Central Retinal Artery Occlusion in a Patient with Polyarteritis Nodosa

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INTRODUCTION

- Central retinal artery occlusion (CRAO) is the sudden blockage of the central retinal artery. Compromised blood supply to the retina leads to progressive retinal cellular damage and rapid and painless vision loss.[1]
- Polyarteritis nodosa (PAN) is a necrotizing inflammatory disease affecting medium-sized arteries in various organs, including skin, kidneys, and gastrointestinal tract.[2,3,4] Delayed or inadequate interventions can lead to a poor PAN prognosis.[5]
- CRAO is not a well-known complication of PAN. Here, we describe a case of CRAO presumed secondary to PAN. We describe the clinical course and diagnostic measures and discuss medical management of this rare entity.

CASE PRESENTATION

A 65-year-old female with history of Sjogren's syndrome, asthma, hypereosinophilia syndrome, chronic sinusitis and irritable bowel syndrome presented to emergency department with acute, painless vision loss in the right eye.

One week prior, the patient had presented to a different hospital for right upper quadrant abdominal discomfort. She was found to have elevated liver function tests, and her abdominal computed tomography (CT) scan revealed diffuse periportal edema, scattered hypodense lesions throughout her liver, and mildly dilated common bile duct without choledocholithiasis. Her viral hepatitis panel was negative. As a possible etiology, a side effect of her new asthma medication mepolizumab vs relapse of hypereosinophilia syndrome was identified. The patient noticed worsening fatigue and headaches with the new medication. She was instructed to hold mepolizumab until rheumatology consultation and discharged from the outside hospital on low-dose oxycodone and a prednisone taper.

On the day of presentation, the patient reported her symptoms had started the day prior to arrival and rapidly progressed to complete vision loss. She complained of transient vision loss in the left eye which resolved after a few hours without intervention. She also complained of jaw claudication. She noted night sweats, weight loss, cold intolerance, and numbness in her right second and third toes.

DIAGNOSTIC WORKUP

- Fundus photography and macular optical coherence tomography scan for a diagnosis of CRAO.
- Temporal artery biopsy and blood work to evaluate for other vasculitides.
- CT angiogram of abdomen to diagnose PAN.

RESULTS

- CT angiography of the head and neck was negative for any acute pathology.
- Temporal artery biopsy was negative for focal granulomatous inflammation.

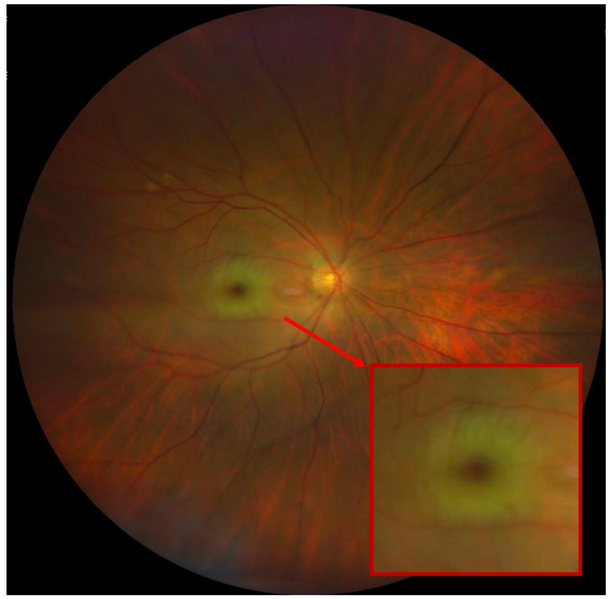

Figure 1. Color fundus photography. Diffuse retinal whitening with a cherry red spot in the right eye.

