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## CASE REPORT

## Retinal and choroidal circulation determined by optical coherence tomography angiography in patient with amyloidosis

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**SUMMARY**

A 43-year-old woman who was diagnosed with the cryopyrine-associated periodic syndrome (CAPS) with severe renal failure and heart failure due to amyloid accumulation was examined by swept source optical coherence tomography (OCT) (SS-OCT; DRI-OCT, Topcon, Tokyo, Japan) and optical coherence tomography angiography (OCTA) (RTVue XR Avanti, Optovue, Fremont, CA). Her best-corrected visual acuity was 20/40 OD and 20/25 OS. A hyporeflective band of about 100 µm thickness was seen just inferior to the retinal pigment epithelium in the cross-sectional SS-OCT images, but the deeper choroidal structures were clearly visible. In the OCTA images, the density of the retinal capillaries in the superficial and deep capillary plexus slabs were reduced, and no signals of the choroidal capillary slab was detected after removing the projection artefacts. The accumulation of amyloid can cause a reduction of both the retinal and choroidal capillary circulations although the circulation in the larger vessels are preserved.

**BACKGROUND**

The cryopyrine-associated periodic syndrome (CAPS) is a hereditary autoinflammatory syndrome with fever, exanthema, arthritis, aseptic meningitis and optic neuritis. The chronic inflammation causes amyloid deposits in multiple organs including the eye.<sup>1-5</sup> A case report showed amyloidosis is associated with chorioretinopathy and a thicker hyporeflective choriocapillaris band at the area of choriocapillaris in optical coherence tomography (OCT). Some histopathological study reported an occlusion of the choriocapillaris due to amyloid deposits. The current study is the first report about amyloidosis using OCT angiography (optical coherence tomography angiography [OCTA]), which proved the capillary abnormalities in the retina and choroid in a patient with amyloidosis.

**CASE PRESENTATION**

The patient was a 43-year-old woman with recurrent uveitis, undergoing haemodialysis for end-stage renal failure and chronic heart failure. She also had a history of arthritis, cold urticaria and recurrent fever since childhood. Her mother had diabetes mellitus and her father had polyarteritis nodosa. Genetic examination revealed a E304K hetero mutation of the NLRP3 gene, and she was diagnosed with CAPS at age of 35 years.

Her uveitis went into remission after she was treated with anakinra (Kineret). She had cataract surgery on the right eye at age of 38 years and in the left eye at 43 years.

Although her visual acuity was 20/40 oculus dexter (OD) and 20/25 oculus sinister (OS), corneal amyloid deposits were observed in both eyes by slit-lamp biomicroscopy. Indirect ophthalmoscopy revealed vitreous opacities, pale optic discs and atrophy in the peripheral retina in both eyes (figure 1).

Cross sectional SS-OCT images showed a shallow foveal depression and a thinning of the inner retinal layer especially temporal to the fovea. An ellipsoid zone could not be detected, and irregularities of the retinal pigment epithelium (RPE) at the temporal area was observed in the both eyes. A hyporeflective band was detected just below the RPE in both eyes even though the structures of the deep choroidal layer below the hyporeflective band were clearly visible in the SS-OCT images (figure 2). The central retinal thickness was 172 µm OD and 208 µm OS. The thickness of the subfoveal choroidal and subfoveal hyporeflective bands were 264 µm and 95 µm OD and 250 µm and 97 µm OS, respectively.

The superficial and deep capillary plexuses of the retinal vessels were visible in the OCTA images although the density of the capillaries was reduced in both images (figure 3). The retinal vessels were also projected onto the choroid capillary slab as projection artefacts. No signal information at the segmentation ranges of the full-thickness of the hyporeflective band including the choriocapillaris was observed after removing the projection artefacts (figure 4).

**OUTCOME AND FOLLOW-UP**

No change within a year.

