Giant Cell Arteritis (GCA)

Authors: Michelle Nguyen, MD and Sonam Kiwalkar, MD

Definition: GCA (formerly temporal arteritis) is a large vessel vasculitis, usually involving the aorta & other great vessels.

Epidemiology:

- Most common vasculitis in adults.
- Race: High incidence in Whites.
- Age: Almost never below the age of 50 years. Mean age 76.7 years.
- Sex: female to male ratio = 2 to 3 : 1.

Symptoms:

- Headache ~ 60-70%. Usually temporal but also frontal or occipital with variable severity. Good response to steroids.
- Constitutional symptoms ~ 50%
    - Fever of unknown origin, weight loss, anorexia
- Jaw and tongue claudication ~ 50%
    - Dull pain with chewing, which improves after rest
- Polymyalgia rheumatica (PMR)[2] ~ 40-50%
- Vision Symptoms ~ 15%. Presents as partial or complete loss of vision, blurry vision, diplopia or amaurosis fugax.
    - If untreated, the second eye is likely to become affected within 1-2 weeks
    - Anterior ischemic optic neuropathy (AION) is the most common pattern characterized by white pallor and edema of optic disc along with flame hemorrhages adjacent to the discs
- Arm claudication ~ 4-15%

Physical Exam:

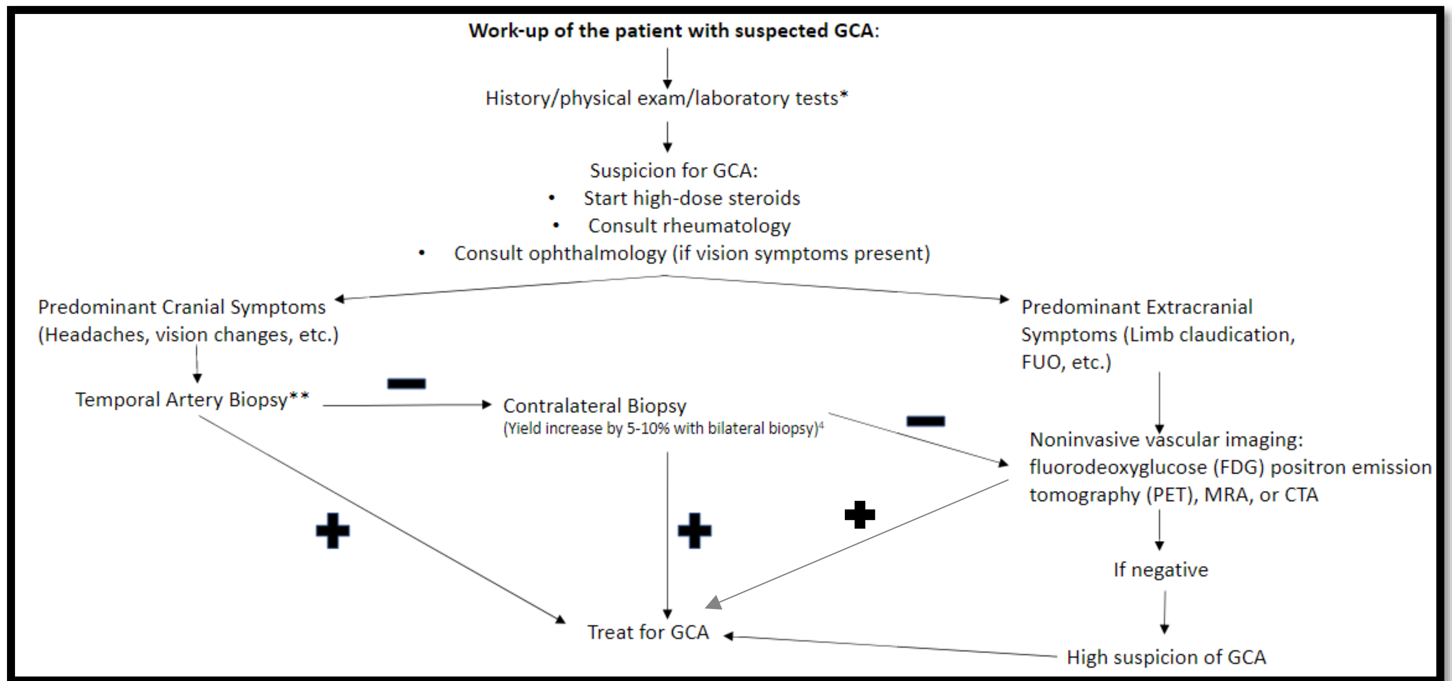
Vitals	Fevers
HEENT	Scalp tenderness Tender, beaded, or pulseless temporal artery
Cardiovascular	Carotid, axillary, or brachial bruits indicating large vessel involvement
BP asymmetry	> 10 mmHg systolic between arms indicating large vessel involvement
Extremities	Asymmetric Pulses

Diagnostic Investigations for GCA:

1) Laboratory Tests*:
- CBC (anemia of chronic disease, thrombocytosis)
- CMP (low albumin, elevated alkaline phosphatase)
- Typically, high inflammatory markers: ESR > 50 mm/hr and CRP > 10 mg/L

2) Temporal Artery (TA) Biopsy**[5,6]:
- Gold standard
- Should biopsy > 1 cm section within 2 weeks of starting steroids.

3) Cranial Doppler Ultrasound (CDUS):
- Operator dependent. Less experience in U.S. 2021 American College of Rheumatology guidelines *conditionally* recommend TA Biopsy over CDUS[3]
- Should perform within days of starting steroids.
- Findings include halo sign (homogenous wall thickening of artery); 68% sensitive and 91% specific[7]



Treatment:[3]

- Long taper of prednisone (1-3 years) along with glucocorticoid sparing agent (preferably tocilizumab over methotrexate[8]). Initial dose for patients without visual loss at presentation: prednisone 1 mg/kg or equivalent, not exceeding 60 mg a day. Initial dose for patients with threatened or established visual loss at presentation: methylprednisolone 500 to 1000 mg intravenous daily, for three days.
- Side effects: Increased risk of infection, high blood sugar, high blood pressure, thinning of the skin, easy bruising, tendon rupture, avascular necrosis, anxiety, insomnia, weight gain, osteoporosis, etc.
- Adjuvant therapies: Aspirin: non-randomized data showed reduction in vision loss and strokes. Statin & PJP prophylaxis: poor data, no concrete recommendations. Screen and treat for glucocorticoid induced osteoporosis.

Clinical Pearls:

- GCA can present in patients > 50 years of age with new onset headache, constitutional symptoms, jaw claudication, vision changes (diplopia, blurred vision, amaurosis fugax, total or partial loss of vision), etc.
- Signs suspicious for GCA include scalp tenderness and tenderness on palpation of temporal artery.
- If suspicious of GCA, get ESR & CRP and start high dose steroids 1mg/kg, not exceeding 60mg/day. If vision is threatened, start IV methylprednisolone 500mg-1000mg/day x 3 days. Refer patient for temporal artery biopsy.
- Urgent consult for rheumatology and ophthalmology (if presenting with ophthalmic symptoms).

References:

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