Optical coherence tomography angiography features of paracentral acute middle maculopathy in a young female in sub-Saharan Africa: A case report

Olufemi Oladimeji Oderinlo, Adekunle Olubola Hassan, Ogugua Ndubuisi Okonkwo

ABSTRACT

To report the Optical Coherence Tomography Angiography features of a 33-year-old female presenting with visual loss secondary to a paracentral acute middle maculopathy. A case report of clinical examination findings, Fundus fluorescein angiography and Angiovue optical coherence tomography angiography (OCTA) findings of a paracentral acute middle maculopathy. The Angiovue optical coherence tomography angiography (OCTA) software of the RTVue XR Avanti (Optovue, Inc., Fremont, CA) was used for imaging. Enface OCTA images demonstrate a hyporeflective plaque extending from the supero temporal area of the optic disc to the perifoveal area. The 3.0 x 3.0 mm enface OCTA retina thickness map showed retinal thickening from oedema and corresponds to the area of the hyporeflective plaque with disruption of the deep retina and choroidal vascular plexi, the superficial retina plexus appeared spared. The FFA also shows adequate capillary perfusion in the area of retina whitening, with a prepapillary vascular loop. Improvement in retina capillary structure and flow is seen after four weeks.

INTRODUCTION

Optical Coherence Tomography Angiography (OCTA) is a novel and non-invasive technique for imaging retinal microvasculature by detecting changes, with respect to time, in reflectivity related to blood flow. OCTA gives 3-dimensional, noninvasive vascular mapping of the retina without the use of a dye. It is superior to the better-known fundus fluorescein angiography (FFA) being able to achieve vascular mapping in the retina and choroid in just a few seconds. With the OCTA distinct images of the superficial and deep retina vascular plexi are seen, as well as the outer retina and choriocapillaries at the same time. It also detects flow characteristics in retina and choroidal vessels but is, however, unable to detect vascular leakage. Since a dye is not used, it avoids the risk of systemic reactions to fluorescein dye, which can be devastating in rare cases [1–4]. With the advent of OCTA images, many
retina vascular disorders can now be better described. Recently Paracentral acute middle maculopathy (PAMM) characterized by deep retinal capillary ischemia, manifesting as hyperreflective bands within the middle retina on spectral-domain optical coherence tomography (SD-OCT) imaging has been identified [5, 6].

One of the main clinical signs of retina capillary ischaemia is a cotton wool spot. With OCTA we are able to see that cotton wool spots are caused predominantly by ischemia of the superficial capillary plexus, while PAMM is caused by ischemia predominantly of the deep capillary plexus (DCP). In PAMM, the hyper-reflective, band-like lesions in the middle retina, extend from the inner nuclear layer (INL)/outer plexiform layer junction to involve the full-thickness INL [6]. Over time, these lesions resolve with atrophy of the INL, resulting in persistent paracentral scotomas for the patient [7]. In the initial description, PAMM lesions were said to occur in isolation, but more recent reports describe them in conjunction with other retinovascular conditions, including branch (BRAO) and central (CRAO) retinal arterial occlusion, central retinal vein occlusion (CRVO), branch retina vein occlusion (BRVO), diabetic retinopathy, and sickle cell retinopathy [8].

OCTA of PAMM lesions demonstrate large areas of middle retinal hyperreflectivity in an arteriolar distribution with more extensive capillary dropout at the level of the deep plexus. The extent of the hyper reflective area is often dependent on the area of retina capillary affection and can be linear or wedge shaped [9]. This case reports the OCTA features of a young female in sub-Saharan Africa with PAMM. Optical Coherence Tomography Angiography (OCTA) features of a 33 year old female presenting with visual loss secondary to a paracentral acute middle maculopathy are discussed in this case-report.

