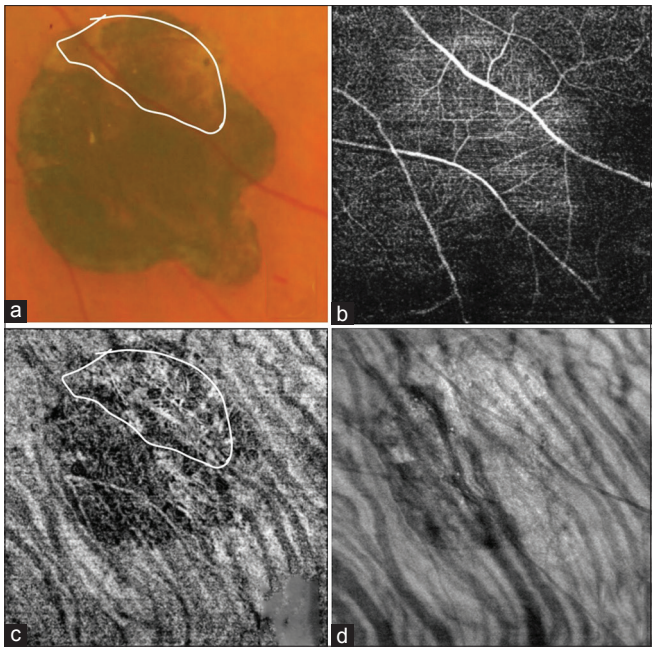


**Figure 2:** (a) Color photograph of case 1 showing a well-demarcated, flat, brown pigmented lesion with relative hypopigmentation in the superior part of the lesion (white line). (b) OCTA deep capillary slab after manual segmentation showing normal vasculature. (c) OCTA at the level of large choroidal vessels shows a signal void area with heterogeneous signals. The superior part of the patch which corresponds to the relatively hypopigmented area of the lesion has high reflective signals due to unmasking. (d) En face OCTA showing normal ribbon-like large choroidal vessels continuing into the lesion

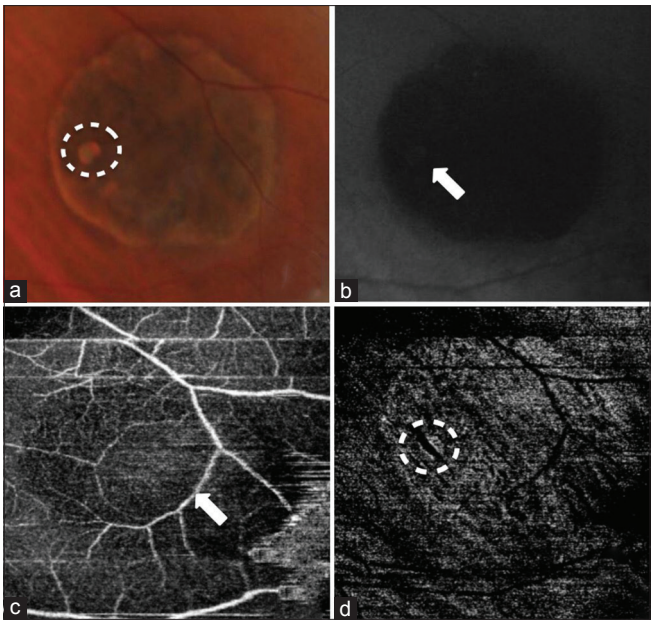


**Figure 3:** (a) Photograph of case 2 showing hyperpigmented lesion, halo, lacunae (dotted circle). (b) FAF showing hypoautofluorescent patch with hyperautofluorescence at lacunae. (c) OCTA superficial slab showing normal vascularity. Motion artefacts were noted. Vessel traversing lesion showing branching inferiorly (white arrow). (d) OCTA at large choroidal vessels showing high flow signals at the lacunae (dotted line)

The granules in the RPE block the visualization of underlying choroidal vessels on FFA and indocyanine green angiography (ICGA).[10] Unlike FFA and ICGA, the choroidal vasculature could be better appreciated on OCTA except for minimal masking owing to thickened RPE.

In case 2, the superficial blood vessel overlying CHRPE which was seen on OCTA was not seen on color photograph [Fig. 3c]. This might be because of the obscuration of the vessel by overlying pigment clumps as described by Touriño *et al.* in their study on FFA features of CHRPE.[2] Such pigments should also cause masking of signal on OCTA, but the reason for absence of masking in our case cannot be explained.

The limitation of OCTA is that it is difficult to image peripheral lesions and hypertrophic RPE obscures the visualization of the choroidal vessels partially. The majority of CHRPE lesions are located anterior to the equator and hence difficult to image by OCTA.[1] With conventional fundus camera, Touriño *et al.* reported that only 27.8% of the lesions could be studied using FFA.[2] With the advent of wide-angle OCTA, this shortcoming can be overcome. The hypertrophic RPE prevents deeper penetration of the signal into deeper choroidal structures to some extent.

To conclude, OCTA is a useful noninvasive tool in assessing vascularity of CHRPE. Using OCTA, we could image the choroidal vasculature which was not possible with FFA and ICGA. The OCTA findings correlated well with the histological features of CHRPE. Superficial large vascular branches over CHRPE which could not be identified on clinical examination could be detected on OCTA.

## Conclusion

OCTA is an excellent noninvasive tool in evaluating the retinal and choroidal vasculature of CHRPE.

### 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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Nil.

### Conflicts of interest

There are no conflicts of interest.

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