The Heart in Sarcoidosis – Imaging

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Gemeinsame Jahrestagung 2016:
Schweizerische Gesellschaft für Kardiologie (SGK), Schweizerische Gesellschaft für Herz- und thorakale Gefässchirurgie (SGHC), Schweizerische Gesellschaft für Pneumologie (SGP)
Topics

- Cardiac sarcoidosis
- Whom to evaluate
- Imaging modalities in sarcoidosis
- Follow-up
Cardiac sarcoidosis

- In 5 % of pts with systemic sarcoidosis
- Subclinical cardiac sarcoidosis in up to 70 % (autopsies) of pts with systemic sarcoidosis
- Death due to cardiac sarcoidosis 13 – 85 %
- May be asymptomatic
Frequency of organ involvement

Clinical evaluation

Autopsy studies

Kusano et al. Heart 2016;102:184–190
Cardiac sarcoidosis clinical presentations

- Heart block and arrhythmias (ventricular)
- Sudden cardiac death (25 – 60 % of deaths)
- Congestive heart failure
- Miscellaneous: «simulated» infarction, cor pulmonale
Cardiac Sarcoidosis Diagnostic criteria

- Biopsy → noncaseating granuloma

- extracardiac sarcoidosis + major / minor criteria

  - major: av-block; basal thinning of interventricular septum; cardiac (gallium), FDG uptake or LGE in CMR; LVEF <50 %
Topics

- Cardiac sarcoidosis
- Whom to evaluate
- Imaging modalities in sarcoidosis
- Follow-up
Specific presentations **without** prior history of sarcoidosis

Screen with CMR and/or FDG PET if any of the following:

- Unexplained Mobitz II or 3rd degree AV block in adults aged <60 years
- Sustained Monomorphc VT of Unknown Etiology
Biopsy proven extra-cardiac sarcoidosis

Screen with CMR and/or FDG PET if any of the following is present

- Symptoms: → significant palpitations
  → pre-syncope / syncope
- Abnormal ECG
- Abnormal echocardiogram

Topics

- Cardiac sarcoidosis
- Whom to evaluate
- Imaging modalities in sarcoidosis
- Follow-up
Evaluation

- History
- ECG
- Echocardiography
- Cardiac magnetic resonance (CMR)
- Positron Emission tomography (PET)
40 year old female patient

- Recurrent dizziness during physical exertion
- Exercise induced RBBB
- Exercise induced syncope (once)
Holter ECG
Initial imaging evaluation algorithm

Suspected Cardiac Sarcoidosis

If contraindication to CMR

CMR

Positive / inconclusive

PET

(Cardiac and whole-body)

* If high clinical suspicion persists

Negative
CMR – cine images
CMR – late gadolinium images
Principle of „late enhancement“

Normal myocardium
Intact cell membrane

Acute infarction
Ruptured cell membrane

Scar
Collagen matrix

Correlation of CMR findings and pathologic findings
Summary of different “late enhancement” patterns

A. Subendocardial Infarct

- Idiopathic Dilated Cardiomyopathy
- Myocarditis

A. Mid-wall HE

- Hypertrophic Cardiomyopathy
- Right ventricular pressure overload (e.g. congenital heart disease, pulmonary HTN)
- Sarcoidosis
- Myocarditis
- Anderson-Fabry
- Chagas Disease

B. Transmural Infarct

B. Epicardial HE

- Sarcoidosis, Myocarditis, Anderson-Fabry, Chagas Disease

C. Global Endocardial HE

- Amyloidosis, Systemic Sclerosis, Post cardiac transplantation

Ednomyocardial Biopsy – Histologic findings

400x HE.
Subendokardiales Granulom bestehend aus Epitheloidzellen, Riesenzellen vom Langhans Typ mit peripher angeordneten Kernen und wenigen Lymphozyten.
FDG – PET CT (Heart and whole body)
Topics

- Cardiac sarcoidosis
- Whom to evaluate
- Imaging modalities in sarcoidosis
- Follow-up
Different stages of sarcoidosis