CASE REPORT

A 33-year-old female professional presented to our hospital with a two week history of sudden visual loss in her left eye, with no preceding aura, nausea or vomiting, no antecedent trauma, redness, pain or previous episode. She is not a known diabetic or hypertensive and not on any regular medication. She is single with no family history of blindness, glaucoma or cataract. Examination revealed visual acuities of 6/6 OD and 6/60 OS. Both pupils were round and reactive with full range extraocular muscle movement. Anterior segments were normal, no rubeosisirides and intraocular pressures (IOP) were 19 mmHg OD, 22mmHg OS by applanation tonometry. Fundoscopy showed flat retinae with cup to disc ratio of 0.1 OU. There was optic disc swelling in the left with an area of whitening of the retina extending from the superotemporal disc margin towards the fovea suggestive of a left branch retinal arteriole occlusion, (Figure 1), however, FFA revealed (Figure 1) adequate capillary perfusion in the area of retina whitening, with a preapillary vascular loop.

On Fourier domain optical coherence tomography (FD-OCT) there was diffuse thickening of the neurosensory retina with increased reflectivity of the inner retinal layers from the surface of the retina to the photoreceptor layers (Figure 1). Using the Angiovue optical coherence tomography angiography (OCTA) software of the RTVue XR Avanti (Optovue, Inc., Fremont, CA), Full-thickness (internal limiting membrane to Bruch’s membrane) 3.0x3.0 mm OCT angiogram of the optic disc and done. This showed decreased capillary perfusion in the supero temporal area of the disc. The 3.0x3.0 mm en-face OCTA retina thickness map showed retinal thickening that corresponds to the decreased capillary perfusion and disruption of the choroidal vascular plexuses in the area of the retina oedema. The superficial vascular plexi are spared (Figure 1).

A diagnosis of paracentral acute middle maculopathy and elevated intraocular pressures was made in her left eye. Haematologic studies revealed, Haemoglobin 13.6 g/dl, PCV 39%, WBC 5,700 per mm₃ , Neutrophils 59%, Lymphocytes 33%, Monocytes 5%, Eosinophils 3%, Basophils 0% and Platelets 196,000. Erythrocyte sedimentation rate (ESR) was 10 by westergreen method. Total cholesterol 170, Triglycerides 30, HDL cholesterol 64, LDL cholesterol 100. Fasting blood sugar 5.3 mmol/l . BMI 29, Patient was also referred to her physician for a cardiovascular risk assessment.

She was commenced on a fixed combination of guttaetimol 0.5% and 2% dorzolamide twice daily, 250mg acetazolamide tablets twice daily with potassium supplement tablets. She was seen two weeks after initial presentation, visual acuities had improved to 6/18 OS and better-controlled IOPs of 13 mmHg OS. On fundoscopy , the retina whitening and optic disc swelling had improved significantly, this was confirmed on OCTA (Figure 2).

DISCUSSION

It is well established that the retina has a multilayered capillary network consisting of the superficial plexus at the level of the ganglion cell layer, and the intermediate and deep plexi at the superficial and deep edges of the inner nuclear layer. The middle retina including the inner nuclear layer and outer plexiform layer is felt to be a watershed zone that may be most vulnerable to ischemia of the deeper capillary plexi [4, 6]. PAMM is a spectral-domain optical coherence tomographic (SD-OCT) finding first reported in 2013 by Sarraf et al and interpreted as a possible more superficial variant of Acute Macular Neuroretinopathy (AMN).

It is characterized by hyperreflective band-like, multiple or isolated focal or diffuse lesions visible at the level of the inner nuclear layer (INL) in patients presenting with acute onset of negative scotoma. It is descriptively termed PAMM due to parafoveal position of
the causative grey lesions with near-infrared reflectance imaging, and the SD-OCT localization of involvement to the middle layer (INL) of the retina. Localized retinal capillary ischemia at the level of intermediate plexus is proposed as the mechanism underlying the development of these lesions [10].

OCTA is capable of demonstrating vascular changes in these different areas of the retina and peripapillary regions [4]. It may have a role in the early diagnosis of branch retina artery occlusions (BRAO), branch retina vein occlusion (BRVO), central retina vein occlusion (CRVO), central retina artery occlusion (CRAO) and more recently PAMM. Particularly in PAMM it can specifically identify the affected deep retina and choroidal vascular plexus, and the sparing of the superficial plexus [9], although further studies are needed to determine sensitivity and specificity.

