

Brief summary of the literature

David J. Browning MD, PhD

Patchy ischemic retinal whitening describes faint macular ischemic whitening in a perivenular distribution found in acute nonischemic central retinal vein occlusion (CRVO), usually found in younger patients. It is associated with relative scotomas, with cilioretinal artery insufficiency in approximately 50% of cases, and usually resolves in 3-6 weeks with incomplete resolution of the scotomas. The fluorescein angiogram is typically unremarkable. OCT has revealed paracentral acute middle maculopathy correlating with the ischemic whitening, mainly involving the inner nuclear layer of the retina, and particularly striking in the en face view. Laboratory workup is typically shows no abnormality. The prognosis is good, and no particular treatment is necessary, although serial monitoring is a good strategy to detect any unexpected deterioration.

Issues raised by the case

1. The main clinical issue is recognition. In contrast to most cases of central retinal vein occlusion, there is a relative paucity of intraretinal hemorrhage, but venous dilation is typically evident. Patients are usually younger than the median age reported in large series of CRVO.
This patient had no risk factors for CRVO, which is often the case. Blood pressure was normal. She was taking no medications.
2. The pathoanatomy and pathophysiology of PIRW and PAMM are controversial.
The competing hypotheses are the serial and parallel conceptions of the intermediate and deep capillary plexuses in the inner retina. The serial hypothesis posits that the blood in the intermediate and deep plexuses is more deoxygenated because these plexuses are downstream of the superficial capillary plexus. The parallel hypothesis denies a serial anatomy and instead posits that the cells of the inner nuclear layer have higher oxygen demands and deplete intracapillary oxygen more than retinal cells of the ganglion cell layer surrounding the superficial plexus.

References

1. Browning DJ. Patchy ischemic retinal whitening in acute central retinal vein occlusion. 2002; 109: 2154-9.
2. Rahimy E, et al. Paracentral acute middle maculopathy in nonischemic central retinal vein occlusion. Am J Ophthalmol. 2014; 158:372-80.
3. Fouquet S, et al. The 3D retinal capillary circulation in pigs reveals a predominantly serial organization. IOVS 2017; 58:5754-63.
4. McLeod D. Misery perfusion, diffusive oxygen shunting and interarterial watershed infarction underlie oxygenation-based hypoperfusion maculopathy. Am J Ophthalmol. 2019; 205:153-64.

Slide 1

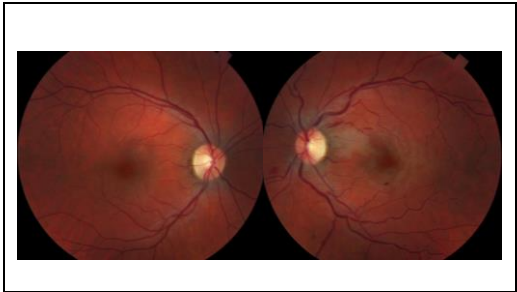
Patchy Ischemic Retinal
Whitening

Slide 2

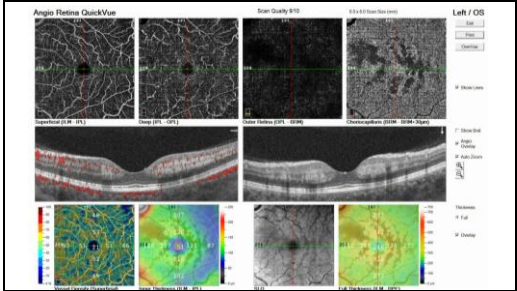
9/3/2019 28 year woman with sudden blurring and a paracentral relative scotoma OS for 3 days

- Negative review of systems
- On no medications
- Denies drug use, OTC use, smoking
- Occasionally drinks alcohol
- No recent overseas travel
- BP 109/70
- VA 20/20 OU
- IOP 15 OU
- Slit lamp normal OU
- Fundus as shown

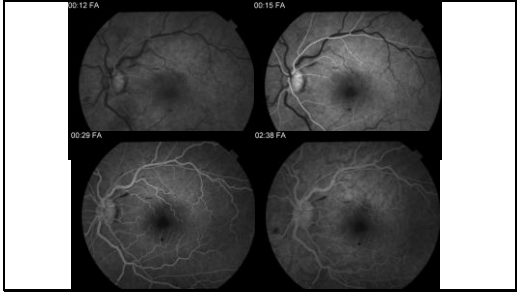
Slide 3



Slide 7



Slide 8



Slide 9

What is your diagnosis?
What workup would you obtain?
What treatment would you recommend?
What prognosis would you give her?

Slide 10

Patchy Ischemic Retinal Whitening in Acute Central Retinal Vein Occlusion

David J. Browning, MD, PhD
Ophthalmology 2002; 109:2154-59

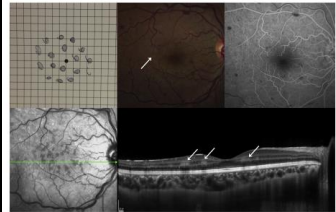
Objective: To describe a new fundus abnormality in nonischemic central retinal vein occlusion and its pathophysiologic basis.
Design: Retrospective, observational case series.
Participants: Eleven consecutive patients from a community-based retina referral practice who had central retinal vein occlusion (CRVO) and patchy ischemic retinal whitening (PIRW) and 235 consecutive patients from

- Occurs in young, healthy patients with CRVO
- Can be seen in ~5% of CRVOs
- PIRW occurs in a perivenular distribution
- Good visual outcomes
- Minimal FA correlates
- Resolves in 2-4 weeks
- Cilioretinal arteriolar insufficiency is common (~50%)
- Laboratory workup is normal

Slide 11

Paracentral Acute Middle Maculopathy in Nonischemic Central Retinal Vein Occlusion

DHAN KAHMY, DAVID SARRAF, MICHAEL L. DOHLIN, JOHN D. PITCHER, AND ALLEN C. HO
AJCO 2014; 158:372-80



- Hypothesized that intermediate and deep capillary plexuses differentially express ischemia compared to the superficial plexus
- Proposed a serial organization of SCP, ICP, DCP

Slide 12

Anatomy and Pathology/Oncology
The 3D Retinal Capillary Circulation in Pigs Reveals a Predominant Serial Organization
IOVS 2017; 58:5754-5763

Stephane Frappet,¹ Ophélie Vacca,¹ Florian Scanziani,¹ and Michel Pequet^{1,2}
¹Centre de la Vision, Institut National de la Santé et de la Recherche Médicale, U1016, Paris, France
²Centre National de la Recherche Scientifique, Institut Pasteur, 75231 Paris, France

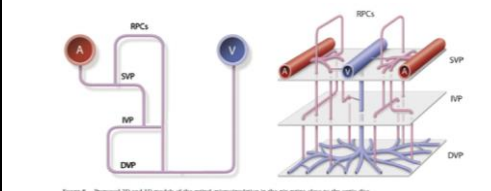


Figure 8. Proposed 2D and 3D models of the retinal microcirculation in the pig retina close to the optic disc.

Slide 16



Slide 17



Slide 18

Questions, Comments, Discussion
