

Focal telangiectatic perifoveal venule associated with sequential documented vitreofoveal adherence/traction, transient intraretinal cystoid spaces and vitreofoveal release

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Background: Perifoveal retinal vascular abnormalities (PRVA) are typically associated with retinal vascular occlusive or inflammatory diseases. PRVA may also develop in macular telangiectasia type 1 (MACTEL1), a congenital or developmental vascular disorder consisting in focal, exudative dilatations which affect both superficial and deep focal capillary density plexuses, preferentially localized in deep capillary plexus as OCTA studies reports. Also, it tends to respond well to anti-VEGF therapy (1,2). PRVA may also be associated with PEVAC (perifoveal exudative vascular anomalous complex), defined as a large unilateral solitary perifoveal aneurysm without adjacent telangiectasia in absence of retinal vascular or inflammatory diseases and do not respond well to anti-VEGF therapy (1). Herein, we present an unpublished case of PRVA particular type characterized as individual telangiectatic perifoveal venule segment (ITEPVES), sparing the foveal thickness onset. Interestingly, we could notice a transient macular intraretinal fluid (as a central bouquet traction) consecutively to spontaneous vitreofoveal traction release. We believe that the diagnosis of ITEPVES may be classified in the spectrum of idiopathic PRVA.

1. Sacconi, Riccardo; Freund, K. Bailey; Yannuzzi, Lawrence A.; Dolz-Marco, Rosa; Souied, Eric; Capuano, Vittorio; Semoun, Oudy; Phasukkijwatana, Nopasak; Sarraf, David; Carnevali, Adriano; Querques, Lea; Bandello, Francesco; Querques, Giuseppe (2017). *The Expanded Spectrum of Perifoveal Exudative Vascular Anomalous Complex*. *American Journal of Ophthalmology*, (), S0002939417304336-.doi:10.1016/j.ajo.2017.10.009

2. Gass JD, Oyakawa RT (1982) Idiopathic juxtafoveolar retinal telangiectasis. *JAMA Ophthalmology* 100:769–780. <https://doi.org/10.1001/archophth.1982.01030030773010>

Case Report : 65-year-old healthier woman with unilateral sudden central visual loss 30' onset. BCVA was 20/70 in affected right eye. Ocular fundus examination on right eye (besides) revealed presence of slightly irregular dilated perifoveal venule at 6 o'clock local as a "venous beading shape". Fellow eye unremarkable.

Methods: The patient have undergone ophthalmic examination including multimodal evaluation with color fundus retinography, fluorescein angiography (FA), near-infrared reflectance (near-IR) scanning laser ophthalmoscopy, blue autofluorescence, spectral-domain optical coherence tomography (SD-OCT) and OCT angiography (Figures 1-3).



