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Unilateral Acute Idiopathic Maculopathy Follow up by Optical Coherence Tomography Angiography a Case Report

Juan Gonzalo Sanchez[1], Milagros Guerra[1], Maria Carolina Sardi[1], Maria Adelaida Piedrahita[2], Claudia Patricia Acosta[1]

[1]National Institute of Research in Ophthalmology, Medellin, Colombia

[2]Faculty of Medicine, CES University, Medellín, Colombia

Email address:

mapipiedrahita@gmail.com (Maria Adelaida Piedrahita)

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Abstract: *Background:* It is described as a unilateral acute idiopathic maculopathy (UAIM) case, including spectral domain optical coherence tomography angiography (SD-OCTA) features and follow-up. *Case information:* the 24-week follow-up description by SD-OCTA of a 39-year-old woman with an episode of UAIM. SD-OCT showed at the beginning an upper yuxtafoveal diffuse hyperreflectivity of the outer nuclear layer (ONL), associated with a slight underlying serous detachment, and a choroidal thickness increase was observed too. *Results:* In SD-OCTA, a dark pattern was observed at the level of the choriocapillaris, with a progressive decrease in the size of this area with an almost complete resolution of the dark pattern during image follow-up a few weeks later. *Conclusions:* In the follow-up with SD-OCTA of a Unilateral Acute Idiopathic Maculopathy case, an impairment of the choriocapillaris was demonstrated that affected the outer retina with the spontaneous recovery of both. SD-OCTA can be a helpful tool to evaluate the recovery of choroidal flow for a longer time, knowing that the resolution of the changes evidenced at the choriocapillaris occurs more slowly, and complement the evaluation of these patients, with other tools that allow to accurately measure the choroidal blood flow to determine the etiology of this disease that remains controversial finally.

Keywords: Choriocapillaris, Choroides, Dark Pattern, Optical Coherence Tomography Angiography (OCT-A), Unilateral Acute Idiopathic Maculopathy (UAIM)

1. Introduction

UAIM is a rare disorder that presents with acute onset of unilateral central painless visual loss often associated with a prodromal flu-like illness. [1] Coxsackievirus infection is considered to be the cause of UAIM but there have been other cases in the literature where it was associated with different infections and vaccines. [2-4] The diagnostic criteria still remain somewhat ill-defined. [2, 5-7] It was originally described by Yannuzzi et al. [1] as an unilateral an acute central vision loss, followed by clinical spontaneous recover few days after.[1, 5, 8] It has been described an irregular hyperfluorescence and hypofluorescence that originate at the level of the retinal pigment epithelium (RPE) on the fluorescein angiography. They described following resolution of the disease process, most maculae have a bull's-eye pattern of pigmentary disturbance with late staining on fluorescein angiography. [1, 6].

Here is reported a case of a 24-week follow-up is presented using SD-OCTA to observe the evolution of an UAIM case.

2. Case Report

A 39- year-old woman in Medellin, Colombia, presented with a 3-day episode of sudden and painless visual loss on her right eye, without any other symptoms. She claims to have, two weeks before, ulcers in her mouth treated with acyclovir. The patient had a history of thyroidectomy for thyroid cancer four years ago and secondary hypothyroidism in treatment with levothyroxine—a history of Lasik treatment in both eyes

and amblyopia in her left eye by anisometropia. There was no other relevant medical and ocular data. The best corrected visual acuity was (Snellen chart) 20/200 in the right eye and 20/100 in the left eye. The slit lamp examination was normal. The dilated fundus examination revealed the right eye whitening of the yuxtafoveal upper retina, which projects superiorly in a torpedo shape that compromises the fovea. The ultra-widefield fluorescein angiography (UWF-FA) showed, in its early phase, an hyperfluorescence lesion in the superonasal border of the foveola, with an increase in leakage around the foveola at the pass of the contrast. The periphery without injuries. SD-OCT (Angiovue Optovue®) presented an upper yuxtafoveal diffuse hyperreflectivity of the outer nuclear layer (ONL) associated with a slight underlying serous detachment. The ellipsoid zone (EZ) looked irregular and seemed to be disrupted. There was an attenuation of the interdigitation of the underlying RPE, with the homogenous focal hyper transmission; Subfoveal choroidal thickness was increased (494 microns).

