Not Another ACS Rule Out

Ryan Nelson, MD
Stephanie Preston, BE
Melissa Watts, MD

Tulane University Health Sciences Center
Department of Internal Medicine
New Orleans, LA
History of Present Illness

- A 50 year old African-American woman presents with chest pain.
  - Acute onset
  - Occurs at rest
  - Sub-sternal, tightness, no radiation
  - Associated with shortness of breath
  - Several episodes in past few weeks
Review of Systems

- 30-pound unintentional weight loss
- Progressive dysphagia
- Dysarthria
- Dry cough
- L-sided facial droop
Medical History:
- Type II Diabetes, HTN, HLD, Bell’s Palsy

Surgical History:
- Tubal Ligation, Right Meniscus Repair

Social History:
- Denies smoking, illicit drugs. 1 drink/month
- NKDA

Home Medications:
- ASA, Ferrous Sulfate, Lisinopril/HCTZ, Metformin, Pravastatin, Naproxen
Physical Exam

- **VITALS:** 99.2°F, 89, 147/80, 18, 100% RA
- **GEN:** NAD, AAO x 3
- **HEENT:** MMM, oropharynx clear
- **NECK:** No cervical LAD, no JVD
- **CV:** Regular rate, normal S1/2, no murmurs
Physical Exam

- **PULM**: CTAB, good effort
- **ABD**: soft, ND,+BS, no TTP
- **NEURO**: L-sided facial droop, no furrowing of ipsilateral brow
- **EXT**: No edema, 2+ DPs, no clubbing
Initial Diagnostic Tests

CBC, CMP, PT, INR, aPTT: normal

Troponin: 0.08 > 0.10 > 0.11 > 0.10

Lipid Panel:
- A1c - 7.2%
- Chol - 220
- TSH - 1.46
- TG - 175
- LDL - 138
- HDL - 47
• NSR, normal axis, no ST-segment changes, T-wave inversions in leads I, aVL, V2, V5, V6
Chest X-Ray
Initial Management

NSTEMI:

- TIMI = 4
- ASCVD Risk = 18.3%
- Heparin gtt
- Aspirin 325 x 1 -> 81 daily, Plavix 300 x 1 -> 75 mg daily, Lisinopril 20 daily, Rosuvastatin 40 daily, Carvedilol 3.125 BID
Initial Management

Dysphagia:
- Modified Barium Swallow Study - failed

Bell’s Palsy:
- Prednisolone 60 mg daily
- Plan to taper after 5 days
Additional Diagnostic Tests

ESR: 108, CRP: 3.50, RF: <10
ANA: negative, Anti-ds DNA: negative
HIV 1 & 2 Ab: NR
c-ANCA, p-ANCA: <1:20 (negative)
Lyme IgG/IgM Ab: <0.91 (negative)
ACE Level: <14 (normal)
Total IgG: 1,026 (normal)
Transthoracic Echo

- Mild LV systolic dysfunction; EF = 40 - 45%
- Severe antero-septal hypokinesis
- Akinesis of the LV apex
Left Heart Catheterization

LM: No disease
LAD: 50% mid stenosis, 70% apical stenosis
LCx: 40% stenosis at level of OM2
RI: mid 65% stenosis
RCA: proximal 95% stenosis, mid 70% stenosis
Cardiac MRI

- Patchy mid-myocardial and sub-epicardial hyper-intensity in the inferior wall, lateral wall, and septum
- No acute infarct
• Focal enhancement and thickening of the dura mater, consistent with pachymeningitis
Bronchoalveolar Lavage

- Sub-carinal LN FNA: Two small epithelioid granulomas
- Flow Cytometry; Anaerobic, Aerobic, and Fungal Cultures; and AFB Smear - negative
Final Diagnosis

Multi-Organ Sarcoidosis, Initial Presentation

- Cardiac Sarcoidosis with NSTEMI
- Neurosarcoidosis with Cranial Nerve Palsy (VII)
- Pulmonary Sarcoidosis
- Gastrointestinal Sarcoidosis - Pharynx [Presumed]
Inpatient Course