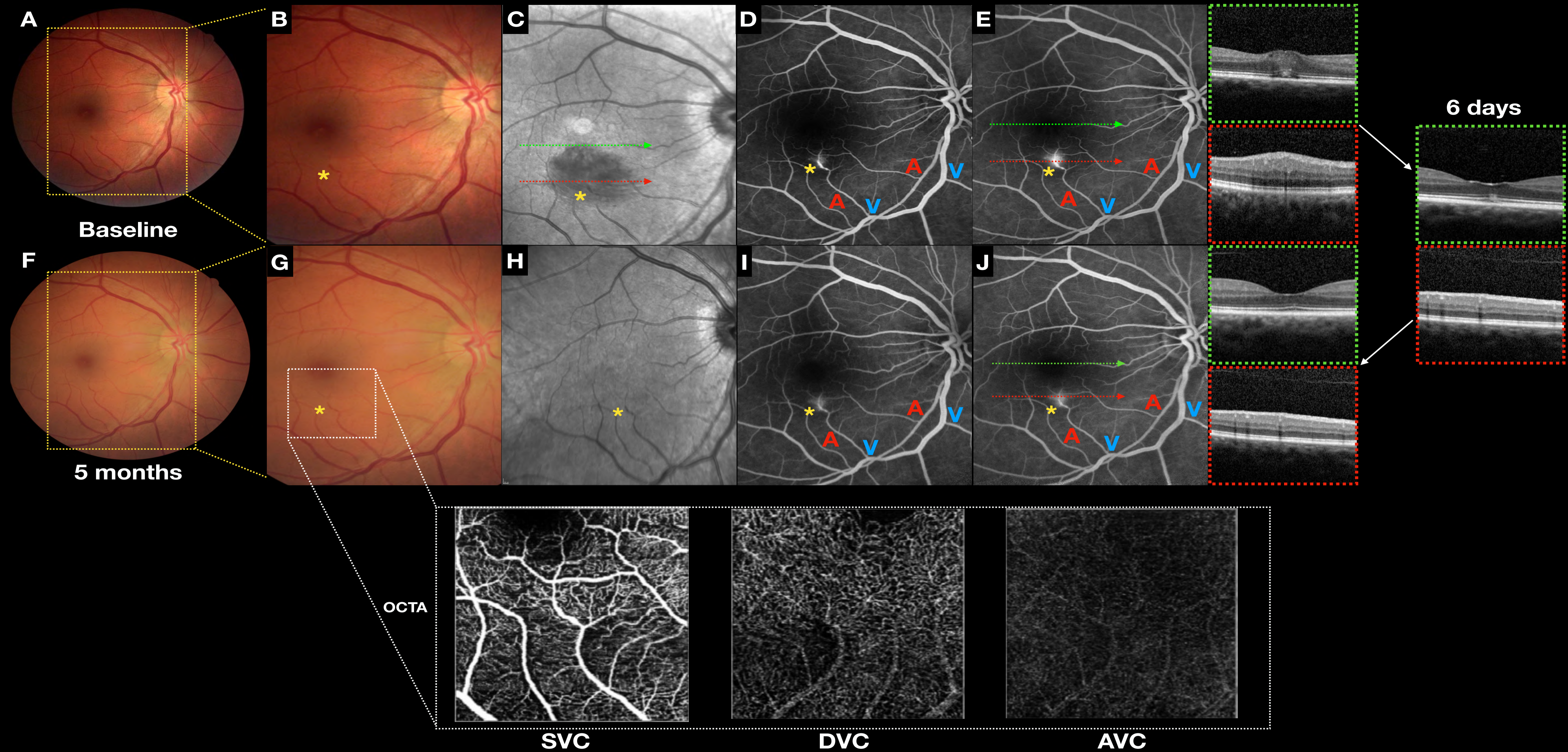


Figure 1. Multimodal imaging Assessment. *Baseline imaging 30 minutes forward ocular symptoms onset (A-E).* (A) Color fundus retinography. (B) Zoomed macular area color retinography and telangiectatic vessel juxtafoveal inferiorly (yellow asterisk). (C) Hyporeflectance near-infrared patch surrounding telangiectatic vessel. Red and green dashed arrow corresponding to telangiectatic vessel and center foveal sectioning, respectively (red and green square-dashed SD-OCT imaging). (D) Early (28") FA revealing initial hyperfluorescence correlated with superficial telangiectatic intraretinal vessel (yellow asterisk). (E) Late (4') FA showing hyperfluorescence staining of telangiectatic vessel (yellow asterisk). Red letter A and blue letter V related to superficial arteries and veins vessels, respectively. Red and green dashed arrow corresponding to telangiectatic vessel and center foveal sectioning, respectively (red and green square-dashed SD-OCT imaging). Next, the 6th day SD-OCT follow-up. *Five months imaging following (F-N).* (F) Color fundus retinography. (G) Zoomed macular area color retinography and telangiectatic vessel juxtafoveal inferiorly (yellow asterisk). White square-dashed area focusing juxtafoveal inferiorly macular area relating to OCT-A analysis. (H) Near-IR regular reflectance. Red and green dashed arrow corresponding to telangiectatic vessel and center foveal sectioning, respectively (red and green square-dashed SD-OCT imaging). (I) Early (38") FA revealing initial weak hyperfluorescence correlated with superficial telangiectatic intraretinal vessel (yellow asterisk). (J) Late (5') FA showing weak hyperfluorescence staining of telangiectatic vessel (yellow asterisk). Red letter A and blue letter V related to superficial arteries and veins vessels, respectively. Red and green dashed arrow corresponding to telangiectatic vessel and center foveal sectioning, respectively (red and green square-dashed SD-OCT imaging). (L) OCT-A from superficial vascular complex involving the telangiectatic (pink asterisk) vessel juxtafoveal inferiorly. (M) OCT-A from deep vascular complex not related to telangiectatic vessel. (N) OCT-A from avascular complex in the outer retinal layer.

