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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)

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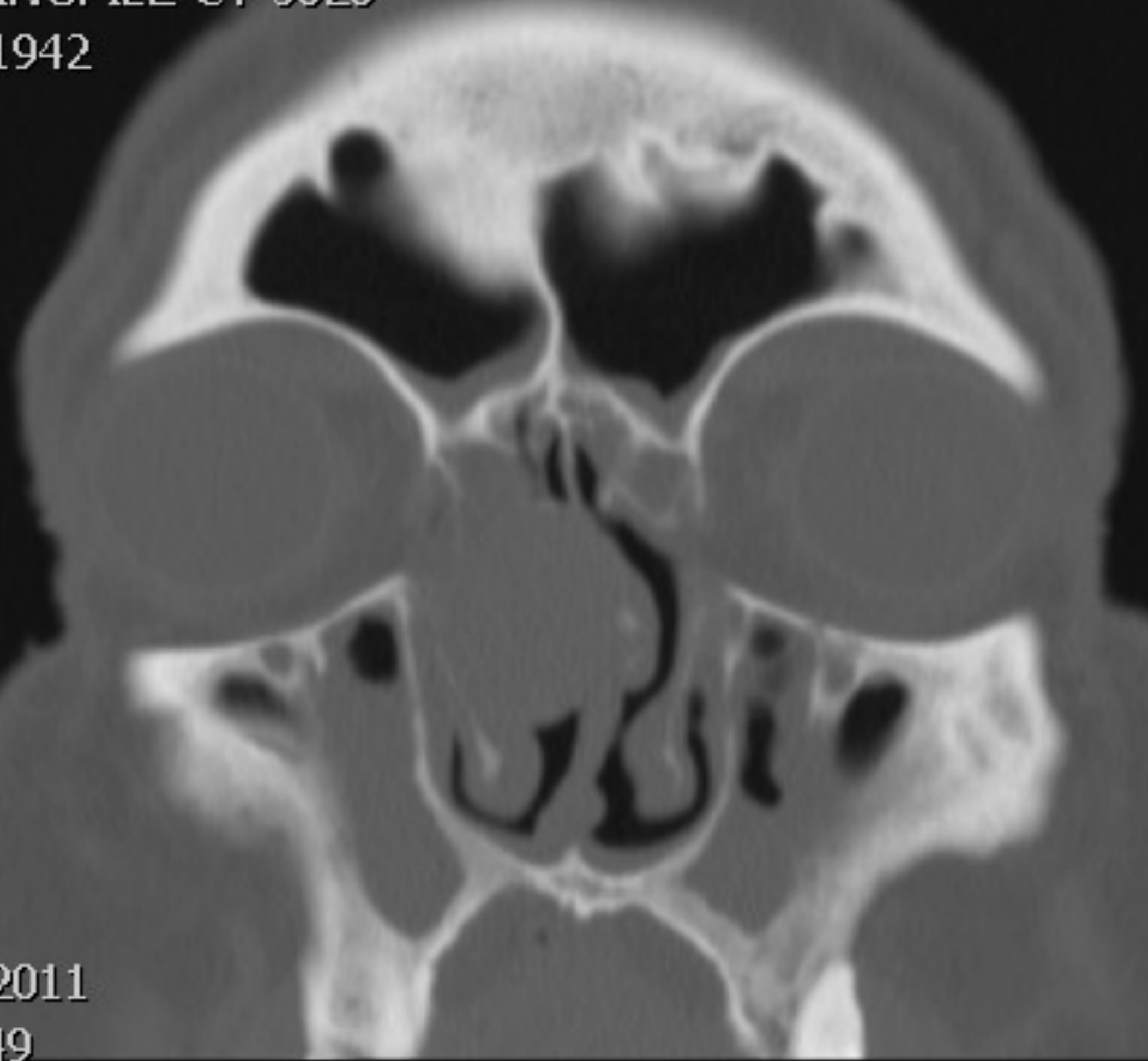
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# Case Report

- ◆ 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%) 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

Myklebust G et al. (1996) A prospective study of 287 patients with polymyalgia rheumatic and temporal arteritis: clinical and laboratory manifestations at onset of disease and at the time of diagnosis. Br J Rheumatol 35: 1161-8.



# 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)

## 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

1. Parikh M et al (2006) Prevalence of a normal C reactive protein with an elevated erythrocyte sedimentation rate in biopsy-proven giant cell arteritis. *Ophthalmology* 113 (10): 1842-5

2. Poole TR et al (2003) Giant cell arteritis with a normal ESR and CRP. *Eye* 17(1): 92-3.

3. Myklebust G et al. (1996) A prospective study of 287 patients with polymyalgia rheumatic and temporal arteritis: clinical and laboratory manifestations at onset of disease and at the time of diagnosis. *Br J Rheumatol* 35: 1161-8.

# 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%

Danesh-Meyer H et al. Low diagnostic yield with second biopsies in suspected giant cell arteritis. J Neuroophthalmol (2000), 20(3):213-5.

◆ 2. concordance rate of two sides = 97%

Boyev LR et al. Efficacy of Unilateral Versus Bilateral Temporal Artery Biopsies for the Diagnosis of Giant Cell Arteritis. Am J Ophthalmol (1999), 128(2):211-215.



# 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.

Achkar AA et al. How Does Previous Corticosteroid Treatment Affect the Biopsy Findings in Giant Cell (Temporal) Arteritis? Ann Intern Med 1994; 120: 987-992

Narvaez J et al (2007) Influence of previous corticosteroid therapy on temporal artery biopsy yield in giant cell arteritis. Semin Arthritis Rheum 37(1): 13-19.

# 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%)

Visual Morbidity in Giant Cell Arteritis. Ophthalmology, November 1994, Vol 101(11), 1779-1785.

Poor Prognosis of Visual Outcome after Visual Loss from Giant Cell Arteritis. Ophthalmology, June 2005, Vol 112(6), 1098-1103.

# 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)

Nesher G et al. (2004) Low-dose aspirin and prevention of cranial ischemic complications in giant cell arteritis. *Arthritis Rheum* 50(4): 1332-7.

# 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 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 varies visual acuity, visual fields, ophthalmoplegia, color blindness, ptosis.