Giant Cell Arteritis:  
Arthritis That Is Really In Your Head

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Terminology

Giant Cell Arteritis  GCA  
Polymyalgia rheumatica  PMR  
Vasculitis (inflammation of blood vessels)

GCA and PMR  
COMMON DISEASES, CLOSELY RELATED  
- (PMR) is second only to Rheumatoid arthritis in lifetime risk of a rheumatic disease.  
- PMR more common in > 50 yo than rheumatoid arthritis  
- Giant cell Arteritis (GCA) / temporal arteritis is by far the most common vasculitis  
- Occur in the same patient population  
  - 15% of PMR patients have GCA  
  - 40% of GCA patients have PMR  
- GCA and PMR Season?
GCA Epidemiology: All ≥ 50 yo

- Incidence of GCA depends on population
  - Highest: 33 per 100,000/year, > 50 years old Scandinavians and Americans of Scandinavian descent
  - Lowest: 0.1 per 100,000/year, > 50 years old Japanese, Asian/Indian, African Americans
- Age
  - 1.5 cases per 100,000 in the 60's
  - 20.7 cases per 100,000 in the 70's
  - 2:1

PMR Epidemiology: All ≥ 50

- Same patient population as GCA
  - High risk: Scandinavians and other people of Northern European descent
  - 3 times the incidence of GCA
  - 20 to 90 per 100,000 persons over the age of 50
  - 10 per 100,000 Italians

Giant Cell Arteritis
Clinical Presentation: All > 50yo

- Tender, indurated Temporal arteries
- New onset temporal (occipital, periorbital) headache (70%), can interfere with sleep
- Scalp soreness
- Jaw (50%) and Tongue (rare) claudication
- Sudden painless visual loss (15%)
- Cough
- Extremity Claudication/ cramp with use (10%)
- Stroke
- Fever / anorexia (15% of FUO in > 65yo)
- PMR (40%)
GCA Sudden Blindness

- Early Diagnosis of GCA is CRITICAL to preventing visual loss (15%)
- Once corticosteroids are started the risk falls to 1%
- Amaurosis Fugax (30.6%)
- Diplopia (5%)
- Visual loss usually irreversible
- Very rapid high dose IV steroids
- Role of low dose ASA


Polymyalgia Rheumatica

Clinical Presentation (all > 50 yo)

- Acute - Subacute onset (1 day-several weeks)
- Morning Stiffness > 60 minutes
- Pain: Shoulders (95%) > Neck (70%) > Hips/thighs (50%)
- No muscle tenderness to touch
- SYMMETRICAL !!!
- Night pain
- Arthritis (knees, wrists, MCPs) transient
- Extremity edema- tenosynovitis (25%)
- Subjective weakness
- Constitutional symptoms (40%)
  - Malaise, fatigue, depression, anorexia/weight loss, fever

PMR: Consider It A Symptom First!

Differential Diagnosis

- Giant Cell Arteritis
- Early Rheumatoid Arthritis
- RS3PE (remitting seronegative symmetrical synovitis with pitting edema)
- Polyarticular CPPD arthritis
- Spondylitis
- Systemic Lupus (particularly in elderly)
- Myositis
- Statin associated myalgia
- Hypothyroidism
- Depression
- Malignancy / Metastasis /MM
- Parkinson’s disease
- Hyperparathyroidism
- Fibromyalgia
- Osteomalacia
Clinical Spectrum of GCA/PMR

- Headache
- Visual change
- Jaw pain
- Stiffness
- Prolonged morning stiffness
- Neck, shoulders, hips
- Cramping of extremities with use (claudication)
- Wasting syndrome
- Fever
- Weakness
- Depression
- Elevated ESR, CRP

Large Vessel GCA: 10% of all GCA

- Aortitis
  - Extremely common by PET scan
  - Pulseless disease
  - Extremity cramping with use
  - BP asymmetry
  - New Raynaud’s phenomenon
  - Fingertip gangrene
  - Thoracic Aortic aneurysm
  - 17 times more common late

PET scan/MRI composite
**GCA: Angiogram**

10% With Large Vessel Involvement

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**Diagnosis Of GCA**

Characteristic Clinical Presentation +

- Thickened, tender temporal arteries, bruits
- + temporal artery biopsy
- Currently the gold standard for dx GCA
- Asymmetric pulses
- Angiogram / HRCT scanning
- **Elevated ESR and/or CRP**
  - Anemia, thrombocytosis
  - Aortitis by PET scan
  - PMR (40%)

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**Why Biopsy?**

< 10% chance of GCA with an appropriate TA bx

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**OUTCOME IN 88 PATIENTS WITH NEGATIVE TAB**

Follow-up, median (range): 56 mo (1-192)

- Miscellaneous: 6%
- Connective tissue diseases: 10%
- Infection: 6%
- Giant cell arteritis: 10%
- Polymyalgia rheumatica: 38%
- Headache: 10%
- ASO/stroke: 10%

Hall, S. Lancet, 1988
GCA: Why Biopsy?
- Alternative Diagnoses
  - < 10% chance of GCA with an appropriate TA bx
- Commitment to Steroids for 9-18 mo is justified
- Biopsy Controversies?
  - Length (> 2 cm)
  - Bilateral vs unilateral (Unilateral)
  - Complications of the biopsy
  - Ultrasound TOO operator dependent?

