Case-Based Studies in CMR: Non-ischemic CMP

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Background

Cardiomyopathy (CMP) is a general term for myocardial diseases that may lead to impairment of its function and is typically on manifestations with various structural changes. CMP may be classified as primary or secondary: while the causes of primary CMPs are “relatively” unknown (idiopathic), secondary CMPs are related/associated with diseases of other organ systems.
Primary CMPs may strike people of all age any may also be differentiated into dilated CMP (DCM), hypertrophic CMP (HCM), restrictive CMP (RCM) and arrhythmiogenic right ventricular CMP (ARVC).
Recently, many genetic conditions, mutations or constellations have been identified as being related to CMPs but they present with various structural and functional phenotypes. As part of possible genetic aberrations they may also run in families and this fact highlights the possible importance of screening cardiac MRI in family members.

MR Imaging Techniques

At present the most important MR techniques for assessing cardiomyopathies are:

- Cine imaging (SSFP, sGRE)
- Late Gadolinium Enhancement (LGE) imaging

As in daily routine cardiac MR cine imaging is performed for assessment of global ventricular size and function, myocardial mass but also for regional judgment of myocardial thickness and wall motion.
LGE imaging allows for major contributions in regard to specific diagnosis of some CMPs and is performed for assessment of presence of any LGE and the distribution pattern of LGE. LGE distribution pattern is also of outmost importance to differentiate non-ischemic from ischemic CMPs as the typically present with distinct differences (Literature LGE patterns).

DCM

DCM is associated with significant morbidity and mortality at younger ages and it has been shown that the patient group outcome benefits from device (AICD) implant.
Patients typically present with various stages of congestive heart failure (HF). Ventricular dilatation may be part of the sequelae of various pathologic conditions including coronary artery disease (CAD) with infarctions and thus differentiation of ischemic to non-ischemic CMP is the most important in this setting. In ~30-40% of DCM patients LGE is present consisting most often of a linear mid-wall distribution but also of patchy enhancement (1, 2). Possible post-myocarditis dilatation though has to be considered as a differential. Any presence of LGE in DCM though is related to higher risk and worse prognosis in recent studies (2, 3).

HCM

Hypertrophic Cardiomyopathy may present with various patterns of myocardial hypertrophy and thickening with typical <50% of the circumference involved and predominately affecting anterior and septal segments (~25%) (4). This location may especially affect the LV outflow tract and lead to obstruction (HOCM). Patients with extensive obstruction may need to undergo surgical myectomy of transseptal ablation. In HCM patients again the presence of LGE is related to outcome and likelihood of arrhythmia (5, 6).

ARVC

Arrhythmogenic RV CMP is most often family associated (30-70%) and the disease is related to mutations of cell adhesion proteins. These changes may lead to fibro-fatty replacement of myocardium. While predominately the RV wall is involved also LV myocardium may be affected. Based on a recently proposed modification to ARVC diagnosis Task Force Criteria cardiac MR may contribute based on the evaluation of global RV function and size (with cut-off values) and regional RV wall motion abnormalities (e.g. delayed contraction, aneurysmatic outpouching,…).
Although fibro-fatty replacement of myocardium may be suitable for visualization with fat-imaging techniques or LGE is does at present not contribute to the diagnosis according to Task Force Criteria (7). Cardiac MR though has recently shown a high sensitivity and moderate to high specificity in a genotyped ARVC population (8).

RCM

Restrictive CMP may have various underlying reasons and results in a substantially ability of the ventricles to relax and thus filling is impaired. Bi-lateral ventricular thrombi may result in this reduction of ventricular blood flow and flow disturbances.
RCM is of special importance as a differential diagnosis in patients with constrictive pericarditis where the ventricular relaxation and dilation in diastole is reduced by external reasons (pericardium) rather than internal, myocardial changes. This differentiation is of importance for therapeutic planning.

Conclusion

The ability of the differential diagnosis of various non-ischemic CMPs is of high importance in today's clinical cardiac MR. The presence of LGE in many CMPs has impact on the patient prognosis. Cardiac MR may not be possible to come to a final diagnosis but patterns of LGE may substantially narrow the differentials (9).

Background Information

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