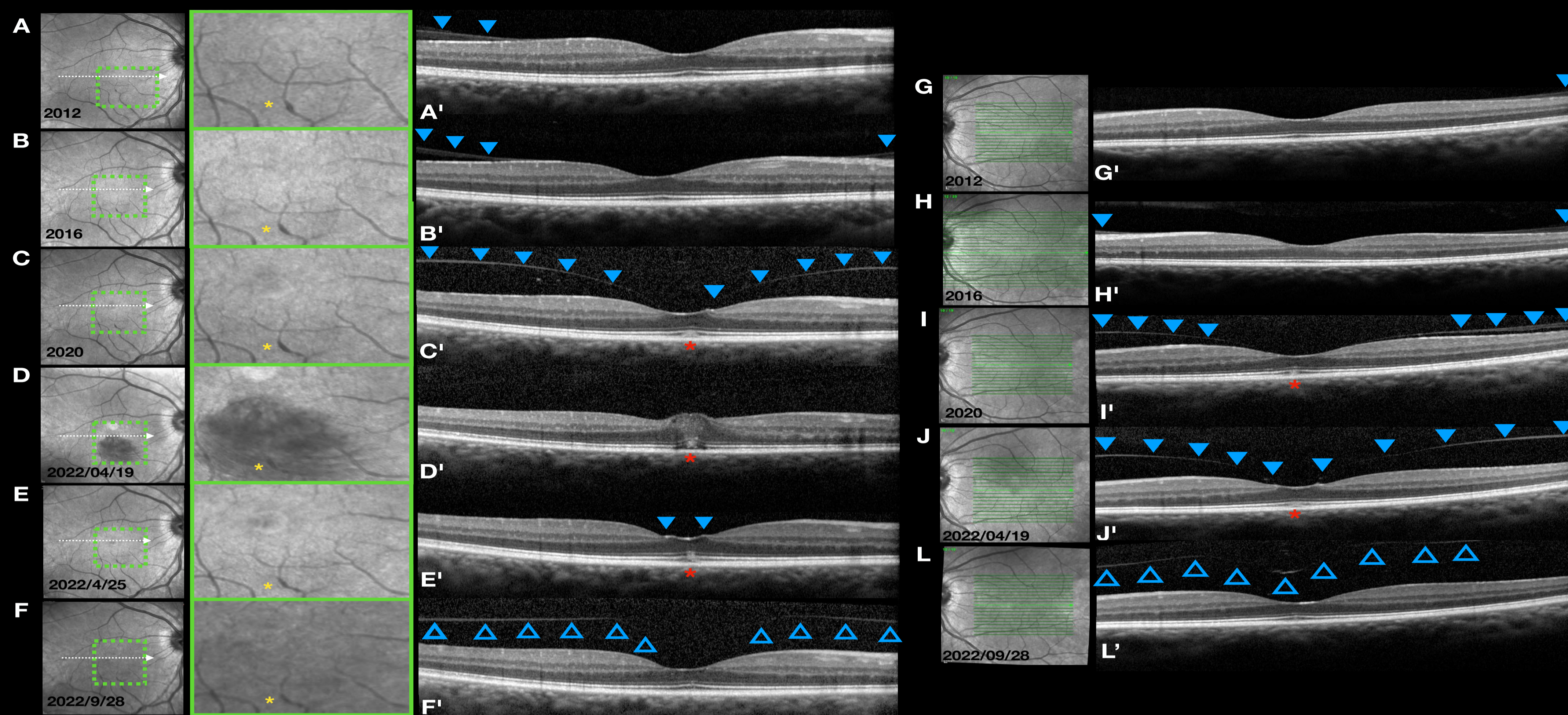


Figure 2 Ten years near-IR and SD-OCT follow-up from both eyes. Right eye (A-F). (A) 2012 year near-IR inferiorly juxtafoveal area selected imaging (green square-dashed). Next, near-IR green square zoomed focusing telangiectatic vessel (yellow asterisk). (A') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing initial posterior vitreous adhesion (PVA) temporally (blue arrowhead). (B) 2016 year near-IR inferiorly juxtafoveal area selected imaging (green square-dashed). Next, near-IR green square zoomed focusing telangiectatic vessel (yellow asterisk). (B') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing nasal PVA (blue arrowhead) and temporal PVA associated. (C) 2020 year near-IR inferiorly juxtafoveal area selected imaging (green square-dashed). Next, near-IR green square zoomed focusing telangiectatic vessel (yellow asterisk). (C') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing PVA relief progress with nasally subtle foveolar inward traction (blue arrowhead) and "cotton-ball" sign below (red asterisk). (D) 2022 april 19th baseline ocular symptoms onset after 30 minutes, near-IR inferiorly juxtafoveal area selected imaging (green square-dashed). Next, green square zoomed imaging showing hyporeflectance near-IR area surrounding telangiectatic vessel (yellow asterisk). (D') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing foveolar inward traction (blue arrowhead), entire intraretinal thickness SD-OCT hyperreflective and "cotton-ball" sign below (red asterisk). (E) 2022 april 25th, six days forward with normal near-IR reflectance recovery at selected area imaging (green