SD-OCTA, 3x3 scan showed at the level of the choriocapillaris, a subfoveal hyporreflectivity area that extended to the upper parafoveal sector. The superficial and deep capillary plexus were preserved, and the outer retinal was avascular. (Figure 1).

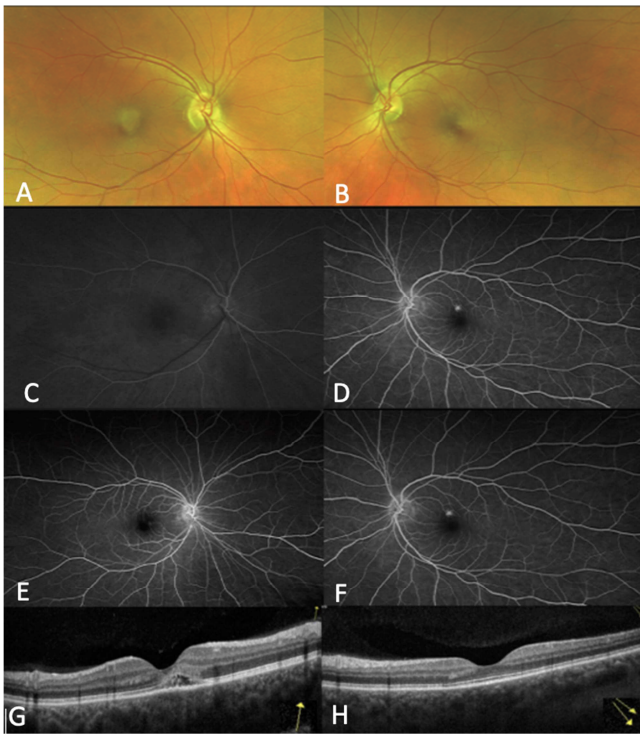

Figure 1. *Patient Baseline. (A-D) Right eye. (A) fundus photography, (B) UWF-FA early phase, (C) UWF-FA late phase, (D) OCT B Scan, (E-H) Left eye. (E) fundus photography, (F) UWF-FA early phase, (G) UWF-FA late phase, (H) OCT B Scan.*

At the dilated fundus examination, the left eye showed a hypopigmented superior parafoveal lesion. UWF-FA on the left eye showed a hyperfluorescence lesion of approximately 200 microns at the macular superonasal border of the fovea without leakage. SD-OCT presented an outer nuclear layer descent paracentral superior to the fovea. The External Limiting Membrane (ELM) and the EZ were not compromised. The subfoveal choroidal thickness was 423 microns.

Evolution:

In the visit of the fourth week, the visual symptoms improved spontaneously, and her best corrected right eye visual acuity was (Snellen chart) 20/40, and in the left eye remained 20/100. Right eye SD-OCT revealed resolution of the hyperreflectivity of the ONL, but with a slight focal descent. The ELM looked unaltered, but the image of the disrupted ellipsoid persisted, with an improvement of the hyper transmission and recovery in the regularity of the RPE. There was a decrease in choroidal thickness compared to the acute phase, measuring 476 microns. In OCTA, a decrease in the size of the hyporreflectivity area at the choriocapillaris level was observed (Figure 2).

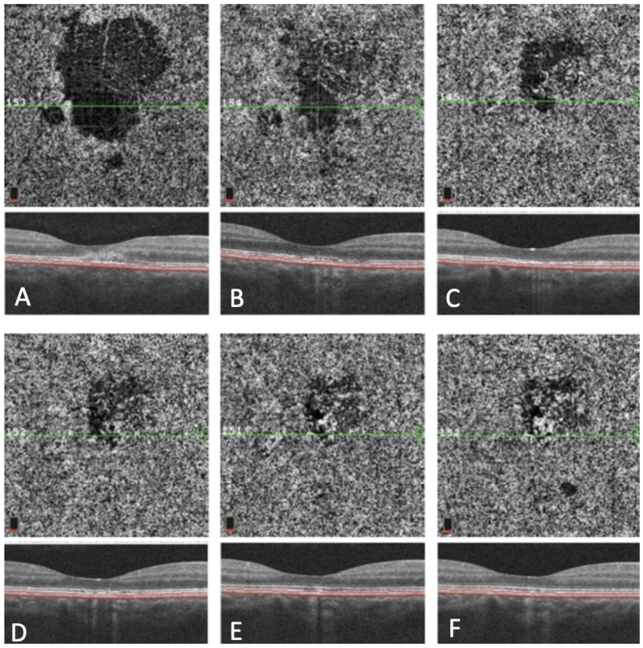

Figure 2. *Optical Coherence Tomography Angiography (OCTA) right eye evolution. (A) Initial, subfoveal hyporreflectivity area that extended to the upper parafoveal sector at the level of the choriocapillaris. (B-C) 1st- 4th week, notice a decrease in the size of the hyporreflectivity area (dark pattern) at the choriocapillaris level. (D) 8th week, the dark pattern seen before at the level of the choriocapillaris continued to diminish. (E-F) 18th -24th week, an almost complete resolution of the dark pattern at the choriocapillaris level was observed.*