<table>
<thead>
<tr>
<th>CMR LGE</th>
<th>$^{18}$F-FDG PET</th>
<th>Interpretation</th>
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</table>
| ![Image](CMR_LGE.jpg) | ![Image]($^{18}$F-FDG_PET.jpg) | **Normal Pattern**  
  - Normal perfusion  
  - No FDG uptake  
  - Physiologic FDG uptake fully suppressed |
| ![Image](CMR_LGE.jpg) | ![Image]($^{18}$F-FDG_PET.jpg) | **Normal Pattern**  
  - Normal perfusion  
  - Diffuse Physiologic uptake |
| ![Image](CMR_LGE.jpg) | ![Image]($^{18}$F-FDG_PET.jpg) | **Early CS vs. Normal Variant**  
  - Active Inflammation  
  - Normal perfusion with focal anterolateral FDG uptake |
| ![Image](CMR_LGE.jpg) | ![Image]($^{18}$F-FDG_PET.jpg) | **Progressive CS**  
  - Active Inflammation  
  - Focal inferolateral perfusion defect with matching FDG uptake |
| ![Image](CMR_LGE.jpg) | ![Image]($^{18}$F-FDG_PET.jpg) | **Advanced CS**  
  - Irreversible fibrosis  
  - Inferoseptal perfusion defect with no FDG uptake |

Modified from Kadkhodayan et al. J Am Coll Cardiol Img 2016;9:603–17
Follow – up of patients with sarcoidosis

Birnie et al. Heart rhythm 2014; 11:1304-1323
CMR in sarcoidosis and prognosis

- 155 pts with systemic sarcoidosis
- 25% of pts had LGE
- Endpoints: death, aborted death

Sarcoidosis and Biomarkers (Troponin)

n=62, age 48y, fup 17 months
Endpoints: Death, ventricular arrhythmia, av-block

Kandolin et al. Am J Cardiol 2015;116:960-964
Summary

Extra-cardiac sarcoidosis with any abnormal:
- Symptoms/exam
- ECG
- EF <50%

No

Annual follow-up:
- Symptoms/exam
- ECG

Yes

Consider CMR

Normal CMR

Annual follow-up:
- Symptoms/exam
- ECG

Abnormal CMR

Consider $^{18}$F-FDG PET

Normal $^{18}$F-FDG PET

Consider ICD

Annual follow-up:
- Symptoms/exam
- ECG

Abnormal $^{18}$F-FDG PET

Consider anti-inflammatory therapy and repeat $^{18}$F-FDG PET to monitor therapy

CMR – cine images
Suggested criteria for screening for cardiac sarcoidosis

**Biopsy proven extra-cardiac sarcoidosis**

Screen with CMR and/or FDG PET if any of the following:
- Symptoms:  significant palpitations
  pre-syncope / syncope
- Abnormal EKG
- Abnormal echocardiogram

**Specific presentations with no prior history of sarcoidosis**

Screen with CMR and/or FDG PET if any of the following:
- Unexplained Mobitz II or 3^{rd} degree AV lock in adults aged < 60 years
- Sustained Monomorphic VT of Unknown Etiology

Who to evaluate?

- Age < 55 y with unexplained II – III av-block
- Age < 55 y with new onset of ECC abnormality
- Inpts with sustained manomarphie VT and/or CMP
- In pts with extracardiac sarcoidosis
Evaluation summary

Biopsy proven extra-cardiac sarcoidosis

Cardiac history, ECG, Echocardiogram

1. Symptom(s) positive (significant palpitations*/pre-syncope/syncope)
2. Abnormal ECG**
3. Abnormal Echocardiogram***

One or more of 1-3

Advanced cardiac Imaging
CMR and/or FDG-PET

None of 1-3

Negative – Low probability of cardiac sarcoidosis

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* palpitations were defined as "prominent patient complaint lasting > 2 weeks"^{25}
** abnormal ECG defined as complete left or right bundle branch block and/or presence of unexplained pathological Q waves in 2 or more leads and/or sustained 2^{th} or 3^{rd} degree AV block and/or sustained or non-sustained VT^{25}
*** abnormal echocardiogram defined as RWMA and/or wall aneurysm and/or basal septum thinning and/or LVEF < 40%^{25}

Birnie et al. Heart rhythm 2014; 11:1304-1323