NON-ISCHEMIC CARDIOMYopathies

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Disclosures and Objectives

Disclosures: Off label use of Gadolinium in cardiac MR

CMR has broad applications outside of ischemic heart disease. Causes of non ischemic cardiomyopathy are broad and have unique appearances on MR. Can optimize scan parameters and prioritize sequences based on pathology you are attempting to visualize. Prognostic value of CMR in cases of non ischemic cardiomyopathy is an expanding field of research and in certain areas has such as HCM has a growing role. CMR is a rapidly evolving field so its role in the diagnosis and management will continue to mature.

NON-ISCHEMIC CMP

1 – ISCHEMIC HEART DISEASE
2 – MYOCARDITIS
3 – HYPERTROPHIC CMP
4 – SARCOIDOSIS
5 – AMYLOIDOSIS
6 – RESTRICTIVE CMP
7 – DILATED CMP
8 – IRON OVERLOAD
9 – LV NON-COMPACTION
10 – ARVD

▶ Definition: NICM are chronic, progressive myocardial diseases with distinct patterns of morphological, functional and electrophysiological changes

▶ Important to determine the etiology in order to guide treatment and prognosis

ISCHEMIC AND NON-ISCHEMIC CMP

LV DYSFUNCTION

ISCHEMIC CMP

NON-ISCHEMIC CMP

ISCHEMIC AND NON-ISCHEMIC CMP

MANAGEMENT

▶ ISCHEMIC CMP – PCI (stent) / CABG / medical treatment

▶ Non-ischemic CMP - Beta-blockade
Causes:
- Viral
- Other infectious agents
- Diabetic
- Autoimmune
- Medications

Imaging findings:
- Global relative enhancement (GRE)
- T2 signal abnormalities
- WMI
- DE
  - Typically affects the epicardial quartile of the LV myocardium
  - DE May decrease during healing
  - DE Can be almost invisible after recovery

• Most common genetically transmitted cardiac disorder
• Leading cause of sudden cardiac death.
• 30% of patients show only isolated hypertrophy
• Bilateral vs concentric
• Myocardial ischemia is common and multifocal
• Myocardial infarcion
• Impaired diastolic dysfunction due to increased rigidity of myocardium
• MR versus Echocardiography

MR findings:
- Asymmetric left ventricular hypertrophy (LVH) confined to LV apex (>15 mm)
- Apical to posterior wall thickness >1.5

Apical Hypertrophic Cardiomyopathy (ApHCM)
- Presentation 41.5 +/- 14.5 years
- Mortality: less than septal variant 83.9 %
- 15 year survival 95%

Malignant ventricular arrhythmias (17%)

Predictors of Mortality: Young age at presentation (<41 years of age) and left atrial enlargement

MR findings:
- Asymmetric left ventricular hypertrophy (LVH) confined to LV apex (>15 mm)
- Apical to posterior wall thickness >1.5

Pure versus Mixed

Prognostic Data from CMR
- Microvascular disease
- Diminished perfusion with hyperemia (increased from baseline reduction) in hypertrophied and non hypertrophied segments
- Leads to myocardial ischemia and scar

Increased amount of scar results in higher susceptibility of NSVT, PVC
- Lower scar in patients without NSVT

Higher myocardial mass correlates with more events and sudden cardiac deaths

- Septal thickness greater than 30 mm carries highest incidence of death

First described in 1929 by Bernstein with non-caseating granuloma on histology

Presentations: 5%
- Symptomatic presentation 5%
- Autopsy 20-50%

Presentations: Arrhythmia (accounts for 60% of fatalities) or restrictive physiology

Diagnosis: MR and endomyocardial biopsy

Scan Protocol: T2 and DE

Key findings: DE, Wall thinning, RVOT, LV

Acute versus chronic phases of sarcoid

Treatment: Early corticosteroid treatment
Protocol: DE, SSFP and T2

Key findings:
- Small left ventricular cavity size
- Biventricular and atrial septal thickening (larger mean mass)
- Subendocardial enhancement
- Rapid clearance of gadolinium from blood pool (diminished T1 difference; expansion of interstitial space)
- Impaired systolic function
- Perfusion anomalies