The best corrected right eye visual acuity for the eighth-week visit was 20/30. Right eye ophthalmoscopy showed an increase in the intensity and size of the pigmentary changes in the area where a retina whitening used to be at the beginning. In FA, a hyperfluorescence lesion was observed from the early phases, which did not increase in intensity or size as the contrast passed (window defect). The SD-OCT looked like an RPE hypertrophy at the site of the lesion, with the recovery of the EZ. The subfoveal choroidal thickness decreased, with an observed thickness of 458 microns. At the follow-up with SD-OCTA, the dark pattern seen before at the

level of the choriocapillaris continued to diminish.

For the 24th week visit, the best corrected right eye visual acuity was (Snellen chart) 20/30. In the fundus photography of the right eye, it has observed subfoveal punctate hypopigmentary changes that alter FAZ, and the UWF-FA in its recirculation phase showed hyperfluorescence points that increase in intensity and not in size, without contrast leakage. SD-OCT showed an increase in the height of EPR hypertrophy with pigment migration to the supra-adjacent retina, surrounded by an area of hyper transmission and persistence of a focal disruption of the EZ. The choroidal thickness was 420 microns. Almost a complete resolution of the dark pattern at the choriocapillaris level in the SD-OCTA (Figure 3). UWF-FA, SD-OCT, and the measurement of subfoveal choroid thickness on the left eye remained unchanged over time.

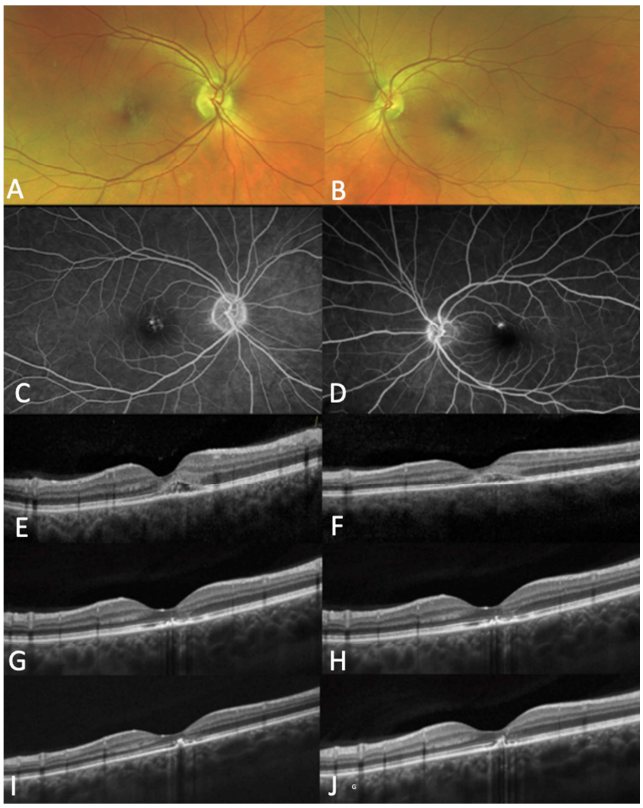

Figure 3. *24th week after the onset (A-D) and right eye evolution OCT B Scan (E-J). (A) Right eye fundus photography 24th week, (B) Left eye fundus photography 24th week, (C) Right eye UWF-FA24th week, (D) Left eye UWF-FA 24th week, (E) Initial OCT B Scan, (F) 1st week OCT B Scan, (G) 4th week OCT B Scan (H) 8th week OCT B Scan, (I) 18th week OCT B Scan right eye, (J) 24th week OCT B Scan right eye.*