Our case study describes the exceptional qualities of the OCTA as an investigatory modality in cases of PAMM. Although a relatively new tool, it gives information that can assist in understanding this pathology better. OCTA might be a more sensitive investigation tool to identify ischaemic retina areas when compared to FFA, it provides a snapshot in time, is non-invasive, acquires volumetric scans that can be segmented to specific depths, uses motion contrast instead of intravenous dye, can be obtained within seconds, provides accurate size and localization information, visualizes both the retinal and choroidal vasculature, and shows structural and blood flow information [9, 11]. OCTA has been shown to be a useful imaging modality for the evaluation of common ophthalmologic diseases such AMD, diabetic retinopathy, artery and vein occlusions, and glaucoma. FFA still remains the gold standard for evaluating ocular blood flow characteristics, however in our patient the main findings was that of an area of hypoflourescent appearance in the superotemporal portion of the optic disc suspicious of a focal kinking of a branch retina artery. There was no corresponding area of hypoflorescence despite the retina oedema and retina vascularization was relatively normal. The first differential considered was a BRAO, however, in most instances a BRAO occurs in individuals older than 65 years, secondary to an embolus, and FFA
will demonstrate an area of capillary dropout, although both diagnosis have been reported to occur together [8]. Emboli typically originate within vessels upstream where they dislodge and travel within the circulatory system to ultimately become lodged downstream in a vessel with a smaller lumen. The most common include cholesterol emboli from aorto-carotid atheromatous plaques, platelet-fibrin emboli from thrombotic disease, and calcific emboli from cardiac valvular disease. Various other endogenous emboli as well as exogenous emboli and nonembolic causes have been reported [12–22]. Other risk factors identified in young adults include cigarette smoking, bhectes disease, obesity, pregnancy, oral contraception use, hypercoagulable states (antithrombin III, protein C, S deficiencies) and intravenous drug abuse [23, 24]. The available results of systemic evaluation of our patient were not suggestive of any of the above. PAMM lesions have been reported to resolve over time, leaving atrophy of the INL, which may result in persistent paracentral scotomas for the patient [7]. Similarly when our patient was seen two weeks after initial presentation, visual acuities had improved to 6/18 OS and better controlled IOPs of 13 mmHg OS, on fundoscopy, the retina whitening and optic disc swelling had improved significantly (Figure 2).

The OCTA angiograms in our patient demonstrated a wedge-shaped area of capillary non-perfusion that correlates with the area of retina oedema. A similar case of vascular lesions not obvious on FFA but seen clearly on OCTA is reported for proliferative diabetic retinopathy by Ishibazawa A et al. [25] This might indicate that OCTA is a more sensitive tool to identify capillary non perfusion. The FFA also showed an area of a probable pre papillary arterial loop (Figure1b), this is likely a predisposing factor to vascular occlusion as also reported by Reichel E et al. [18]. To our knowledge our case report is the first describing PAMM in an individual in sub-Saharan Africa, most other cases report findings in caucasiens and individuals of Asian descent.

Some improvements expected in this technology are faster scanning speeds as well as larger fields of view with higher resolution. More studies are needed to determine OCTA’s utility in the clinical setting and to determine if this technology may offer a noninvasive option of visualizing the retinal vasculature in better detail for a series of other retina vascular disorders. However, at this point OCTA images are best used in conjunction with already existing imaging modalities like FFA and Indocyanine Green Angiography (ICG) for better results.

CONCLUSION

Angiovue optical coherence tomography angiography (OCTA) is a useful tool in evaluating retina vascular plexi affection in paracentral acute middle maculopathy, it gives adequate vascular details to enable diagnosis and assist with follow up.

REFERENCES


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Author Contributions
Olufemi Oladimeji Oderinlo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Adekunle Olubola Hassan – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ogugua Ndubuisi Okonkwo – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

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Written informed consent was obtained from the patient for publication of this case report.

Conflict of Interest
Authors declare no conflict of interest.

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