Chronic Granulomatous Panuveitis with ANCA Positive Testing

Kristina Shpountova, O.D.

Edward Hines Jr. VAH - Hines, IL 60141
Jesse Brown VAMC - Chicago, IL 60612

3241 South Michigan Avenue, Chicago, Illinois 60616

INTRODUCTION

ANCA-associated vasculitis (AAV) is a group of disorders optometrists do not often encounter, but knowing their clinical manifestations and diagnosis is critical for patient care. This case report outlines a rare ocular manifestation of AAV.

CLINICAL FINDINGS

Case history: 30 y/o AA male lost to follow-up with complaint of blurred vision and photophobia OU for multiple years

- **OcHx**
  1. Chronic bilateral granulomatous panuveitis OU
     -s/p sub-tenons Kenalog OS and methotrexate treatment
     -MPO+(PR3-) GPA w/o other systemic involvement
  2. Glaucoma suspect OU with h/o steroid response
  3. Pseudophakia OU; s/p YAG OS; PCO OD

- **MHx**
  1. Depression
  2. LBP, chronic knee pain
  3. Recent negative STD screen
  4. Medications: none

Exam	OD	OS
BCVA	20/25	20/25
Pupils/EOMs/CVF	NL	NL
IOP	15	15
L/L	NL	NL
Conjunctiva	NL	NL
Cornea	CE scar, Poor TF	CE scar, Poor TF
AC	1 isolated WBC, (-) flare	NL
Iris	Dyscoria, temp atrophy	Dyscoria
Lens	PCIOL; 2+ central PCO	PCIOL; s/p YAG OS
ONH	0.60v/0.55h	0.45v/0.45h
Macula	NL	NL
Vitreous	tr-1+ cell	+snowballs
Vessels	Early focal venous sheathing along ST/I arcades	Early focal venous sheathing along ST/I arcades
Periphery	CR scarring	CR scarring

FIGURE 1
Fundus Photo OD

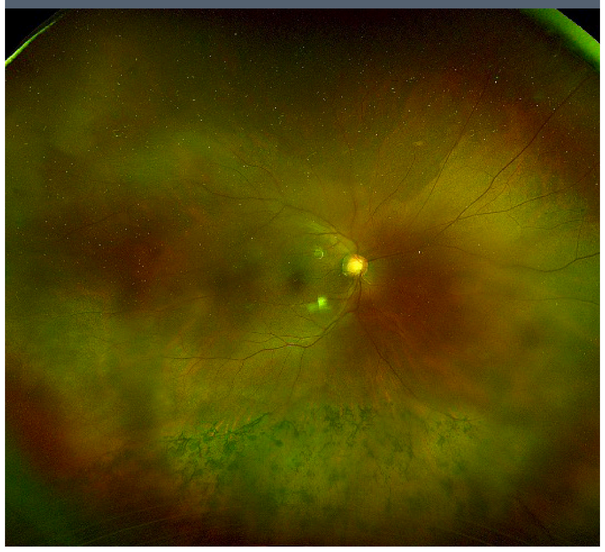
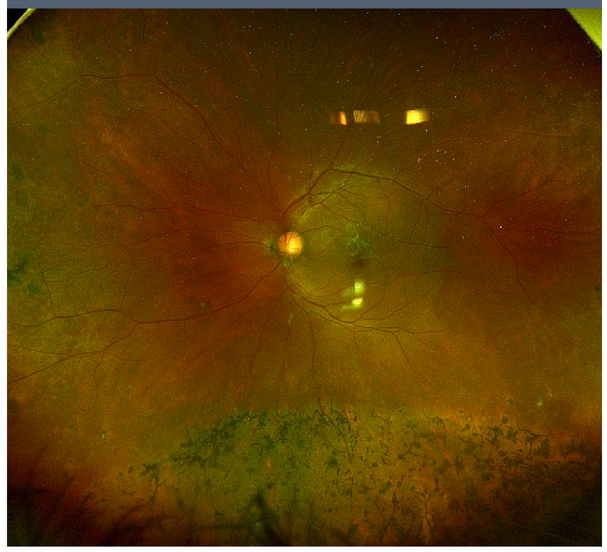
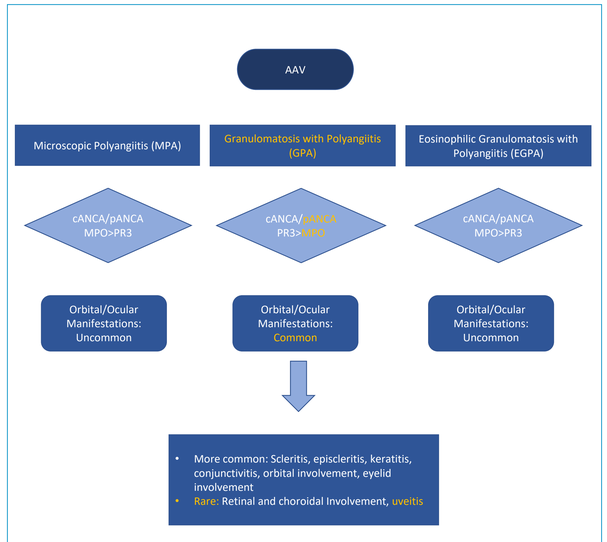


FIGURE 2
Fundus Photo OS



Imaging	OD	OS
ONH OCT	NL	NL
Macula OCT	NL	NL
24-2 HVF	Full	Full
Gonioscopy	No structures sup (+) PAS, CB T/I/N	No structures S/N (+) PAS, CB T/I
B-scan	Retina intact w/ vitreal debris	Retina intact w/ vitreal debris
FANG	No vasculitis	No vasculitis

Lab/Radiology Study	Result
ANA, RPR/MHATP, HLA-B27, Quant TB, Lyme screen	Negative
RF, ACE, Lysozyme, CRP, ESR	NL
cANCA	Negative
pANCA	Positive
PR3	Negtive
MPO	Positive
Chest X-ray, SI Joint X-ray, Spine X-ray	NL



DISCUSSION

- This patient was diagnosed with a recurrent panuveitis from MPO(+) GPA AAV.
- AAV is a group of rare autoimmune disorders that affect small blood vessels.
- ANCA or anti-neutrophil cytoplasm autoantibodies target proteins inside neutrophils and release toxic substances which damage blood vessel walls and cause inflammation.
- AAV can affect any part of the body but most commonly affects kidneys, lungs, joints, ears, nose, and nerves.
- Ocular complications can occur in every structure in the eye and orbit.
- Ocular disease is often the dominant presentation with GPA.
- Granulomatous pan-uveitis is a rare yet documented ocular manifestation of GPA, occurring in up to 10% of patients, and is extremely rare amongst African-Americans.
- This case shows how ANCA testing is valuable with uveitis when other uveitic testing is normal.

MANAGEMENT

- There is no cure for AAV.
- With ocular manifestations of GPA, treatment is aimed at the systemic disorder itself.
- Treatment options include glucocorticoids, immune suppression, and maintenance therapies.
- This patient was referred to rheumatology for continued systemic treatment.

CONCLUSION

- AAV is rare and life threatening as it affects vital organs.
- Early detection and treatment are key to prevent long-term complications.
- Co-management is essential for patients to receive appropriate therapy for best possible visual and systemic outcome.

BIBLIOGRAPHY

Available upon request

CONTACT

Kristina Shpountova, O.D.
kshpountova@eyedoc.ico.edu • www.ico.edu