• Prednisolone 60 mg daily course extended to 4 weeks with taper
• L-sided CN VII Palsy resolved
• Dysphagia resolved, passed repeat MBSS
• Repeat TTE:
  – EF > 55%, improved apical contraction
• Discharge HD #14
Outpatient Course

- Multi-disciplinary treatment team:
  - Rheumatology, Dermatology, Cardiology, Pulmonology, Neurology, and Primary Care
- Hydroxychloroquine 200 mg BID
- Methotrexate 10 mg weekly
- Prednisone tapered to 5 mg daily
Outpatient Course

- **Holter Monitor:** 1% PVC’s, otherwise NSR
- **EP Study:** no inducible ventricular arrhythmia
  - no indication for AICD
- **Staged PCI:** DES x 2 to the RCA - DAPT x 1 year
- **Patient doing well, no recent hospitalizations**
Sarcoidosis

- Peak incidence, ages 20-39
- More common in women
- Incidence 3 x higher in African-Americans
- Non-caseating granulomas:
  - Epithelioid cells, macrophages, lymphocytes
  - Later - fibroblasts and collagen
- CD4+ T-cell response maintains granulomas:
  - IL-2, IFN-γ, TNF-α
Sarcoidosis - Diagnosis

- Compatible clinical and radiographic findings
- Biopsy of most accessible involved organ
- Stain for AFB, Fungal and Bacterial Cultures
- ACE - poor sensitivity and specificity
- Diagnosis of exclusion
Cardiac Sarcoidosis

- 5% of patients symptomatic
- 25% of Sarcoidosis autopsies
- Restrictive cardiomyopathy
- Tachy- and brady-arrhythmias
- ACS very rare as initial presentation
CENTRAL ILLUSTRATION  Clinical Features of Cardiac Sarcoidosis

Small patches of basal involvement, usually clinically silent

Large area of septal involvement, often clinically manifest as heart block

Re-entrant circuit involving area of granuloma/fibrosis leading to VT

Extensive areas of LV and RV involvement, often clinically manifest as heart failure +/- heart block +/- VT

<table>
<thead>
<tr>
<th>Table 4 Heart Rhythm Society (HRS) consensus statement for diagnosis of cardiac sarcoidosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Histological diagnosis of cardiac sarcoidosis</strong></td>
</tr>
<tr>
<td>Endomyocardial biopsy specimens with non-caseating epithelioid granulomas and no alternative cause identified</td>
</tr>
<tr>
<td><strong>Clinical diagnosis of probable cardiac sarcoidosis</strong></td>
</tr>
<tr>
<td>Histologic diagnosis of extracardiac sarcoidosis and one or more of the following is present while reasonable alternative cardiac causes other than CS have been excluded:</td>
</tr>
<tr>
<td><strong>Corticosteroid or immunosuppressive therapy responsive cardiomyopathy or heart block</strong></td>
</tr>
<tr>
<td>Unexplained reduced LVEF (&lt;40%)</td>
</tr>
<tr>
<td>Mobitz type two second degree heart block or third degree heart block</td>
</tr>
<tr>
<td><strong>Depressed left ventricular ejection fraction &lt;50%</strong></td>
</tr>
<tr>
<td>Patchy uptake on cardiac FDG-PET in a pattern consistent with CS</td>
</tr>
<tr>
<td><strong>Late gadolinium enhancement (LGE) on cardiac magnetic resonance imaging in a pattern consistent with CS</strong></td>
</tr>
<tr>
<td>Positive gallium uptake in a pattern consistent with CS</td>
</tr>
</tbody>
</table>
Treatment

- Corticosteroids
  - Prednisone: 20 - 60 mg daily x 1-3 months then slow taper
- Immunomodulating Therapy:
  - Methotrexate: can decrease corticosteroid dose requirement
  - Hydroxychloroquine: for neurologic or cutaneous involvement
References


Special Thanks

Deepa Bhatnagar, MD
- Tulane Department of Internal Medicine

David Spruill, MD
- Tulane Department of Internal Medicine

David Smith, MD
- LSU Department of Radiology

Tracey Dewenter, MD
- LSU Department of Pathology
Questions?