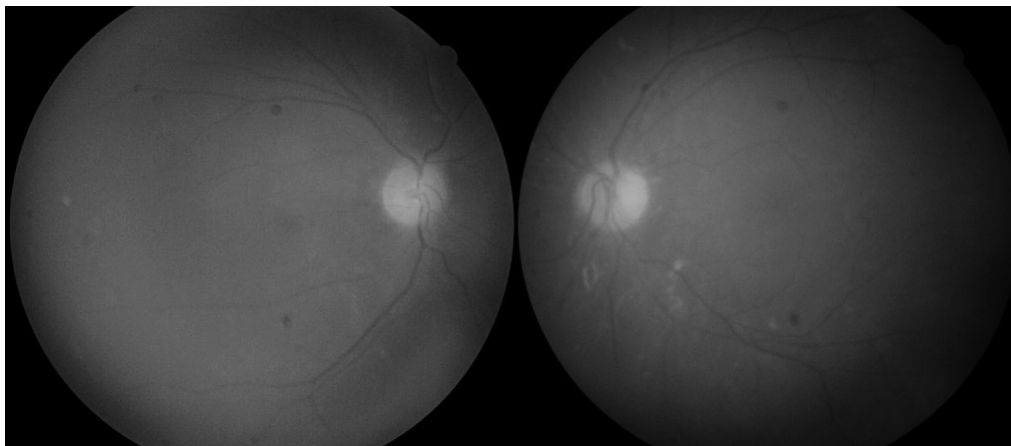
**DISCUSSION**

The OCTA images of the eyes of our patient with severe systemic amyloidosis due to CAPS showed not only a reduction of the retinal capillary density at the inner retinal layer but also the loss of choriocapillaris blood flow at the hyporeflective choriocapillaris band in the cross sectional OCT. Roybal *et al*<sup>6</sup> conducted a retrospective analysis of four patients with amyloidosis-induced chorioretinopathy, and they reported a thicker hyporeflective choriocapillaris band in the OCT images. This band



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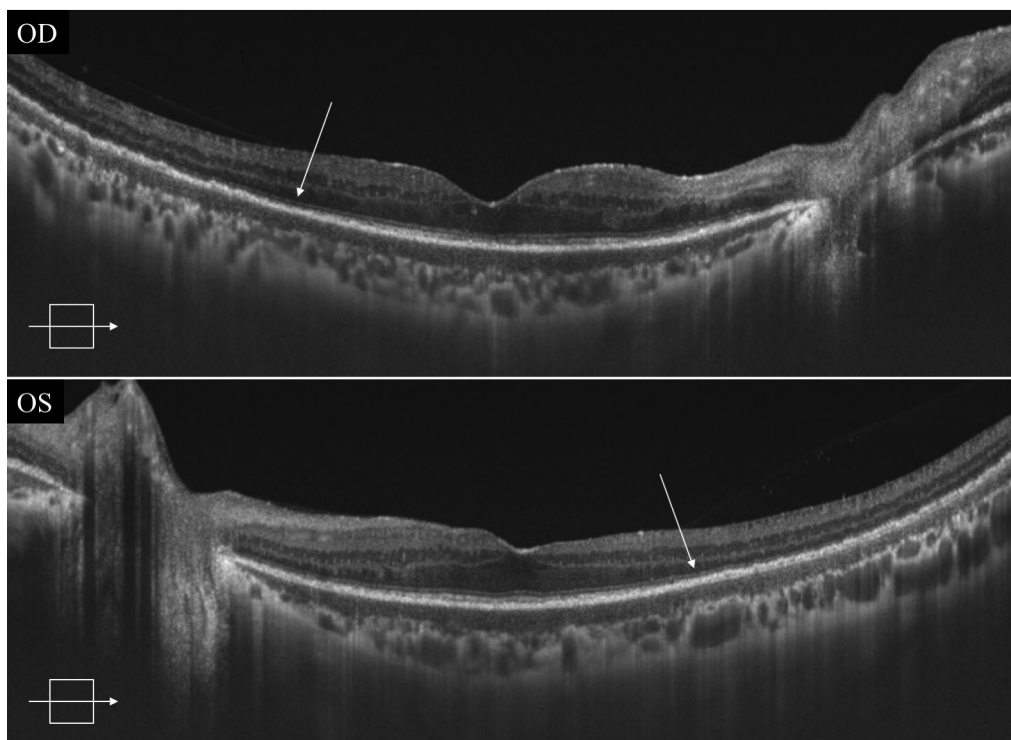


**Figure 1** Fundus photographs showing the blurred images due to the vitreous opacities and the pale optic discs in both eyes.

