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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this correspondence.

Rare case of extramacular choroidal macrovessel

Choroidal macrovessel (CM) is a rare, mostly unilateral vascular anomaly of the choroidal circulation.[1] These abnormally large vessels may cause disturbances in the overlying retinal pigment epithelium (RPE) and the development of subretinal fluid. Some CMs can even masquerade as a choroidal tumour or parasitic infestation.[1–3] Reports showing early filling of CMs during indocyanine green angiography (ICGA) suggest that most CMs are arterial in nature.[1,3,4] The short posterior ciliary circulation is comprised of a medial and lateral posterior choroidal artery (PCA)[5,6]; the latter may represent the origin of most CMs and may explain why almost all previously reported CMs arise in the macula. An extramacular origin for CM is rare, and to our knowledge, there are only 2 cases described in the literature. Of these 2 cases, only 1 was illustrated with retinal imaging, which appears to show an origin of the anomalous vessel within the macula.[1]

In this correspondence, we describe and illustrate an extramacular CM that clearly originates in an extramacular location superior to the optic disc, as shown with ultra-wide-field (UWF) ICGA. This work was completed at the retinal disorders and ophthalmic genetics division at the Stein Eye Institute, University of California–Los Angeles.

A 58-year-old patient was referred for an evaluation of floaters and photopsia in the left eye for 5 days. Past medical history included hypertension and hypothyroidism secondary to thyroidectomy to treat thyroid cancer 8 years prior. Ocular history was remarkable for anisometropic myopia with a spherical equivalent of −2.25 D in the right eye and −6.50 D in the left eye.

On examination, visual acuity was 20/25 OD and 20/20 OS with normal anterior segments OU. Ophthalmoscopy demonstrated RPE mottling superior to the optic disc OS (Fig. 1A). UWF fundus autofluorescence showed a corresponding area of hyperautofluorescence (Fig. 1B). Optical coherence tomography B-scans through this region showed a large (380 μm in height) choroidal vessel with overlying subretinal fluid, focal elevation of the RPE–Bruch membrane complex, and elongated photoreceptor outer segments (Fig. 2). UWF ICGA revealed early arterial filling of a large dilated and tortuous extramacular CM originating superior to the optic disc and extending into the superior periphery without late leakage or staining (Fig. 3A). There was marked asymmetry of choroidal venous drainage, with reduced venous drainage directed toward the inferonasal vortex ampulla (Fig. 3B).

Otero-Marquez et al.[7] reported a possible association of CM with myopia. They reviewed the fundus images of 13 cases of CM and identified features of high myopia, including fundus tessellation and peripapillary atrophy in 11 of these patients. The patient reported herein exhibited high myopia with a refraction of −6.50 D OS. Of interest, retinal arterial tortuosity was identified only in this subject's left eye. The coexistence of CM with retinal arterial tortuosity

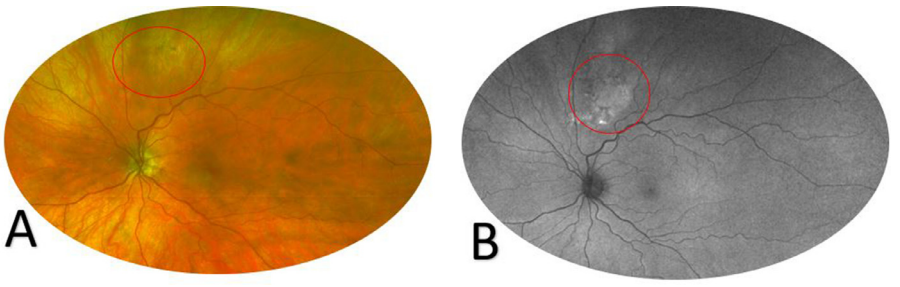

Fig. 1—(A) Ultra-wide-field colour image of the left eye illustrating retinal pigment epithelium mottling (red circle) superior to the optic disc. (B) Ultra-wide-field fundus autofluorescence showing the corresponding area, which is hyperautofluorescent (red circle).

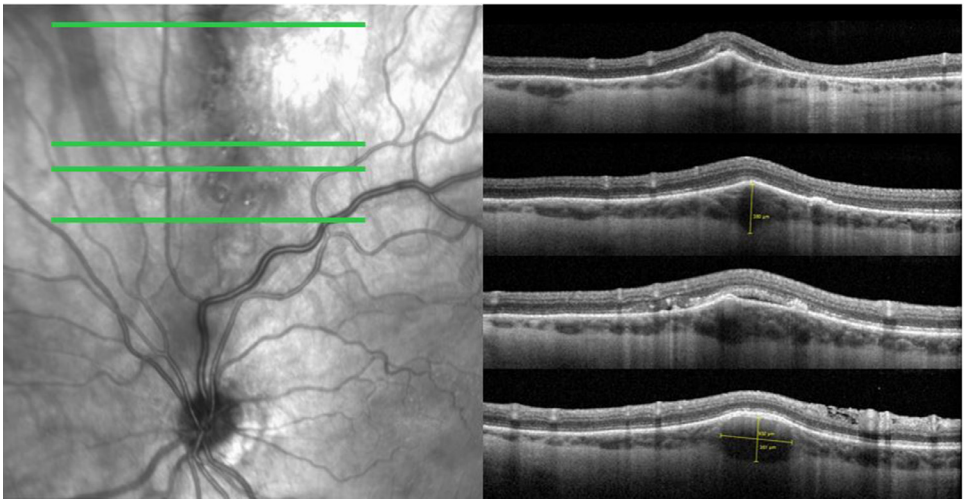

Fig. 2—Near-infrared reflectance (left) and spectral-domain optical coherence tomography (OCT) B-scans (right). The OCT B-scans show the choroidal macrovessel to measure 380 μm in greatest vertical diameter and 932 μm in greatest horizontal diameter. There are areas of overlying subretinal fluid, focal elevation of the retinal pigment epithelium–Bruch membrane complex, and elongated photoreceptor outer segments. The green lines in the near-infrared reflectance show the location of the OCT B-scans.

