

Multimodal Imaging in the Diagnosis of Exophytic Juxtapapillary Retinal Capillary Hemangioblastoma

Abstract

Purpose: Exophytic juxtapapillary retinal capillary hemangioblastoma (JRCH) can be difficult to diagnose. We explore the value of multimodal imaging to aid in the diagnosis.

Methods: Medical records and multimodal imaging studies were reviewed on all patients diagnosed with RCH at Bascom Palmer Eye Institute, Miami, Florida, between January 2013 and December 2019. Patients with exophytic lesions within 2 mm of the disc were included. One patient from the Baylor College of Medicine, Houston, Texas was included. Patient demographics, referring diagnosis, history of von Hippel-Lindau syndrome, initial and last visual acuity, and treatments were recorded. Fundus photography, fundus autofluorescence, fluorescein angiography, indocyanine green angiography, optical coherence tomography (OCT), OCT angiography, and B-scan images were reviewed.

Results: Twelve patients were identified with exophytic JRCH. The mean age was 54 years (range 38-73 years). Five patients had von Hippel-Lindau syndrome. The most common referral diagnoses were choroidal neovascularization and neuroretinitis. Imaging features included nodular outer retinal thickening with shadowing and intra-/subretinal fluid on OCT, hypoautofluorescence on fundus autofluorescence, middle to outer retinal hypervascularity on OCT angiography, early hyperfluorescence with late leakage on fluorescein angiography, and lack of choroidal vascular lesion on indocyanine green angiography. Treatments included photodynamic therapy (6 patients), intravitreal anti-vascular endothelial growth factor therapy (6 patients), argon laser photocoagulation (2 patients), intravitreal or sub-Tenon's triamcinolone (3 patients), and observation (4 patients).

Conclusions: A key to the accurate diagnosis of exophytic JRCH is its intraretinal location, typically involving the outer retinal layers, which results in a clinical appearance that is distinct from the more common and easily recognizable endophytic RCH. Multimodal imaging can aid in ruling out choroidal neovascularization and disc edema by demonstrating an absence of involvement of those structures.