Genesis of Retinal-Choroidal Anastomosis in Macular Telangiectasia Type 2: A Longitudinal Analysis

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Purpose:
To longitudinally characterize structural and angiographic findings of retinal choroidal anastomosis (RCA) in macular telangiectasia type 2 (MacTel2).

Methods:
Design:
Retrospective chart review at a community-based, retina referral practice.

Subjects:
MacTel2 patients with two or more visits and minimum 2 months follow-up were included.

Testing:
Complete ophthalmologic examination, fundus photography, fluorescein angiography, spectral-domain optical coherence tomography (SD-OCT), and projection-resolved OCT angiography.

Main Outcome Measures:
The development of RCA and associated retinal defects.

Results:
There were 24 eyes of 12 patients with mean age 61.1 (±7.3 [SD]) years and mean follow-up 652 (range 81-1511) days. There were 5 eyes diagnosed with new RCA after not having any previously. RCA was diagnosed in 8 (33%) eyes initially and eventually in 13 (54%). In the 8 eyes initially diagnosed with RCA, there was an increase in number of RCAs and outer retinal hyperreflective lesions over the follow-up. The RCAs were found in clusters, generally temporal to the fovea. RCA was uniformly associated with complete retinal subsidence, defined as the loss of substance in the outer nuclear layer such that the middle layers of the retina sank down toward the retinal pigment epithelium, was also present in each case. Each RCA colocalized with a region of hyperreflectivity, previously termed an outer retinal hyperreflective lesion (ORHL). There was a lack of fluorescein leakage, lipid, hemorrhage, or any structural correlates to fluid exudation, and no signs of subretinal or subretinal pigment epithelial neovascularization.

Conclusions:
De novo RCA development appears to be associated with retinal subsidence, and after RCAs form, they increase in number and topographic distribution over time. Initially after formation, there was no sign of exudation. These data suggest that descent of the deep capillary plexus occurs with RCA, devoid any evidence of subretinal neovascularization in MacTel2.