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Case Report

- 68 years old white male with h/o recurrent sinusitis for many years
- h/o FESS for nasal polyps 1995 outside ENT
- Left maxillary tooth cyst removed 2001 by oral surgeon
- 2 months of right maxillary/periorbital pain/pressure, loss of smell, headache, nausea, vomiting (denies nasal obstruction, cough, vision change, diplopia, fever)
Case Report

- Underwent right concha bullosa mucocele resection/decompression and FESS without improvement in symptoms
- Neurologist treated maximally for migraine headache without success
Case Report

- Patient mentioned scalp tenderness
- ESR = 62
- CRP = 64.7
- Bilateral temporal artery biopsy positive for GCA
- Symptoms resolved on prednisone 60mg/day
- Referred to rheumatologist...
Giant Cell Arteritis (GCA)

- Most common type of systemic vasculitis affecting white patients over 50 years and incidence increases with age
- Characterized by granulomatous involvement of large and medium sized blood vessels of the aorta with predilection for the extracranial branches of the carotid artery
Demographics of GCA

- 2:1 female to male ratio (64% F vs 36% M)
- Mean age 70 years old
- White 74%, Black 18%, Hispanic 6%, Asian 1.6%
- 15-30 cases per year per 100,000 > 50 yrs age
- associated with HLA-DR4 possible genetic
Symptoms of GCA

- New headache (60%)
- Jaw/tongue/palate claudication (23%) \textit{pathognomonic, odds ratio}=9
- Visual symptoms/ocular findings (35%)
- Temporal artery exam finding (53%) such as TA beading, prominence, or enlargement
- Polymyalgia rheumatica (27%)
- Synovitis (12%), fatigue, fever, anorexia, fever, weight loss
- Scalp tenderness or necrosis
- Mean delay in diagnosis = 1.5 months

Polymyalgia Rheumatica

- pain & stiffness in proximal muscles (shoulders) worse in morning & after exertion
- elevated ESR
- responds rapidly to low dose prednisolone (10 mg/day)
- can occur alone or with GCA
American College of Rheumatology
1990 Criteria for Giant Cell Arteritis

A score of 3 or more has a sensitivity of 93.5% and a specificity of 91.2%.

1. age > 50 years at onset
2. new onset of localized headache
3. temporal artery tenderness or decreased pulse
4. ESR > 50
5. TA biopsy showing necrotizing arteritis

**Also scalp tenderness and claudication of jaw/tongue or on deglutition**

Labs for GCA

- Normocytic anemia (hgb < 12) 55%
- Leukocytosis (>11,000) 28%
- Elevated alkaline phosphatase 25%
- Low albumin (<3) 28%
- *Thrombocytosis (>400k) 49%
- *Mean ESR 93 (high ESR >20), 1-2% of GCA normal ESR
- *Mean CRP 94 (high CRP >2.45)

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Odds Ratio of Positive Biopsy

- 1.5x greater with ESR 47-107
- 4.2x greater with platelets > 400,000
- 5.3x greater with CRP > 2.45.

ESR & CRP in GCA

- multi-center review 119 patients TAB positive
- Sensitivity of ESR & CRP together was 99%
- Both are elevated in most patients (93.4%)
- normal ESR with elevated CRP (1.6%)
- elevated ESR with normal CRP (1.7% - 3.7%)
- 1% of GCA had both normal ESR & CRP

Pathologic Findings of Temporal Artery Biopsy

- Typical temporal arteritis (49%)
  - at least one giant cell with mixed mononuclear cells (lymphocytes, histiocytes, and plasma cells)

- Atypical temporal arteritis (51%)
  - inflammation without giant cells or
  - inflammation mainly in the adventitia (rather than media)
Biopsy of Temporal Artery

- TAB is gold standard for diagnosis of GCA
- Sensitivity of TAB = 87%
- 15% of GCA will be biopsy negative

Unilateral vs. Bilateral Biopsy?

Unilateral *if the specimen length and processing are adequate  

**TWO STUDIES:**

- **1. concordance rate of two sides = 99%**
  

- **2. concordance rate of two sides = 97%**
  
Surgeon’s Intraop Findings

- Thick artery
- Nodular & tortuous artery
- Pale artery
- Minimal bleeding/back flow
- Occluded lumen

4-5 considered grossly positive, 2-3 indeterminate, <2 negative.

Specificity 97.9%; accuracy 98.2%. (retrospective study of 108 patients)
Shrinkage
(of temporal artery biopsy specimen)

- Mean shrinkage of specimen 15%
- Biopsy should be at least 2.5 cm length
- Proper meticulous sectioning of specimen by pathologist is required
Does Previous Steroid Treatment Affect Biopsy Findings?

- Mayo study 535 patients
- Biopsy shows arteritis even after more than 14 days of steroids!
- Untreated group had biopsy positive rate of 31%
- Treated group had biopsy positive rate of 35%
- Trend towards atypical path with higher dose/longer duration of steroid therapy but arteritis was still detectable.


Vision loss in GCA

- Risk factors for visual loss in giant cell (temporal) arteritis: a prospective study of 174 biopsy proven patients.

- Transient ischemic visual symptoms 28% with permanent vision loss in 13%.

- Two risk factors for permanent visual loss
  - 1. h/o transient visual ischemic symptoms (odds ratio 6.3)
  - 2. higher platelet count > 400,000 (odds ratio 3.7)

Vision loss in GCA

- Visual recovery is uncommon
- Visual deterioration can occur despite high dose (250mg solumedrol q 6 h) IV steroids (3%) greatest risk is in the first 6 days
- Pale swollen optic disc with flame-shaped hemorrhages, cupping of optic disc
- Loss of vision, visual field defects, complex visual hallucinations, loss of color vision, ptosis, diplopia, tonic pupils.

Vision loss in GCA

- 185 patient retrospective study
- 41 (22%) with vision loss
  - 46% unilateral, 37% sequential, 17% simultaneous
- Sequential eye involvement was only seen with oral steroid treatment (not IV)
- In patients with vision loss (treated with IV)
  - 34% improved, 49% unchanged, 17% worsened
  - acuity (15%) may improve without better visual fields (5%)

Treatment of GCA

- Oral prednisone 1 mg/kg/day
- Low dose ASA with steroids can reduce incidence of CVA & vision loss
- Methotrexate can be used if steroid-resistant or steroids contraindicated
  (studies on efficacy of methotrexate are mixed)

Top 10 Take Home Points for GCA

- 10. Check ESR, CRP, CBC, CMP
- 9. Normal ESR & CRP do not rule out GCA (1%)
- 8. TAB has specificity and PPV of 100%, but sensitivity of 85%
- 7. Biopsy is ok within 2 weeks of starting steroids
- 6. Unilateral biopsy of 2.5cm length is sufficient, but proper processing of biopsy specimen is important
Top 10 Take Home Points for GCA

- 5. GCA patients will have negative biopsy 15% of the time
- 4. 20% of GCA cases have loss of vision and may present without other symptoms of arteritis
- 3. Outpatient treatment is prednisone 1mg/kg/day and daily 81mg aspirin
- 2. Need inpatient high dose IV steroids for any transient visual symptom or platelets > 400k
- 1. Visual symptoms are severe, and vary visual acuity, visual fields, ophthalmoplegia, color blindness, ptosis.