A CRYSTAL CLEAR MOMENT

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67 YEAR OLD MAN PRESENTS TO ED

ACUTE ON CHRONIC STABBING NECK PAIN
SUBJECTIVE FEVERS
NEW RIGHT JAW PAIN
WORSENING RIGHT KNEE PAIN
### OTHER PERTINENT HISTORY

**REVIEW OF SYSTEMS**
- No headaches
- No vision loss
- No photophobia
- No rashes
- No neurological deficits

**SOCIAL HISTORY**
- Remote history of tobacco and alcohol use
- No illicit drug use

**PAST MEDICAL HISTORY**
- Atrial fibrillation on warfarin
- Oligoarthritis
  - Recent presumed acute CPPD flare, no tap
- Seizure disorder, well controlled

**FAMILY HISTORY**
- Non-contributory
# PHYSICAL EXAM

<table>
<thead>
<tr>
<th>VITALS</th>
<th>EXAM</th>
</tr>
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<tbody>
<tr>
<td>T</td>
<td>36.1 °C</td>
</tr>
<tr>
<td>P</td>
<td>80</td>
</tr>
<tr>
<td>BP</td>
<td>127/85</td>
</tr>
<tr>
<td>RR</td>
<td>17</td>
</tr>
<tr>
<td>SpO₂</td>
<td>98% on room air</td>
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<table>
<thead>
<tr>
<th>EXAM</th>
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<tbody>
<tr>
<td>HEENT</td>
</tr>
<tr>
<td>NECK</td>
</tr>
<tr>
<td>MUSCULO-SKELETAL</td>
</tr>
<tr>
<td>NEURO</td>
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LABORATORY WORKUP

- LFTs: WITHIN NORMAL LIMITS
- LACTIC ACID: 1.17
- TSH: 3.31
- INR: 2.8

- CALCIUM: 9.1
- PHOSPHATE: 2.6
- BLOOD CULTURES X 2 SETS: PENDING
## IN THE ED, THE CLINICAL PICTURE EVOLVES

### TEMPERATURE

<table>
<thead>
<tr>
<th>Temperature</th>
<th>Value</th>
<th>Reference</th>
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<tbody>
<tr>
<td>36.1 °C</td>
<td>39.1 °C</td>
<td>≤ 3.0</td>
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</tbody>
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### CRP

- 123
- (ref ≤ 3.0)

### ESR

- 30
- (ref 0 -15)
An older man with history of atrial fibrillation on warfarin, arthritis, and chronic neck pain who presents with **acute neck and jaw pain, fever, and** is found to have elevated **inflammatory markers.**
## Problem Statement to Differential

<table>
<thead>
<tr>
<th>Infectious</th>
<th>Inflammatory</th>
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<tbody>
<tr>
<td>meningitis</td>
<td>giant cell arteritis</td>
</tr>
<tr>
<td>epidural abscess</td>
<td>polymyalgia rheumatica</td>
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<tr>
<td>osteomyelitis/discitis</td>
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</table>
WE PERFORMED A CT OF THE NECK

RESULTS INTERPRETED AS BENIGN

- DJD at C5-C7
- Radiology concerned about appearance of meninges
- Subsequent MRI normal
HOW DO OUR FINDINGS FIT WITHIN OUR DIFFERENTIAL?
WE LOOKED BACK TO 2008 FOR KEY EVIDENCE

Chondrocalcinosis of bilateral knee joints
An older man with history of atrial fibrillation on warfarin, arthritis, and chronic neck pain who presents with acute neck and jaw pain, fever, and is found to have elevated inflammatory markers.

An older man with history of arthritis, chronic neck pain, and chondrocalcinosis on imaging who presents with acute neck and jaw pain, fever, and is found to have elevated inflammatory markers and an inflamed right knee.
ARTHROCENTESIS

- No organisms
- WBC 14,700
- Positively birefringent crystals

A QUICK REFRESHER ON CPPD

- Usually causes monoarthritis
- Most commonly affects peripheral joints
- Increased risk with age and joint disease
BUT WHY DOES HIS NECK HURT?

NEXT DIAGNOSTIC STEP

LUMBAR PUNCTURE? | PET SCAN? | SEROLOGIC MARKER? | OTHER LAB TEST?
CT NECK WAS RE-REVIEWED

Dens

Transverse Atlantal Ligament
An older man with history of arthritis, chronic neck pain, and chondrocalcinosis on imaging who presents with acute neck and jaw pain, found to have elevated inflammatory markers, CPPD of right knee, and C1-C2 microcrystalline deposition on CT of the neck.
DIAGNOSIS
CROWNED DENS SYNDROME
CROWNED DENS SYNDROME

- Atypical presentation of CPPD
- Described by Bouvet et al., in 1985
- In patients with diagnosis of acute CPPD, 18.3% had neck symptoms
CROWNED DENS SYNDROME (CDS)

- **CLINICAL PRESENTATION**
  - Cervico-occipital stiffness
  - Evidence of inflammation

- **CT SCAN OF NECK IS BEST IMAGING MODALITY**
  - Identification of calcifications of the retro-odontoid ligament the neck

- **ULTIMATELY, SYMPTOM RESOLUTION WITH NSAID/COLCHICINE**

- **FREQUENT REPORTS OF MISDIAGNOSIS AS PMR, GCA, AND MENINGITIS IN THE LITERATURE**
HOSPITAL COURSE AND FOLLOW UP

NAPROXEN

SYMPTOMS IMPROVED OVERNIGHT, DISCHARGED NEXT DAY

SWITCHED TO PREDNISONE TAPER GIVEN INCREASED RISK OF BLEEDING
CROWNED DENS SYNDROME IS AN UNDER-RECOGNIZED CAUSE OF NECK PAIN

CPPD FREQUENTLY PRESENTS ATYPICALLY

REFORMULATION OF THE PROBLEM REPRESENTATION CAN LEAD TO ALTERNATIVE DIAGNOSES
REFERENCES


