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Review

Review of cardiomyopathy imaging

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ABSTRACT

Cardiomyopathies are increasingly being detected on both routine and non-routine imaging. Furthermore, the diagnosis of cardiomyopathy is changing from the traditional method of clinical presentation and cardiac morphology to a quantifiable method based on both cardiac morphology and function. With cardiac magnetic resonance imaging, coronary computed tomography and nuclear medicine increasingly being utilized along with echocardiography in the diagnostic process, it is important for the radiologist to be aware of the relevant criteria in formulating a diagnosis. We aim to provide an overview of the imaging characteristics of the most commonly encountered cardiomyopathies.

Cardiomyopathy is a disease of the myocardium, which usually demonstrates inappropriate ventricular morphology, function or both [1]. It is classified into two groups based on organ involvement. Primary cardiomyopathy is predominantly confined to the myocardium and can be genetic, non-genetic or acquired. Secondary cardiomyopathy is due to a generalized systemic disorder.

We will provide an overview of the imaging of the most commonly encountered cardiomyopathies including dilated, ischaemic, arrhythmogenic right ventricular, left ventricular non-compaction, hypertrophic, restrictive, hypertensive, Takotsubo cardiomyopathies as well as infiltrative aetiologies such as cardiac sarcoidosis and amyloidosis.

Echocardiography usually suffices as the initial and likely only imaging in many patients. However, technical factors limit transthoracic echocardiography (TTE). Poor acoustic windowing is a recognized limitation. Transoesophageal echocardiography (TOE) can overcome most of these imaging issues, however, it is invasive with additional anaesthetic risks.

Cardiac magnetic resonance imaging (CMRI) is currently the gold standard due to high temporal and excellent spatial resolution enabling accurate assessment. It is also viewed as a valuable prognostic tool whereby left ventricular ejection fraction (LVEF) <35% is thought to represent a risk marker for sudden cardiac death in dilated cardiomyopathy (DCM) [2].

Myocardial tissue characterization is possible using contrast-enhanced sequences with early enhancement suggesting ischaemic change or acute inflammation. When gadolinium contrast is administered, it has a slow washout rate in areas of increased extracellular space and fibrosis. Thus, late gadolinium enhancement (LGE) (10–20 min) with nullification of the normal myocardial signal is utilized to identify fibrosis or necrosis. The effect of enhancement is based on divisions of the myocardium: namely the subendocardial, subepicardial and midwall regions.

Cardiac computed tomography (CCT) is used in the investigation of patients with suspected symptomatic obstructive coronary artery disease. It has a high negative predictive value for the exclusion of coronary artery disease. CCT can also assess cardiac chamber size, morphology, and wall thickness. Due to recent advances, electrocardiogram (ECG) gated myocardial blood pool and single photon emission computed tomography (SPECT) studies in nuclear medicine could provide both right and left ventricular volume, ejection fraction and wall motion assessment.

1. Dilated cardiomyopathy

DCM is the most common form of non-ischaemic cardiomyopathy. Most are idiopathic (50%) with other causes including familial (20–35%), drugs (e.g. alcohol) and post-viral.

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DCM is characterized by dilatation and dysfunction of the left ventricle or both ventricles with a subsequent increase in end diastolic and end systolic volumes and a reduced ejection fraction (<50%). Clinically it usually manifests as progressive cardiac failure.

2D echocardiographic findings characteristically show left ventricular (LV) dilatation (with variable involvement of the right ventricle and to a lesser degree the atria), poor systolic myocardial thickening and reduced systolic indices (i.e. LVEF < 50%) (Fig. 1). Additional features may include mitral or aortic valve incompetence. It is believed that mitral valve regurgitation is due to migration of the valve secondary to the change in shape of the left ventricle from ellipsoid to spherical, resulting in poor coaptation of the leaflets.

CMRI has become the gold standard in assessing DCM. Chamber enlargement and a reduced ejection fraction (EF) are easily assessed via 2D and cine imaging (i.e. end-diastolic volumes >140 mL for the left ventricle and >150 mL for the right ventricle) [3].

LGE imaging is based on the accumulation of gadolinium in the extra-cellular space, a feature of fibrosis that replaces normal myocardium (Fig. 1). LGE has been described as being present in 12–35% of cases, with the most common pattern being enhancement affecting the midmyocardial layer.

The role of CCT is currently in excluding obstructive coronary artery disease. Other characteristic findings of DCM on CCT include increased diastolic LV internal diameter measurement greater than 5.6 cm (upper limits of normal) [4]. CCT also plays a role in determining DCM prognosis, based on ejection fraction (<35% for symptomatic patients and <30% for asymptomatic patients.

SPECT myocardial perfusion scintigraphy (MPS) and positron emission tomography (PET) can quantify myocardial perfusion to exclude ischaemic aetiology. DCM can demonstrate homogeneous distribution of blood flow on SPECT MPS and glucose metabolism on the F-18 fluorodeoxyglucose (^{18}F-FDG) images respectively in contrast to ischaemic cardiomyopathy cases where left ventricular segments demonstrate discrete reductions in perfusion and or reduced glucose utilization in segmental vascular territorial distribution. With the utilization of medical therapy, defibrillator placement and cardiac transplantation, survival rate has improved to 80% [5].