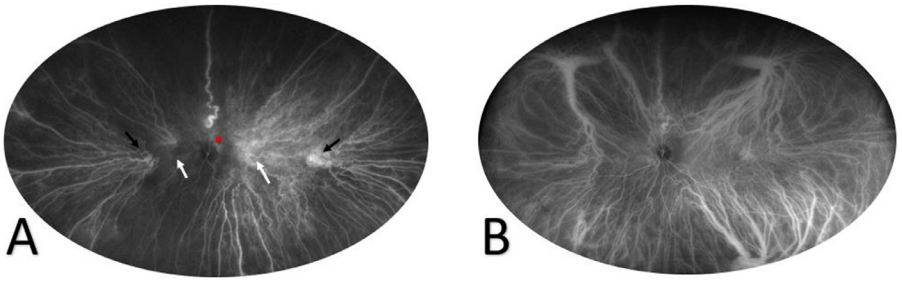

Fig. 3—(A) Ultra-wide-field indocyanine green angiography showing filling of the dilated and tortuous extramacular choroidal macrovessel originating superior to the optic disc and extending into the superior periphery without late leakage or staining. Note the lateral and medial distal short posterior ciliary arteries (white arrows) and the long posterior ciliary arteries (black arrows), which all exhibit a triangular distribution. The paraoptic vessels represent the short posterior ciliary arteries that originate around the optic disc (red asterisk). (B) There is marked asymmetry of the choroidal venous drainage, with reduced venous drainage directed toward the inferonasal vortex ampulla.

may be coincidental or may be related to an underlying vascular disorder involving both systems.

The presence of RPE mottling and subretinal fluid can be associated with CM.[1,3,8] These pathoanatomical complications may develop from compression of the overlying inner choroid leading to RPE dysfunction. A similar pathogenesis may explain the development of pigmentary changes and fluid in eyes with choroidal nevi or overlying dilated veins (pachyvessels) in eyes with pachychoroid disease.[9,10]

Hayreh[5,6] studied the PCA circulation and noted the presence of typically 1 to 5 PCAs. The lateral and medial PCAs (of the short posterior ciliary system) are most commonly identified (97%—100% of eyes), but the superior PCA is an uncommon branch that exists in 9% of eyes. The PCAs branch into short and long PCAs. The short PCAs pierce the sclera in 2 locations. The paraoptic short PCAs enter adjacent to the optic disc and the distal short PCAs enter nasal or temporal to the disc. In contrast, the long lateral PCA enters the sclera temporal to the macula. Use of UWF ICGA in our case clearly demonstrates the medial and lateral distal short PCAs as well as the long PCAs. Remarkably, a single superior PCA that is dilated and tortuous is also identified and represents the CM (Fig. 3). This anomalous pattern of circulation can be detected in less than 10% of normal subjects. Most previously reported CMs originate from the temporal distal short PCA, which may explain the remarkable macular predominance for this vascular entity.

To our knowledge, this is the only reported case of a CM originating from the superior PCA and the only illustration of this uncommon vascular pattern with ICGA. The extramacular origin of the CM is rare and can be easily overlooked because of the peripheral location, which may explain the scarcity of reports describing this vascular anomaly. The novelty of this case is further underscored by the associated presence of unilateral retinal arterial tortuosity in

the ipsilateral eye. Multimodal retinal imaging including UWF ICGA and fundus autofluorescence can facilitate identification of these abnormalities, which can be complicated by the development of pigmentary alterations and even serous retinal detachment with the potential to cause vision loss.

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Footnotes and Disclosure

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Retinal pigment epithelium apertures associated with subretinal fluid and acquired vitellifom lesions in non-neovascular age-related macular degeneration

Classic features of non-neovascular age-related macular degeneration (AMD) include drusen, drusenoid retinal pigment epithelial (RPE) detachment, and RPE atrophy. More recent studies have shown that intra- and subretinal fluid also may complicate the non-neovascular form of AMD.[1]

RPE aperture has been described as an atypical feature of eyes with non-neovascular AMD. Querques et al.[2] studied 10 eyes, most with drusenoid pigment epithelial detachment (PED) that developed such RPE defects. Similarly, Giannakaki-Zimmermann et al.[3] reported 7 eyes with apertures, of which 4 displayed drusenoid PED, whereas Yoshinaga et al.[4] studied 5 cases of RPE defect all associated with drusenoid PED.

In this correspondence, we describe 7 cases of RPE aperture that developed in eyes with acquired vitelliform lesions and subretinal fluid in the absence of drusenoid PED at the time of presentation. In this retrospective case series, eyes with RPE aperture associated with subretinal fluid (SRF) secondary to non-neovascular AMD were captured. The study was approved by the institutional ethics committee and adhered to the tenets of Declaration of Helsinki. Inclusion criteria were age greater than 55 years, presence of classic features of non-neovascular AMD including macular drusen and/or outer retina and RPE atrophy associated with an RPE aperture, and the absence of macular neovascularization (MNV) with multimodal imaging including optical coherence tomography (OCT; Spectralis, Heidelberg Engineering, Heidelberg, Germany, or Avanti, Optovue, Calif.), OCT angiography (Spectralis, Heidelberg Engineering or