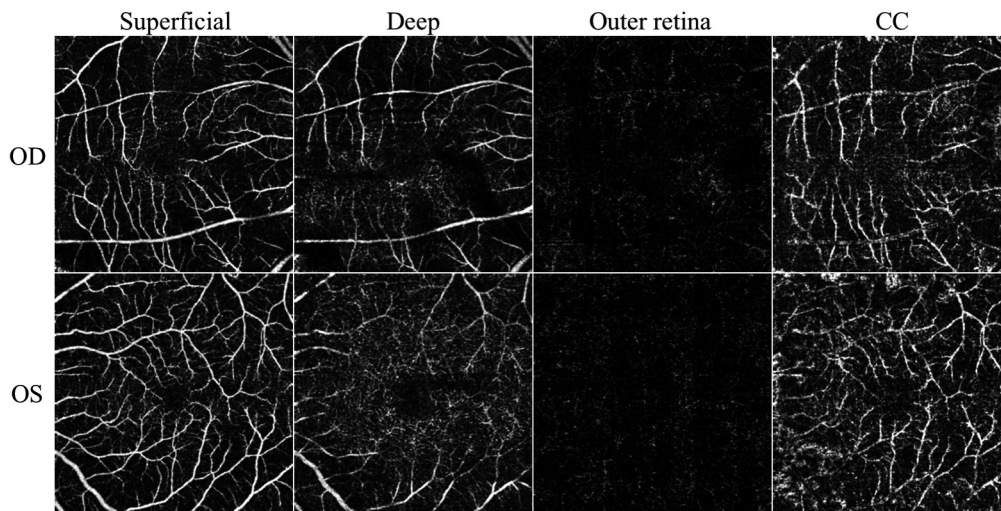
is similar to the hyporeflective band in the SS-OCT images in our case. They stated that the appearance of this band was caused by the accumulation of amyloid and the choriocapillaris appeared to be occluded. They also stated that the outer nuclear layer was thinner which could have been due to a reduction of the choroidal circulation. Some of their cases also had a thinning of the inner retina, and they might have included these cases with the retinal capillary abnormalities. However, this can not be proven because they did not have OCTA data.

Amyloid deposits in the choroid, especially in the choriocapillaris, were reported in a histopathological study by Ts'o and Bettman.<sup>7</sup> They documented the occlusion of the choriocapillaris by amyloid deposits in a patient with primary systemic

amyloidosis. They also stated that there were no abnormal retinal capillaries and no amyloid deposits in the entire inner retinal layer. On the other hand, Yamashita *et al*<sup>8</sup> reported the presence of amyloid deposits in the inner retinal layer, and these were connected to the vitreous. Other studies reported amyloid deposits extending from the retina to the vitreous body, and these investigators raised some concern on the effects of the amyloid deposits on the inner retinal layer.<sup>9 10</sup> Our case had not only severe amyloidosis, but was also undergoing haemodialysis which could have affected the retinal and choroidal capillaries. Even so, amyloid deposition may occur in both retinal and choroidal capillaries if it is in quite severe cases.



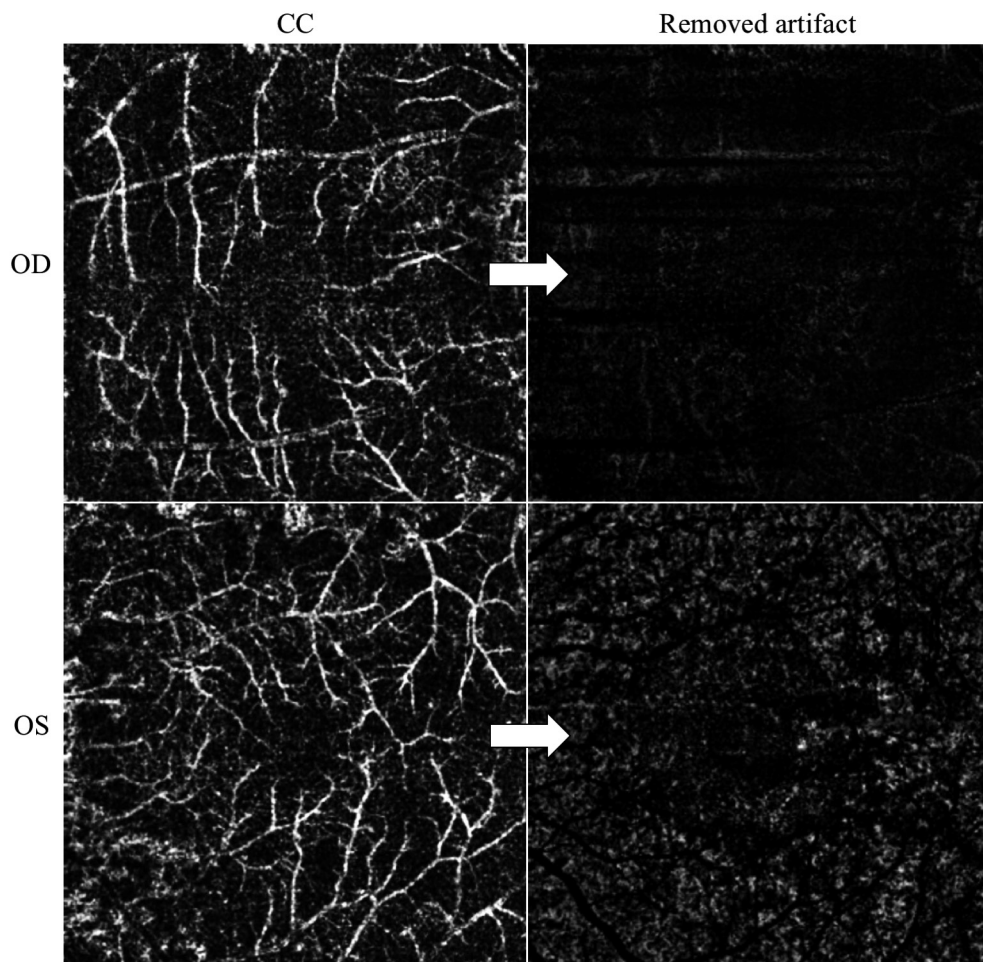
**Figure 2** Swept source optical coherence tomographic image showing an absence of the ellipsoid zone on the temporal side of the retina (arrows), and the presence of a hyporeflective band just below the retinal pigment epithelium in both eyes. The thickness of the hyporeflective band at the subfovea is 95  $\mu\text{m}$  OD and 97  $\mu\text{m}$  OS.