HALO SIGN IS Very Specific

REMEMBER
- Negative TA biopsy does NOT rule out large vessel GCA
- OPMR suspects must be carefully evaluated for GCA (the older at dx of PMR, the more likely they have GCA)

Diagnosis of PMR
- Abrupt / Subacute Onset of SYMMETRIC Stiffness and pain (2 out of 3 areas)
  - Neck
  - Shoulder muscle girdle
  - Hip muscle girdle
- Morning stiffness > 1 hour
- Gelling to the MAX!!
- AGE ≥ 50
- Elevated ESR and / or CRP
  - Negative ANA, RF, CCP
- 100% Better after 5 days of prednisone 20 mg/day
Diagnosing PMR

Subclinical Synovitis

- Joint scan
- MRI
  - Biceps tendonitis
  - Glenohumeral synovitis
  - Bursitis (thickened, edema)
- U/S
  - Fluid filled subacromial bursa

Diagnosis of PMR- PET Scanning

GCA treatment

- Non-specific vascular maintenance
  - Low dose Aspirin
  - Lipid management (ATP 4 guidelines) × 2
  - Stop smoking
  - Control blood pressure and diabetes
- Prednisone 1 mg/ kg (unusual for > 60 mg / day)
- Protect the bones
  - Everyone should get a bisphosphonate
  - Calcium and Vitamin D
GCA Treatment: Prednisone

- 10 days to get the biopsy after starting prednisone.
- Prednisone and reduced risk of blindness.
- < 1% risk after ANY dose of prednisone.
- Continue initial dose of prednisone UNTIL ESR and/or CRP are back to normal.
- 1/3 relapse and the clock starts over.
- Relapse of PMR sx only, DO NOT increase prednisone, just stop taper temporarily for no less than 4 weeks.

GCA Treatment: Prednisone Sparing RX

- Low dose prednisone options? NO!!
- Methotrexate
- Tocilizumab / Actemra
  - 7 patients
  - Pretreatment steroid dose 21 mg, after treatment 4 mg
  - 1 pt autopsy, after fatal MI, with active vasculitis
- NOTHING ELSE
  - Not anti-TNFs, Not Ocrecia, Not Rituximab, Not Mycophenolate.

PMR Treatment

- Treatment with Prednisone 20 mg/day or less.
- QOD steroid does not work in > 50%.
- Protect the bones: bisphosphonate +Ca + Vit D.
- Rare patients can be managed with NSAIDs.
- Start to taper ONLY when inflammatory parameters controlled / symptoms gone.
- Too rapid a taper and the clock starts over again.
- Those that cannot taper below 10 mg/day have a different disease.
  - 13% develop seronegative inflammatory polyarthritis.
  - 15% develop GCA.
- Steroid sparing therapy only modestly successful.
  - HCQ
  - MTX.

**PMR Prednisone Taper**

- Initial dose ≤ 20 mg /day
- Continue that dose for a minimum of 14 days
- Reduce initially by 2.5 mg every 14-21 days
- Doses < 10 mg, should be reduced in 1 mg increments

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**GCA: Outcomes and Late Complications**

- Most go into remission
- 1/3 relapse in the first 10 years after dx
- Thoracic and abdominal Aortic aneurysms
  - 17 times, 3 times normal
  - Monitoring schemes?
    - I check a CXR every 3 years

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**PMR Outcomes**

- Most patients will enter remission, off all steroids
- > 50% are off all steroids by 18 months
- Average dose of prednisone by the 4th month should be < 7 mg/day
- 1/3 Relapse in the first 10 years after remission
- 13% with chronic inflammatory polyarthritis (sero- RA)
- 15% with GCA
Newer Entities

- Isolated aortitis (5% of all AAA repairs)
- IgG4 Disease and Lymphoplasmacytic aortitis

Summary

- PMR and GCA are complex diseases with potentially morbid treatments and rheumatologists should always be part of the management team
- New understanding of the relationship between these conditions is just around the corner
  - Infectious precipitants
  - Biochemical pathogenesis