Diagnostics:
- Echocardiography
- MR

Seeger A, et al. BJR. 2009; 82, 337-342

ENDOMYOCARDIAL FIBROSIS

Braunwald

- Geographical distribution near the Equator
  - Most frequent in children and young adults
  - 50% involvement of RV and LV / 40%
  - 50% involvement of RV and LV / 10% involvement of RV
  - Pericardial effusion (may be quite large)
  - RA is often dilated
- Tricuspid valve is often distorted by the disease process leading to regurgitation
- IV involvement - Fibrosis thickening of the inflow tract and apices, with involvement of the papillary muscles and tricuspid valve
- LV involvement - Fibrosis extending from the apex up the inflow portion of the LV to the posterior aortic valve basin

Our patient

- Tricuspid
  - L V
  - MRI findings suggesting involvement of RV and LV
  - Moderate to large pericardial effusion
  - LA dilation
  - Distortion of tricuspid valve with TR
  - Evidence of papillary muscle involvement
  - RV myocardial hypertrophy
  - Delayed Enhancement demonstrating subendocardial fibrosis in the LV apex

Braunwald 2001: 1720-83

Pericarditis

Acute or Chronic
- Pericarditis
- Acute or Chronic
- Pericardial effusion
- T2 signal of pericardial fluid
- MR features: Restricted pericardial motion
- SSFP: Ventricular coupling increases with strong respiratory dependence

Curt RC et al. Circulation. 2005 Mar 8;111(9):e115-7

ENDOMETRIAL FIBROSIS

Braunwald

- Geographical distribution near the Equator
- Most frequent in children and young adults
- 50% involvement of RV and LV / 40%
- Venezuela
- 18 y / o
- MRI findings suggesting involvement of 50%
- Involvement of RV and LV / 40%
- Pericardial effusion (may be quite large)
- RA is often dilated
- Distortion of tricuspid valve with TR
- Tricuspid valve is often distorted by the fibrous process leading to regurgitation
- RV involvement – Fibrous thickening of the inflow tract and apex with involvement of the papillary muscles and chordae tendineae
- LV involvement – Fibrosis extending from the apex up the inflow portion of the LV to the posterior aortic valve basin

Clinical signs: Progressive heart failure
Causes: Not well understood, suggested ischemic, genetic, viral, toxins, etc.
Feature: Dilated ventricular and atrial size

CMR:
- DE: 41% with DE (28% neomyocardial, 13% pattern similar to ischemia)
- SSFP
- Ejection fraction
- Functional measurements
- EDV>140 for LV
- EDV> 150 for RV
- Prognostic value: DE (endomyocardial) was a predictor of all cause mortality, hospitalization, death and tachycardia

Pathogenesis: Increased trabeculated (non compacted) myocardium relative to compacted myocardium.

History: Ischemia within the trabeculated myocardium and ischemia or fibrosis along the interventricular septum.

Clinical spectrum: Broad range of presentation. Asymptomatic—Heart failure, cavitary thrombus, and death.

Prognostic value: Small studies with small series of patients. Association between amount of delayed enhancement and worse left ventricular function.

Imaging Techniques: 2 Plane assessment for WMA and DE.

Iron Overload: Hemochromatosis and blood dyscrasia.

Clinical: Elevated serum iron, transferrin saturation, and ferritin; death from free iron bound to non transferrin bound iron, which is toxic.

Deposition: skin, liver, pancreas, thyroid and heart.

Protocol: T2* with assessment of interventricular septum and anterior left ventricular wall (shortening of relaxation times).

T2* does not always correlate with ferritin times).

Treatment: Chelation or liver storage.

Deposition: skin, liver, pancreas, thyroid.

Prognostic Value of CMR T2*

Increases serum iron and T2* falls (<50 to >20): No impact on LV function

Increase serum iron and T2* (<20): Severe decrease in LV systolic function.

Prognostic value of CMR T2*

Cardiomyopathy characterized by fibro- fatty infiltration of the right ventricular wall.

May present with arrhythmias of RV origin and even sudden cardiac death.

Difficult diagnosis but important because treatment is available (ICD).

Biopsies unreliable secondary to patchy distribution of fatty infiltration.

Clinical diagnosis based on major and minor criteria.

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Thank You
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