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INTRODUCTION

The cilioretinal artery is a common congenital variant of the retinal blood vessels with a prevalence range of 23.8-50.0% in the United States¹. Location, length and diameter of cilioretinal arteries vary and the cilioretinal arteries located temporally to the optic disc are more likely to supply blood to the macula¹. Cilioretinal artery occlusion (CLRAO) is a rare condition with unique characteristic features like vision loss from blood flow disruption to the macula, occurring in only about 5-7% of retinal artery occlusion cases². We reviewed 2 case studies and one retrospective study of cilioretinal artery occlusion in which symptoms and visual acuity resolved to 20/20.

CASE 1

Patient 1: 42-year-old female with recent diagnosis of hypertension, presented to the ED with persistent binocular vision changes described as a “gray highlighter” in the left eye for three days.

HPI: denied pain, diplopia, flashing lights, new floaters, and curtain-like vision loss.

PMHx, SHx, FHx: none

VS: BP 166/112

• **NVA:** 20/20 OU

• **IOP:** 20, 21

• **CVE, Color plates:** full OU

• **DFE OS:** area of pallor of the inner retina, temporal to the optic disc, with white, fluffy cotton wool spots

Upon retinal exam:

• **NVA:** 20/20 OU

• **DFE:** well-demarcated whitening in the cilioretinal artery distribution OS

• **OCT OS:** revealed normal foveal contour with hyper-reflectivity in the inner nasal layer of the macula.

• **F/A OS:** hypo-perfusion in the nasal macula in the cilioretinal artery distribution.

CASE 1 CONTINUED...

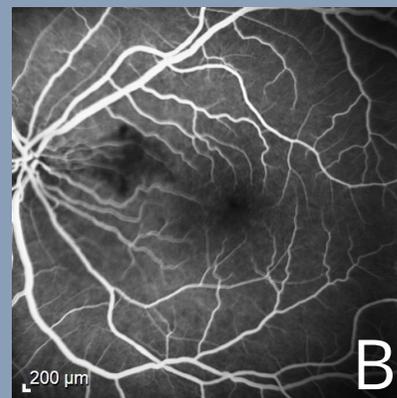
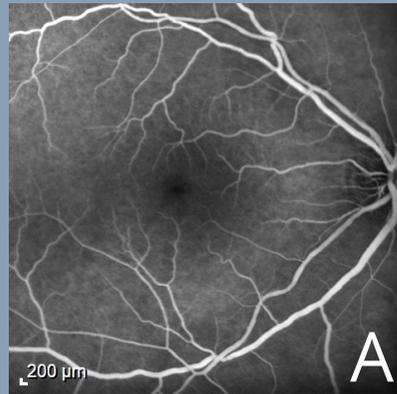


Figure 1: Fluorescein angiography of retina
A: Right eye. B: Left eye: hypo-perfusion in nasal macula

LITERATURE REVIEW

CASE 2: Cilioretinal arterial occlusion phenomenon: A rare cause of loss of vision in pregnancy⁴

A 21-year-old primigravid female at 30 weeks presented with sudden painless central vision loss OD.

- **VA:** 6/36 (20/120), 6/9 (20/30)
- **DFE OD:** linear hemorrhage in the superior nasal quadrant with pallor between the macula and optic disc.

- Small dot hemorrhage in the right temporal quadrant with mild blurring of optic disc margin

CASE 3: Cilioretinal obstruction during pregnancy⁵

A retrospective study was conducted including 135 patients between 2007 and 2012; 20 patients with cilioretinal artery occlusion. **Two** of these patients were pregnant and with no other predisposing risk factors.

Patient 1: A 34 y/o female at 20 weeks gestation reported a spot in OD without a reduction in VA.

• **VA:** 1 (20/20) OU with normal anterior poles.

• **DFE OD:** cotton-like spot 15mm in diameter and a visualized white funnel in the cilioretinal artery

• **OCT OD:** edema and campimetry showed central scotoma

• After 5 weeks the retinal edema disappeared, retinal atrophy was present in the area of the cotton like spot.

Patient 2: A 13 y/o patient, 16 weeks pregnant, reported a spot in OD after one day.

• **VA:** 1 (20/20) OU with normal anterior pole.

• **DFE:** retinal infarct in the inter-papillomacular area

• **VF:** showed a central scotoma OD

• After 4 weeks symptoms improved.

DISCUSSION

- Isolated cilioretinal artery occlusion (without CRAO) is rare and represent only 5-7% of retinal artery occlusion case
- In a longitudinal study, isolated cilioretinal artery occlusions found that 85% of patients were correctable to only 20/50 visual acuity³
- The visual outcome is based on location and its supply to the macula
- The differential diagnosis includes hypertension, hypercoagulable state, and vasculitis (e.g., giant cell arteritis)

CONCLUSION

- Predisposing factors to retinal obstruction include hypertension, atherosclerosis, hypercoagulability and embolism.
- All four patients with CLRAO within these cases had temporal cilioretinal arteries in which a decline in VA would be initially suspected because temporal cilioretinal arteries are more likely to supply blood to the macula.
- It is rare for a patient with an isolated CLRAO to present with 20/20 vision when the blood flow to the macula is impacted.
- Clinicians should be aware that the location and macular supply of the cilioretinal artery will contribute to the visual outcome.

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