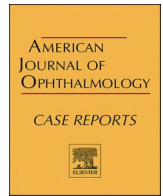


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Non-invasive imaging of a choroidal macrovessel

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ABSTRACT

Purpose: To describe novel anatomic findings of an apparent choroidal macrovessel, originally misdiagnosed as a choroidal tumor, using non-invasive imaging tools.

Observations: Initial ophthalmic examination revealed an elevated hypopigmented choroidal mass in the macular area, with a serpentine track extending temporally to the equator. Enhanced depth imaging optical coherence tomography (EDI-OCT) revealed an optically hollow lesion just outside the choroid-scleral junction (CSJ), indenting the retina and compressing the choroid from the scleral side. Optical coherence tomography angiography (OCTA) at the choroidal level showed relative low flow within the lesion. En face OCT at the level of the choroid demonstrated similar reflectivity to the physiological adjacent choroidal vessels.

Conclusion and importance: Non-invasive imaging can be used to demonstrate the presence and anatomy of a choroidal macrovessel. OCTA is presented as a useful diagnostic imaging test that can distinguish this lesion from alternative diagnoses without the use of dye injection. In addition to the previously published reports of such vessels in the choroid, we suggest a possible anatomic variant infra-choroidal location of a macrovessel and hypothesize its origin.

1. Introduction

The entity “Choroidal macrovessel” was first described in 2011 by Lima et al.¹ as a large, tortuous anomalous vascular lesion in the choroid with early filling and hypocyanescence in the late phase on indocyanine green angiography (ICG) without leakage. In the Dalvin et al.² review of existing reports of choroidal macrovessels, they are found to predominantly affect middle to older aged white women. In this article we report a case of a choroidal macrovessel in a non-white, Asian male, in a variant infra-choroidal location, with non-invasive imaging including fundus color photography, fundus autofluorescence (FAF), enhanced depth imaging optical coherence tomography (EDI-OCT), en face OCT and optical coherence tomography angiography (OCTA) as diagnostic methods. We highlight the advantage of OCTA as a diagnostic tool not requiring dye injection with high resolution to detect changes in the chorio-retinal vasculature and blood vessel flow.

2. Case report

A 69-year-old Asian male with medical history of Hypertension, Coronary Artery Disease and low myopia (spherical equivalent of −2.50D) was referred for a possible choroidal tumor in his left eye. The

patient denied flashes, floaters pain or discomfort in his left eye. Visual acuity was OD 20/40 and OS 20/100 with intraocular pressures of 12 mmHg in his right eye and 13 mmHg in his left eye. Anterior segment evaluation revealed prominent nuclear cataracts in both eyes, NS 2+ in the right and NS 3+ in the left eye. Funduscopy examination of the right eye was unremarkable. Funduscopy examination of the left eye revealed an elevated, hypopigmented choroidal mass in the macular area, with a serpentine hypopigmented track extending temporally to the equator (Fig. 1A). FAF showed a healthy retinal pigment epithelium (RPE) along the lesion (Fig. 1B). Infrared reflectance image (Fig. 1C) and EDI-OCT scan (Spectralis OCT2, Heidelberg Engineering Inc., Heidelberg, Germany) that crossed the lesion below the fovea and in two places temporally (Fig. 1D and E), demonstrated a optically hollow lesion just outside the choroid-scleral junction (CSJ) with an estimated measurement of 400 μm at widest girth markedly compressing the choroid from the scleral side with mild elevation and thickening of the overlying RPE, an intact but indented outer retina and a tapering width without any point of contact with the choroid itself, suggesting an infra-choroidal location. The patient had a myocardial infarction three months prior to the assessment and voluntarily denied dye-based angiography (fluorescein angiography (FA) and/or indocyanine green angiography (ICGA)). OCTA (Optovue RTVue XR Avanti, Optovue, Inc., Fremont,