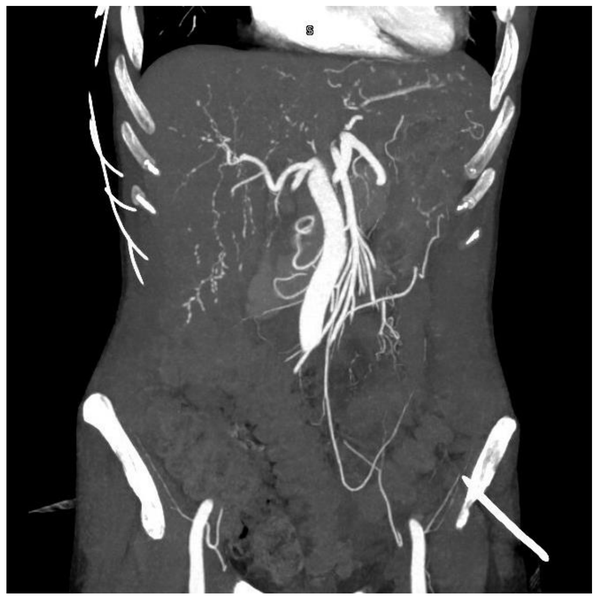

Figure 3. CT angiogram of the abdomen. Microaneurysms scattered near the periphery of the liver.

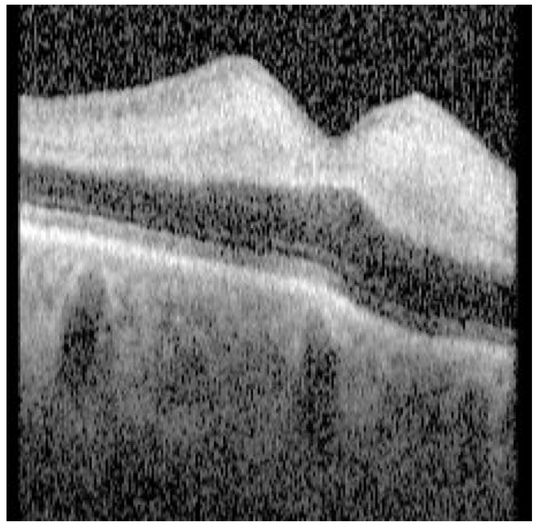

Figure 2. Optical coherence tomography scan of macula. Diffuse inner retinal thickening and hyperreflectivity in the right eye.

Patient's Lab Values	Reference Range & Units
Albumin **3.4 (L)**	3.5 – 5.2 g/dL
Alkaline Phosphatase **354 (H)**	35 – 104 U/L
AST **79 (H)**	<32 U/L
ALT **139 (H)**	<33 U/L
ESR **66 (H)**	<30 mm/hr
Rheumatoid factor **46 (H)**	<14 IU/ml
CRP **68.6 (H)**	<8.0 mg/L
SSA autoantibody **143 (H)**	0 – 99 [AU]/mL
ANA speckled pattern **80 (H)**	<80 1/dil
WBC **10.2 (H)**	4 – 10 x10^3/uL
Absolute Eosinophil **0.70 (H)**	0 – 0.5 x10^3/uL
Anti-Myeloperoxidase Ab <5.0	<20.0 CU
Anti-Proteinase 3 Ab <5.0	<20.0 CU

Table 1. Patient's laboratory values.

DISCUSSION

- Although the patient reported headaches and jaw claudication, an absence of granulomatous inflammation in her temporal artery biopsy ruled out giant cell arteritis.
- Negative anti-neutrophil cytoplasmic antibody (ANCA) ruled out eosinophilic granulomatosis with polyangiitis (p-ANCA), granulomatosis with polyangiitis (c-ANCA), and microscopic polyangiitis (p-ANCA).
- Although not specific for PAN, elevations in erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) indicate systemic inflammation associated with PAN.
- The patient's complaints of neuropathy in feet, headaches, weight loss, and abdominal discomfort align with common symptoms of PAN.
- CT evidence of microaneurysms (string of pearls appearance) involving the hepatic arteries supports the diagnosis of PAN.
- During her hospitalization, the patient received seven doses of cyclophosphamide along with methylprednisolone which improved her vision (to hand motion) and sensation.
- Currently, the patient is on prednisone 4mg daily and closely follows up with her rheumatologist. The patient's vision remains stable (counting fingers at 3 feet).

CONCLUSION

- We report an uncommon occurrence of CRAO associated with PAN.
- Early diagnosis and proper management of PAN are crucial for preventing irreversible complications such as vision loss from CRAO.

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