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Diffuse, Subendocardial Vasculitis Identified by Cardiovascular Magnetic Resonance. Use of Images to Learn Pathophysiology

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Abstract

Diffuse Subendocardial Vasculitis (DSV), either primary or secondary, may occur during autoimmune diseases and lead to high mortality and morbidity. Recently, Cardiovascular Magnetic Resonance (CMR), a non-invasive, non-radiating technique, capable to perform tissue characterization has been used in the evaluation of cardiac vasculitis in the autoimmune diseases. CMR by performing tissue characterisation can detect diffuse, subendocardial oedema during the acute phase of DSV. To assess oedema, T2STIR images, T2 mapping and native T1 mapping can be used. Furthermore, diffuse subendocardial fibrosis can be also detected in both acute and chronic DSV. For this purpose, Late Enhanced Images (LGE) should be taken 15 minutes after the intravenous injection of the paramagnetic contrast agent gadolinium. Diffuse, subendocardial scar can be quantified and expressed as percentage of LV mass and this index have an important clinical value together with the LV ejection fraction in both ischemic and nonischemic cardiomyopathy.

DSV can be identified by CMR in both acute and chronic autoimmune diseases. Acute DSV has the potential to be reversed completely by autoimmune and cardiac treatment modification. CMR is a powerful tool to early diagnose DSV and guide therapeutic decisions. However, further studies are needed to establish its impact in the patients' risk stratification and treatment follow up.

Keywords: Vasculitis; Autoimmune diseases; Cardiac magnetic resonance

Introduction

Vasculitis, either primary or secondary, may occur during autoimmune diseases and lead to high mortality [1,2]. Primary (idiopathic) inflammatory vasculitides may involve large vessels as in Giant Cell Arteritis (GCA) or Takayasu Arteritis (TA), and medium- or small-sized vessels as in Kawasaki Disease (KD), polyarteritis nodosa, Antineutrophilic Cytoplasmic Antibody (ANCA) associated vasculitides, Behcet Disease (BD), primary CNS angiitis, and thromboangiitis obliterans.

Secondary vasculitides may develop during the course of sarcoidosis, systemic lupus erythematosus, antiphospholipid syndrome, rheumatoid arthritis, infections and angiitis after radiation exposure) or conditions that may mimick vasculitis, such as vascular neoplasias and ergotism may also involve large and medium-sized arteries [3].

While advances in serologic testing have succeeded earlier diagnosis, affected patients still present poor outcome, due to lack of accurate diagnostic tool [4]. Recently, Cardiovascular Magnetic Resonance (CMR), a non-invasive, non-radiating technique, capable to perform tissue characterization has been already used in the evaluation of autoimmune diseases [5-8].

Until now, the typical CMR pattern of Diffuse, Subendocardial Vasculitis (DSV) has been described in selected cases including CSS, anti-phospholipid syndrome, systemic lupus erythematosus, and rheumatoid arthritis [8-10].

How Can CMR Detect Diffuse, Subendocardial Vasculitis?

CMR, due to its capability to perform tissue characterisation, can detect diffuse, subendocardial oedema during the acute phase of DSV (Figure 1). To assess oedema, T2STIR images, T2 mapping and native T1 mapping can be used [10,11].

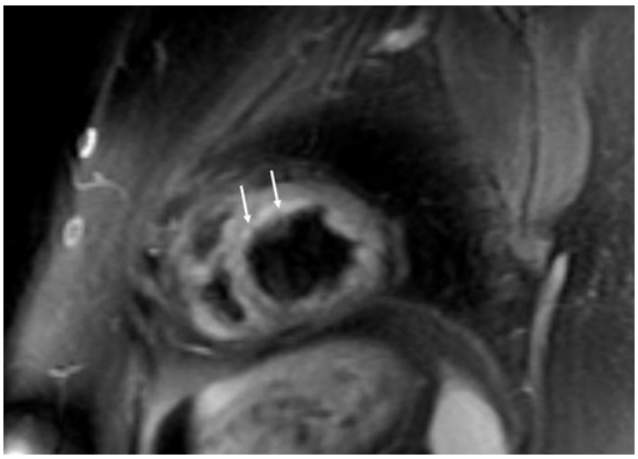

Figure 1: Diffuse subendocardial oedema, due to cardiac vasculitis in a patient with CSS