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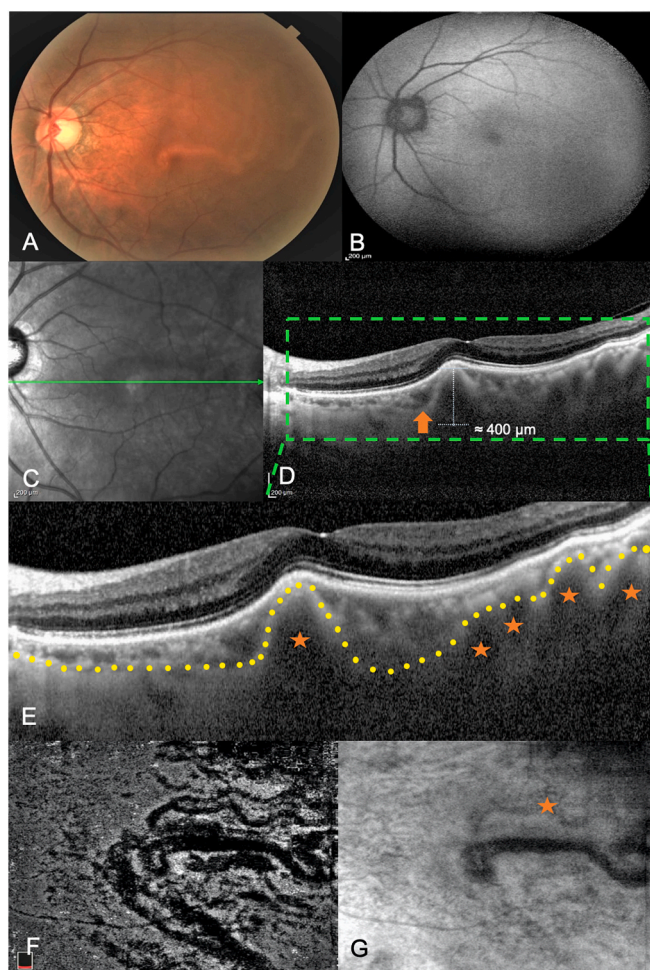


Fig. 1. (A) Fundus color photography revealed a tortuous, hypopigmented serpentine track originating from an elevated lesion in the macular area and extending temporally. Media opacity is due to cataract. (B) Fundus autofluorescence image showed a healthy retinal pigment epithelium along the lesion. (C) Infrared reflectance image and (D) EDI-OCT scan that crossed the lesion below the fovea and in two places temporally showed an optically hollow lesion just outside the choroid-scleral junction with an estimated measurement of 400 μm at widest girth markedly compressing the choroid from the scleral side. Looking at the lateral wall of the vessel, the choroid-scleral junction is more evident (orange arrowhead) and the location of the macrovessel, in fact, is infra-choroidal. (E) Magnified scan showed mild elevation and thickening of the overlying RPE, an intact but indented outer retina and a tapering width without any point of contact of the macrovessel (orange asterisks) with the choroid itself. The choroid-scleral junction is highlighted in dashed yellow. (F) OCTA at the choroidal level showed relative low flow within the lesion. (G) En face OCT at the choroid showed that the reflectivity of the lesion was similar to that of the physiological adjacent choroidal vessels (orange asterisk). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

CA, USA) at the choroidal level (Fig. 1F) showed relative low flow within the lesion. En face OCT (Fig. 1G) of the choroid showed that the reflectivity of the lesion was similar to that of the physiological adjacent choroidal vessels. With the information given by non-invasive imaging tools, a diagnosis of an infra-choroidal macrovessel was made. Observation of the macrovessel in his left eye and cataract surgery in both eyes was proposed to improve the visual acuity of the patient.

3. Discussion

Choroidal macrovessels are a rare condition with only twelve

previously reported cases in the literature (Table 1). The track-like nature of these lesions with accompanying retinal RPE changes can lead to confusion with parasitic tracks, such as ophthalmomyiasis, choroidal vascular neoplasms, retinochoroidal anastomosis and inflammatory choroidopathies, among others.³

Choroidal macrovessels seem to be a lesion of adulthood, with patients in their middle to older age (range 39–80 years), affecting predominantly females, most of them Caucasian women. This is a report of a choroidal macrovessel in an Asian, and only the second in a male, suggesting a broader range in gender and race. New cases will help establish the epidemiology further.