2. Hypertrophic cardiomyopathy

Hypertrophic cardiomyopathy (HCM) has an incidence of 1:500. An autosomal dominant inherited condition with variable penetrance and expression, HCM is defined by the presence of LV hypertrophy in the absence of LV dilatation. Although clinical presentations range from asymptomatic to sudden cardiac death, particularly important in young athletes, dyspnoea is the most common symptom due to a combination of LV diastolic impairment, LV outflow tract obstruction and/or mitral regurgitation caused by systolic anterior motion (SAM) of the mitral valve. SAM is the paradoxical movement of the anterior mitral valve leaflet towards the left ventricular outflow tract (LVOT) during systole. Mechanisms include interventricular septal hypertrophy causing LVOT stenosis, with increased flow velocity and decreased pressure (Venturi effect) of ejected blood; or displacement of the papillary muscles with altered chordal tension on the mitral valve leaflet.

Diagnosis is usually established with echocardiography confirming LV hypertrophy typically involving the interventricular septum. However, any diffuse or segmental pattern of LV wall thickening is possible. Diagnostic criterion is a maximal LV wall thickness greater than or equal to 15 mm in the end-diastolic phase (normal range $\leq$12 mm) [6]. However, the degree of hypertrophy may be underestimated by echocardiography leading to delayed diagnosis.

Variation of myocardial hypertrophy may include asymmetric or non-contiguous myocardial involvement. Right ventricular (RV) muscular hypertrophy (17%) [7] may also be seen, most commonly involving the mid to apical portion of the right ventricle.

CCT may demonstrate basal myocardial hypertrophy with a small outflow tract, and an enlarged and elongated mitral valve. The associated haemodynamic obstruction may also demonstrate SAM of the mitral valve leaflets and mid-systolic contact with the ventricular septum with either subaortic obstruction or mitral regurgitation. Differentials include hypertensive cardiomyopathy however HCM is usually non-concentric and asymmetric.

CMRI can help differentiate HCM from other causes of LV hypertrophy such as increased afterload (i.e. hypertension or aortic stenosis) by measuring the ratio of end-diastolic wall thickness to end-diastolic volume, a measure of wall thickness in relation to heart size. A cut-off value of 0.15 mm mL m^2 gives positive and negative predictive values of 95% and 94% respectively [8]. A majority of patients with true HCM have LGE, usually involving the interventricular septum or points of insertion of the RV free wall into the LV (Fig. 2). The presence of LGE is an independent predictor of both morbidity and mortality.

Medical management of HCM is largely aimed at ventricular relaxation and filling, via the utilization of beta-blockers and calcium channel blockers. Implantable cardiac defibrillator (ICD) is advised for patients of HCM deemed at high risk of sudden cardiac death as it effectively reduces the chance of lethal ventricular tachyarrhythmias.

3. Arrhythmogenic right ventricular cardiomyopathy

Arrythmogenic right ventricular cardiomyopathy (ARVC) is characterized prototypically by the total or partial replacement of the right ventricular myocytes by adipose and fibrous tissue. The most common location for tissue transformation is between the anterior infundibulum, the right ventricular apex, and the inferior/diaphragmatic aspect of the right ventricle. This is termed the “triangle of dysplasia”. It may also involve the left ventricular myocardium in severe cases.

The pathogenesis is not established but four main hypotheses are now considered: myocyte apoptosis giving rise to myocardial muscle loss and subsequent fibrofatty replacement, dysontogenetic RV development, degenerative RV disorders due to metabolic aetiology, and healing by fibrofatty replacement after an inflammatory process such as myocarditis.

ARVC usually manifests in adolescents or young adults with a prevalence of 1 in 5000 with a 3:1 male to female ratio. It is thought to be transmitted by an autosomal dominant mode of inheritance with incomplete penetrance. The most common presentation is ventricular arrhythmias with left bundle branch block and can predispose to sudden death. ARVC accounts for 20 percent of sudden cardiac death cases [9,10].

The diagnosis is based on the presence of major and minor criteria encompassing genetic, electrocardiographic, pathophysiologic, and histopathologic factors. Echocardiographic investigation is limited because much of the right ventricular free wall lies posterior to the sternum and ribs. Furthermore, it is not useful at demonstrating intramyocardial fat. Despite this, there are major and minor echocardiographic findings detailed in the modified criteria of the World Heart Federation task force revised in 2010, listed in Table 1. Furthermore, echocardiography can demonstrate a hypokinetic and dilated right ventricle. A right ventricular outflow tract (RVOT) dimension >30 mm from the parasternal long-axis view has a sensitivity (89%) and specificity (86%) for the diagnosis of ARVC [11].