A Case of Cardiac Sarcoidosis

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Our patient

- 33 year old woman
- Brother with sarcoidosis
- 4 years ago: salivary gland enlargement, bilateral hilar adenopathy on CXR
  - Presumed sarcoidosis: not biopsy proven
  - Prednisone x 1 month
- 2-3 weeks ago: admitted with DVT, bil PE
  - Incidentally noted 1st degree AV block on EKG
  - Incidentally noted to have diffuse thoracic/abdominal lymphadenopathy on CT
• Presents now with intermittent chest pain, worsening dyspnea/orthopnea
• Intermittent palpitations
• Noted to be bradycardic
Cardiac MRI

Cardiac MRI:
- sensitivity:
  75-100%
- Specificity:
  77-78%


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Advanced Imaging of Cardiac Sarcoïdosis

<table>
<thead>
<tr>
<th>Diagnosis group</th>
<th>Major criteria</th>
<th>Minor criteria</th>
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<tbody>
<tr>
<td>Histologic: CS is confirmed when endomyocardial biopsy specimen demonstrates noncaseating epithelioid cell granuloma with histologic or clinical diagnosis of extracardiac sarcoïdosis</td>
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<td>Clinical: although endomyocardial biopsy specimens do not demonstrate noncaseating epithelioid granuloma, extracardiac sarcoïdosis is diagnosed histologically or clinically and satisfies more than 2 major criteria, or 1 major criterion and 3 or more minor criteria</td>
<td>Advanced atrioventricular block</td>
<td>Abnormal electrocardiogram showing ventricular arrhythmias (multifocal or frequent ventricular contractions), complete right bundle branch block, axis deviation, or abnormal Q wave</td>
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<td>Basal thinning of interventricular septum</td>
<td>Abnormal echocardiography showing regional abnormal wall motion or morphologic abnormality (ventricular aneurysm, wall thickening)</td>
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<td>Positive cardiac 123I on uptake</td>
<td>99mTc myocardial scintigraphy showing perfusion defect</td>
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<td>Depressed left ventricular ejection fraction (&lt;50%)</td>
<td>MRI imaging showing delayed gadolinium enhancement</td>
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• Treated with:
  • Prednisone
  • Methotrexate
  • Biventricular internal cardio-defibrillator placed
    • Pacemaker – conduction disease
    • Defibrillator – primary prevention, reduced EF
  • Diuresis
  • Beta-blockers
  • ACE-inhibitors
• On follow up appears to be doing well clinically

Sarcoidosis

• Multisystem disorder of unknown etiology
• Noncaseating granulomas in tissue
  • Pulmonary, cutaneous, ocular, lymphatic, MSK, exocrine, endocrine, renal, neurologic, GI
• Accumulation of T lymphocytes, mononuclear phagocytes

Cardiac Sarcoidosis

- Prevalence varies geographically
  - Japan: up to 50-78% cardiac involvement\(^1\)
  - United States: 20-28% cardiac involvement\(^2\)
- Most commonly asymptomatic
  - Good prognosis, high remission rate, low mortality
- Symptomatic cardiac involvement drastically worsens prognosis\(^3\)
- Signs, symptoms depend on location of granulomas
  - Conduction abnormalities
  - Arrhythmias
  - Myocardial damage, simulating infarction
  - Congestive heart failure
  - Valvular disease
  - Sudden cardiac death

1. Sarcoidosis 1994; 11 (1) 26-31
2. Circulation 1978; 58 (6) 1204-121

Implications

- Cardiac sarcoidosis should be on the differential for young patients who present with new arrhythmias, conduction disease, ischemic symptoms, or heart failure
- Patients with extracardiac sarcoidosis should undergo evaluation for clandestine cardiac involvement\(^1\)
  - EKG

Pacemaker indications:
- High degree AV block

ICD indications:
- Reduced EF – primary prevention
- Ventricular arrest – secondary prevention
- Some experts believe all patients with cardiac sarcoidosis should undergo long-term rhythm monitoring
  - If frequent ventricular arrhythmias then can consider ICD even with preserved EF
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