Choroidal macrovessels seem to be lesions that remain unnoticed because of their asymptomatic, subtle clinical findings, although metamorphopsia and blurry vision have been reported in four cases.^{2–5} When visual symptoms are present, they could be explained by a spectrum of clinical presentations, which range from mild EZ and RPE changes³ to mounding and hyperpigmentation of the RPE, debris, subretinal fluid (SRF), and changes in the outer nuclear layer thickness.⁶ Although observation is the treatment of choice, other options might be considered when subretinal fluid is present in the fovea.²

A choroidal macrovessel appears ophthalmoscopically as a single, hypopigmented, tortuous, dilated vessel extending from the central macula toward the equator in a predominantly horizontal orientation with a serpentine path along the temporal meridian. The macrovessel is most prominent in the perifoveal region as an ampulla that can resemble an elevated mass giving the impression of a choroidal tumor.²

After reviewing the fundus color photographs of the previous cases and the one shown in this report, there seems to be a relationship between choroidal macrovessels and myopia. Myopia is known to be associated with thin choroid and large choroidal vessels occasionally causing RPE elevation,⁷ however, the diameter of the vessels in myopia does not reach the $140 \pm 40 \mu\text{m}^2$ seen in macrovessels. We identified a tessellated fundus appearance with visualization of choroidal vasculature and peripapillary atrophy in eleven of thirteen cases. Nevertheless, more studies that detail the refractive status of the patients are needed to assess this feature as a risk factor for the presentation of a macrovessel.

Diagnostic imaging tests including invasive test such as FA and ICGA, and non-invasive tests such as fundus color photography, FAF, EDI-OCT, en face OCT and OCTA are useful in the distinction of a choroidal macrovessel from other conditions. Although invasive tests have been widely used, non-invasive imaging tools can be used as well to describe the features of this condition as reported in three previous cases.^{3,8,9}

EDI-OCT is an important diagnostic tool that helps determine the choroidal vascular nature of the retinal lesion. Typically the OCT scan shows an enlarged, hollow structure with elevation in the macula area, accompanied by compression of the overlying choroid, elevation of the RPE and irregularity of the overlying EZ.³ However, as discussed above, the presentation varies among patients. In our case, although the outer retina was indented, there was no presence of subretinal fluid and the EZ was unaltered, which explains the lack of additional visual symptoms in the patient, aside from those conditioned by the cataract. Likewise, there are no reported cases associated with aneurysm, exudation, pigment epithelial detachment, or choroidal neovascularization.⁶ After a detailed review of the EDI-OCT scan of our case, the location of the macrovessel appears to differ from previous descriptions. Looking at the vessel “wall”, we see it extends outside with what appears to be the CSJ. Thus, the vessel itself must be posteriorly in the sclera, just outside the choroid, in what we call an infra-choroidal location. It is also notable how the vessel markedly compresses the choroid from the scleral side, not described in other reports. The strategy used to assess the infra-choroidal location was evaluating the lateral wall of the macrovessel and its relationship with the CSJ. We noticed that the CSJ was more evident on the lateral wall of the vessel and its location was, in fact, below the choroid, as detailed in Fig. 1D (see arrowhead). Our hypothesis for infra-choroidal location is that at least some macrovessels may be aberrant posterior ciliary branches that ‘never made it’, i.e.,

Table 1
Features of choroidal macrovessel in patients based on a literature review.