square-dashed). Next, near-IR green square zoomed focusing telangiectatic vessel (yellow asterisk). (E') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing traction solving progress with subtle foveolar inward traction (blue arrowhead) and presence of "cotton-ball" sign below (red asterisk). (F) 2022 september 28th, five months forward with normal near-IR reflectance at selected area imaging (green square-dashed). Next, near-IR green square zoomed focusing telangiectatic vessel (yellow asterisk). (F') SD-OCT horizontal sectioning at foveolar centre (white dashed arrow) revealing complete macular posterior vitreous detachment (PVD) (blue arrowhead) and EZ/IZ regularity in the outer retinal layers. *Left eye (G-L).* (G-G') 2012 year near-IR and SD-OCT horizontal sectioning at foveolar centre (green line arrow) revealing PVA temporally (blue arrowhead). (H-H') 2016 year near-IR and SD-OCT horizontal sectioning at foveolar centre (green line arrow) revealing PVA temporally and nasally (blue arrowhead). (I-I') 2020 year near-IR and SD-OCT horizontal sectioning at foveolar centre (green line arrow) revealing PVA progress temporally and nasally (blue arrowhead) and "cotton-ball" sign below (red asterisk). (J-J') 2022 april 19th near-IR and SD-OCT horizontal sectioning at foveolar centre (green line arrow) revealing vitreous macular traction (blue arrowhead) and "cotton-ball" sign below (red asterisk). (L-L') 2022 september 28th near-IR and SD-OCT horizontal sectioning at foveolar centre (green line arrow) revealing complete macular posterior vitreous detachment (PVD) (blue arrowhead) and EZ/IZ regularity in the outer retinal layers

