Brief summary of the literature

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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

**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**

![Fundus Images]
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

- 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

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

AJO 2014; 158:372-80

Slide 12

The 3D Retinal Capillary Circulation in Vivo Reveals a Peculiar Arteriovenous Organization

IOVS 2017; 58:5754-5763
9/18/2019 Blurring improving, scotoma smaller

- VA 20/20 OU
- IOP 15 OU
- Pupils: No RAPD

9/30/2019 Continues to improve
- Advised to return in 1 year or prn