3. Discussion

Acute idiopathic maculopathy is a self-limiting, usually unilateral disorder that presents with acute onset of central vision loss. [8] The diagnosis requires a combination of history, fundus examination, and imaging. The association with a previous flu disease, clinical or serologic evidence of coxsackievirus infection, history of N1H1 and COVID vaccination, and also infections like Yellow fever, Streptococcal Pharyngitis, and SARS-COv2 due to the ACE-2 receptors located in the retina, can also be followed by UAIM as it has been reported in the past year. [2–5, 9, 10] This report presents a patient with the typical symptoms of UAIM, with findings of fundoscopic examination consistent with those previously described in others cases reports. [1, 6] In addition, OCT and OCTA findings supported clinical diagnosis and evidenced benign behavior. To our knowledge, this is the first case with follow-up by SD-OCTA of 6 months without any complication evidenced.

SD-OCT observed it at the beginning of an upper yuxtafoveal diffuse hyperreflectivity of the outer nuclear layer (ONL), associated with a slight underlying serous detachment. The ellipsoidal band looked irregular and disrupted, and the measurement of choroidal thickness was increased, as previously reported. [11, 12].

The SD-OCT follow-up showed an improvement of the initial injury with an area in the RPE that looked like hypertrophy, recovery of the ellipsoid, and a progressive decrease of choroidal thickness. The residual damage to the retinal pigment epithelium (RPE) was morphologically characteristic by Yannuzzi et al. [1] in a series of cases and also was described as not typical of any previous maculopathies. In our case, the bull's-eye pattern was not observed as classically described. However, the pigmentary changes were observed at the level of RPE.

It is observed in the SD-OCT and SD-OCTA an increase of choroidal thickness and a dark pattern at the choriocapillaris level, respectively, in the acute phase, as described previously. In the follow-up, this case showed an almost complete spontaneous resolution from the fourth month of both: choroidal thickness and dark pattern without any treatment, so the recovery could be close to the four months. SD-OCT usually cannot detect the blood flow at the choriocapillaris level. However, in our case using SD-OCTA, there was no limitation on the visualization of the choriocapillaris through the RPE. This tool showed the impairment at the choroidal flow expressed as a "dark pattern" at the choriocapillaris level.

Others have proposed choroidal involvement during UAIM; Haruta et al. [7] demonstrated thickened choroid in the foveal region; they speculated that the primary site of inflammation might be the choroid. Later on, Srour et al. [13], with SD-OCT, suggested that the choroid could be the primary originating anatomical site of the disease process.

On the other hand, Hashimoto et al. [14], using laser speckle flowgraph to evaluate the mean blur rate (MBR), a quantitative index of relative blood flow velocity, observed that patients with UAIM in the acute phase presented a decrease of choroidal blood flow velocity concerning the contralateral eye; this blood flow velocity normalizing as the improvement occurs around the third month of follow up. Indocyanine green angiography (ICGA) was described as an hypofluorescence in the acute phase that corresponds to this finding.

Recently Nicolo et al. [15] showed, in a case with follow-up by SS-OCTA for a month, a dark pattern at the level of choriocapillaris in the acute phase that persisted after a month

and confirmed that a hyperperfusion of the choriocapillaris might be involved primarily in the disease process.

In literature, it has been suggested that in some patients, the inflammatory process may be associated with RPE alterations and choriocapillaris ischemia, with a breakdown of the outer blood-retina barrier and the development of macular detachment. [5].

It suggests that the transient impairment of blood flow in the choriocapillaris, both by ischemia and by hyperperfusion, will produce an alteration in the signal reproduced by the OCTA, resulting in a dark pattern. The complete normalization of the choroidal blood flow will be represented in the OCTA as the complete resolution of the dark pattern, observing how white noise is expected.

It is recommended to follow up with OCTA for a longer time, knowing that the resolution of the changes evidenced at the choriocapillaris occurs more slowly, and complement the evaluation of these patients with other tools that allow an accurate measure of the choroidal blood flow to determine the etiology of this disease that remains controversial finally.

4. Conclusion

UAIM can be defined as a pathology with choroidal circulation impairment whose mechanism remains unknown, with secondary involvement of the external retina that can have favorable evolution without treatment. SD-OCTA can be a valuable tool to evaluate the recovery of choroidal flow.

Consent

A well-written informed consent was obtained from the patient, along with the approval of the institutional ethics committee. An explicit written consent regarding publishing the photographs was taken from the patient.

Conflicts of Interest

The authors declare that there is no conflict of interest.

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