**Figure 3** Optical coherence tomography angiography (6×6 mm) shows the decrease in the density of the retinal capillaries in both the superficial and deep capillary plexuses, and the obvious retinal vessels projected at the choroid capillary slab. CC, choroid capillary.

To our best knowledge, this is the first report of a signal loss of not only the retinal capillaries but also the choriocapillaris in a patient with severe systemic amyloidosis as determined by OCTA. Although amyloid deposits have been confirmed pathologically in the retina and choroid, our findings were obtained

noninvasively and in situ by OCTA. Our findings indicate that it is important to evaluate the circulation of choriocapillaris as its occlusion can cause the atrophy of outer retina and dysfunction of RPE. Because the deeper choroidal structures were clearly visible in the cross-sectional SS-OCT images, the full-thickness



**Figure 4** Optical coherence tomography angiography (6×6 mm) shows an absence of blood flow signals in the choroid capillary slab after removing the projection artefacts. CC, choroid capillary.



hyporeflective band might be caused by the occlusion of the choriocapillaris. The findings in this patient demonstrate the usefulness of OCTA in monitoring the chorioretinal condition in patients with amyloidosis-induced chorioretinopathy.

### Learning points

- ▶ Amyloid deposits with multiple organs including the eye, and is sometimes associated with chorioretinopathy.
- ▶ Optical coherence tomography shows the thick hyporeflective choriocapillaris band.
- ▶ Optical coherence tomography angiography shows not only a reduction of the retinal capillary but also the loss of choriocapillaris blood flow.

**Contributors** Designing and conducting of study: MT, IM. Data collection: MT, IM, EU. Analysis and interpretation of data: MT, IM, EU. Manuscript preparation and review: MT, IM, EU, TI. Approval of the manuscript: MT, IM, EU, TI.

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### REFERENCES

- 1 Cuisset L, Jeru I, Dumont B, *et al.* Mutations in the autoinflammatory cryopyrin-associated periodic syndrome gene: epidemiological study and lessons from eight years of genetic analysis in France. *Ann Rheum Dis* 2011;70:495–9.
- 2 Pece A, Yannuzzi L, Sannace C, *et al.* Chorioretinal involvement in primary systemic nonfamilial amyloidosis. *Am J Ophthalmol* 2000;130:250–3.
- 3 Scuderi G, Abdolrahimzadeh S, Troccola A, *et al.* Surgical treatment of a rare case of bilateral ptosis due to localised ocular amyloidosis. *Saudi J Ophthalmol* 2016;30:201–3.
- 4 Ando T, Oshitari T, Saito M, *et al.* A case of conjunctival amyloidosis with repeated subconjunctival haemorrhage. *Case Rep Ophthalmol Med* 2017;2017:1–5.
- 5 Venkatesh P, Selvan H, Singh SB, *et al.* Vitreous amyloidosis: Ocular, systemic, and genetic insights. *Ophthalmology* 2017;124:1014–22.
- 6 Roybal CN, Sanfilippo CJ, Nazari H, *et al.* Multimodal imaging of the retina and choroid in systemic amyloidosis. *Retin Cases Brief Rep* 2015;9:339–46.
- 7 Ts'o MO, Bettman JW. Occlusion of choriocapillaris in primary nonfamilial amyloidosis. *Arch Ophthalmol* 1971;86:281–6.
- 8 Yamashita H, Ishi Y, Ando K, *et al.* Systemic amyloidosis: Histochemical observation of the eye. *J Clin Electron Microscopy* 1990;23:183–93.
- 9 Inomata H, Okayama M, Oshima K. Familial primary amyloidosis: Light and electron microscopic histopathology of the eye. *Jpn J Ophthalmol* 1976;20:51–62.
- 10 Hattori T, Shimada H, Yuzawa M, *et al.* Needle-shaped deposits on retinal surface in a case of ocular amyloidosis. *Eur J Ophthalmol* 2008;18:473–5.

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