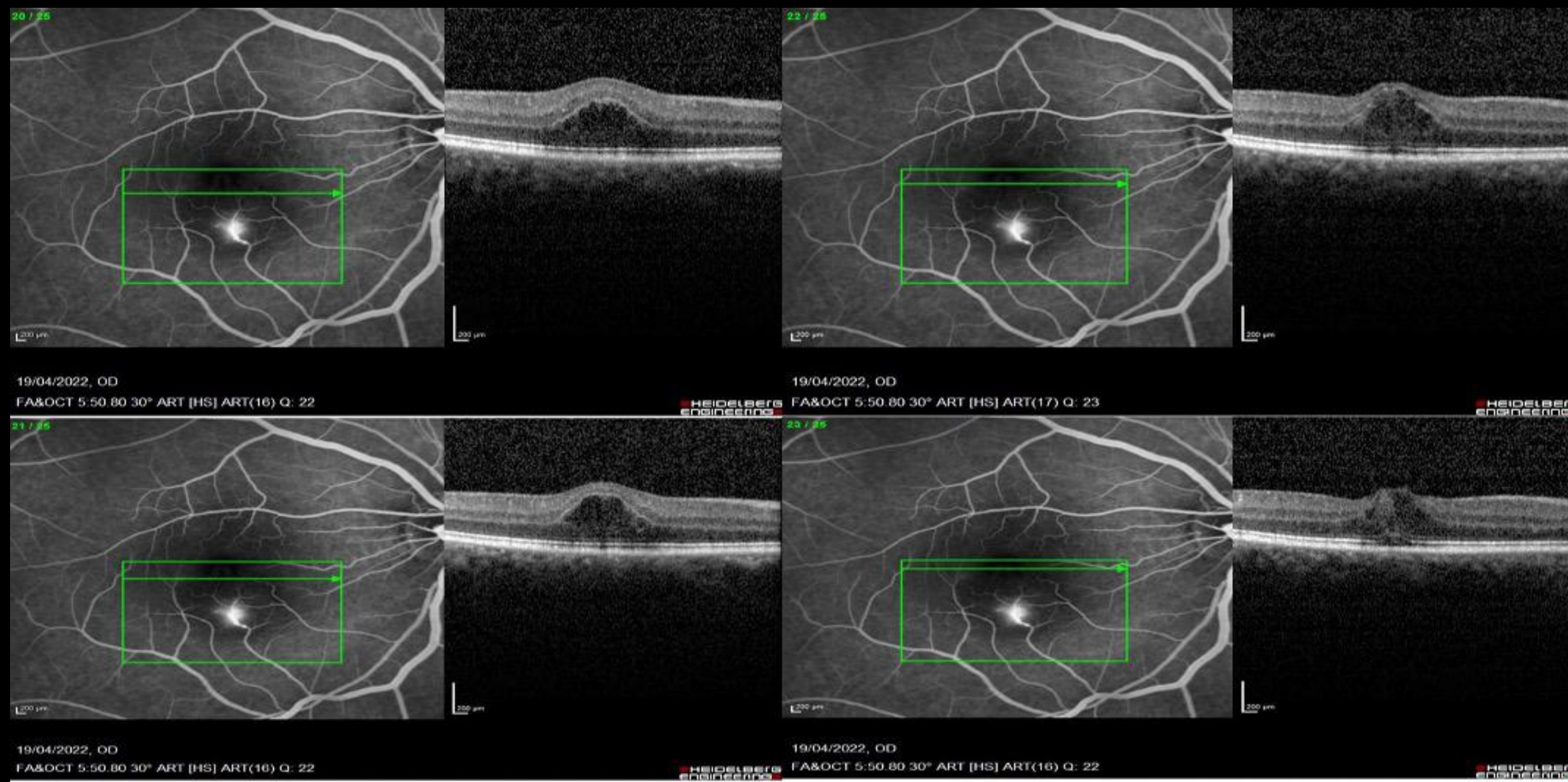


Figure 3 From 6 to 12 o'clock, this area showed increasing sized multiple cystic cavities affecting the outer nuclear and plexiform layers with a coexistent detached (or recently detached) hyaloid

Discussion: MACTEL1 was the first diagnosis impression based on the initial fundoscopic and angiographic findings. However, MACTEL 1 usually occurs in young male patients and is characterized by multiple capillary, venular, and arteriolar aneurysms involving the temporal side of the fovea (3). Small branch vein occlusion could also be cogitated based on fundus examination, but a careful multimodal exams discard the presence of blood retinal hemorrhages and/or retinal ischemic status. PEVAC was also ruled out due to the absence of a perifoveal aneurysm. The distinct central foveal edema detected was triggered to inward vitreous traction pulling the superficial abnormal telangiectatic venule. The mechanical traction is confirmed with the presence of “cotton ball sign” on the outer retinal layers. Finally, the patient was followed closely for 6 months with serial multimodal imaging. OCT-A confirmed the location of the telangiectatic venule at the superficial retinal level and a repeated fluorescein angiogram enhanced the vascular abnormality with less hyperfluorescence and perivascular leakage.

Conclusion: We conclude this case with the possibility of atypical forms of macular telangiectasias, as well as the possibility of intraretinal cysts caused by foveal vitreous traction.