No.	Authors Year published	Demographics			Clinical Features			OCT				FA	ICGA		Other findings
		Age (years)	Sex	Race	Symptoms	Visual Acuity	Tessellated fundus/ Peripapillary atrophy	RPE elevation	Irregular EZ	SRF	OCTA		Initial phase	Late phase	
1	Lima et al. 2011	42	M	White	ND	1.0	+/+	+	-	-	ND	ND	Normal	Hypo	
2	Ehrles et al. 2014	76	F	ND	Asymptomatic	0.66	+/+	+	+	-	ND	Hyper	Hyper	ND	
3	Choudry et al. 2016	42	F	White	Metamorphopsia	ND	+/+	+	+	-	ND	ND	ND	ND	
4	Pichi et al. 2016	ND	ND	ND	ND	ND	+/+	+	+	-	ND	ND	ND	ND	
5	Kovach 2016	70s	F	White	Asymptomatic	0.66	+/+	+	+	-	ND	ND	ND	ND	
6	Hamptom et al. 2017	80	F	ND	ND	ND	+/+	+	+	-	Low flow	ND	ND	ND	
7	Mahroo et al. 2017	79	F	ND	ND	ND	-/-	+	+	-	ND	ND	Hyper	ND	
8	Dalvin et al. 2018	55	F	White	Asymptomatic	0.8	-/-	+	+	+	ND	Hyper	Hyper	ND	
9	Dalvin et al. 2018	68	F	White	Blurry vision	0.8	+/+	+	+	+	ND	Hyper	Hyper	ND	
10	Casalino et al. 2019	65	F	White	Asymptomatic	1.0	+/+	+	+	+	Low flow	Hyper	Hyper	ND	
11	Mori et al. 2020	79	F	ND	Metamorphopsia	0.4*	+/+	+	+	-	Low flow	Hyper	Hyper	ND	AV connection
12	Katakoa et al. 2020	39	F	ND	Blurry vision	1.0	+/+	+	+	-	ND	Hyper	Hyper	Hypo	LSFG Hyper- perfusion
13	Otero-Marquez et al. This case	69	M	Asian	Asymptomatic	0.2*	+/+	+	-	-	Low flow	ND	ND	ND	

EZ, ellipsoid zone; F, female; FA, fluorescein angiography; hyper, hyperfluorescence; hypo, hypofluorescence; ICGA, indocyanine green angiography; LSFG, laser speckle flowgraphy; M, male; ND, no data; OCT, optical coherence tomography; OCTA optical coherence tomography angiography; RPE, retinal pigment epithelium; SRF, subretinal fluid.

*Visual Acuity conditioned by cataract.

never penetrated into the choroid itself, have no branches, and instead came to a bulbous terminus near the fovea, remaining in the sclera as an infra-choroidal vessel.

OCTA is a novel diagnostic tool in the diagnosis of choroidal macrovessels. Hampton et al.¹⁰ were the first to show the relatively low-flow of a macrovessel in the choriocapillaris slab with OCTA. More recently, Casalino et al.¹¹ and Mori et al.⁵ published another two cases of a choroidal macrovessel with OCTA also revealing relatively low flow. In our case the low flow within the macrovessel is also evident, unexpected if it were an arteriole, but which would be explained by the lack of an outflow, i.e., the blood has “no place to go”, with a closed bulbous termination and no connection to the rest of the choroid. However, a recent publication⁵ has shown a possible connection between a macrovessel and a vortex vein, suggesting a choroidal arteriovenous malformation. Moreover a possible focal hyper-perfusion within a macrovessel was recently demonstrated by Kataoka et al.⁴ using laser speckle flowgraphy. Thus, further observation and clinical-pathological studies are needed to confirm our impression.

4. Conclusion

Choroidal macrovessels are a rare, underdiagnosed condition mostly described in middle to older aged Caucasian women, but not exclusively. In most of the cases macrovessels do not produce visual symptoms although metamorphopsia or blurry vision can occur if subretinal fluid underlying the fovea is present. There seems to be a relationship with myopia, and more information from the refractive status of the previous cases, as well as reporting new cases would help in this regard. In our case the location of the macrovessel appears to infra-choroidal, and our hypothesis is that at least some macrovessels may be aberrant posterior ciliary branches that never penetrated into the choroid itself, have no branches, lack of an outflow, have low flow and instead came to a bulbous terminus near the fovea, remaining in the sclera as an infra-choroidal vessel. Choroidal macrovessels are a rare clinical entity, and non-invasive imaging can be useful to facilitate the diagnosis. OCTA is presented as a diagnostic imaging test that can distinguish this lesion from alternative diagnoses without the use of dye injection in patients who refuse its use or in whom it is contraindicated.

Patient consent

The patient consented to publication of the case orally. This report does not contain any personal information that could lead to the identification of the patient.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interestCOI

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