Furthermore, diffuse subendocardial fibrosis can be also detected (Figure 2) in both acute and chronic DSV. For this purpose, Late

Enhanced Images (LGE) should be taken 15 minutes after the intravenous injection of the paramagnetic contrast agent gadolinium. Diffuse, subendocardial scar can be quantified and expressed as percentage of LV mass and this index have an important clinical value together with the LV ejection fraction in both ischemic and nonischemic cardiomyopathy [12,13] and specifically in Churg Strauss syndrome [14].

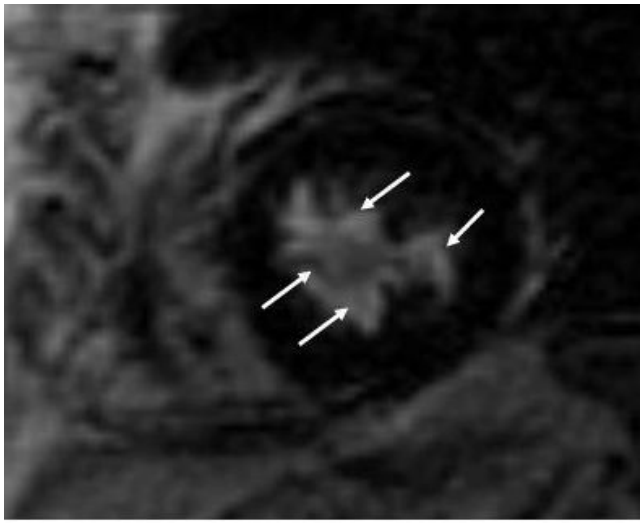

Figure 2: Diffuse subendocardial fibrosis, due to cardiac vasculitis in a patient with CSS

Clinical Implications in Diagnosis, Follow Up and Treatment

Until recently, there were only scarce data about vasculitis in autoimmune diseases, due to lack of an accurate diagnostic tool. This is a preliminary review about the clinical implications of CMR-documented DSV in autoimmune diseases. LV impairment can be the common endpoint in a significant percentage of our autoimmune patients, presented with DSV that is in agreement with previous studies, supporting that DSV may contribute to severe LV dysfunction and death [10,15,16]. In these studies, CMR not only identified this entity, but also disease acuity that led to treatment modification and final improvement of both clinical and CMR pattern in acute cases. Furthermore, in patients with Churg Strauss syndrome, CMR identified the cardiac lesions, early before symptoms' development [17,18], detected the pathophysiologic background of ventricular tachycardia (VT) [19] and revealed complete regression of cardiac lesions after successful rheumatic and cardiac treatment [20].

In chronic cases, the detection of DSV and the early start of ACE-inhibitors even in those with normal LVEF have the potential to prevent overt LV dysfunction [21]. Finally, according to French Vasculitis Study group, CMR is promising to detect those patients with a less favorable cardiac outcome [22].

According to previous studies, vasculitis can be a sign of cardiac involvement in sarcoidosis [23], SLE [24,25], SSc [26,27] and RA [8,28]. In a pathology study of 161 RA, systemic vasculitis was observed in 22.4percentage and the most frequently involved organ was the heart (66.7percentage), although the presence of clinically detected vasculitis in RA was substantially low [28]. However, all these studies have various limitations including 1) referral bias 2) lack of endomyocardial biopsy 4) Lack of long-term follow up 5) Lack of comparison with autoimmune patients presenting different CMR lesions (myocarditis or myocardial infarction) 6) lack of treatment evaluation. Additionally, limitations concerning the CMR application per se, such as claustrophobia, metallic clips, cardiac devices, impaired renal function, lack of availability/expertise and high cost should be taken under consideration, before a CMR evaluation will be asked.

Conclusion

DSV can be identified by CMR both in acute and chronic autoimmune diseases. Acute DSV has the potential to be reversed completely by autoimmune and cardiac treatment modification. CMR is a powerful tool to early diagnose DSV and guide therapeutic decisions. However, further studies are needed to establish its impact in the patients risk stratification and treatment follow up.

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