

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

Sophie Mavrogeni<sup>†</sup>, George Markousis-Mavrogenis and Genovefa Kolovou

Onassis Cardiac Surgery Center, Athens, Greece

<sup>†</sup>Corresponding author: Sophie Mavrogeni, Onassis Cardiac Surgery Center, Athens, Greece, Tel: 302109882797; E-mail: soma13@otenet.gr

Received date: April 06, 2016; Accepted date: May 05, 2016; Published date: May 08, 2016

Copyright: © 2016 Mavrogeni S, et al. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

## 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.

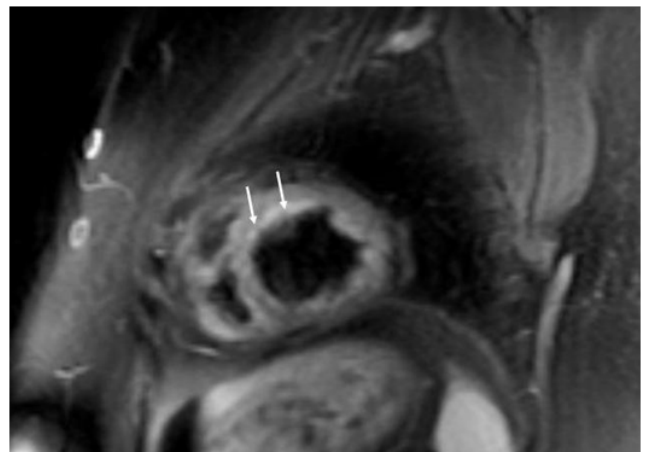
Secondary vasculitides may develop during the course of sarcoidosis, systemic lupus erythematosus, antiphospholipid syndrome, rheumatoid arthritis, infections and angiitis after radiation exposure) or conditions that may mimic vasculitis, such as vascular neoplasias and ergotism may also involve large 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?

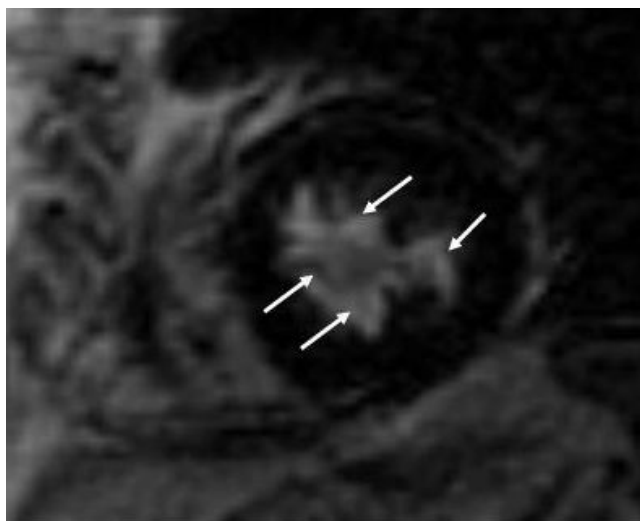
CMR, due to its capability to perform tissue characterisation, can detect diffuse, subendocardial oedema during the acute phase of DSV (Figure 1). To assess oedema, T2STIR images, T2 mapping and native T1 mapping can be used [10,11].



**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.

### References

1. Scott DG, Watts RA (2000) Systemic vasculitis: epidemiology, classification and environmental factors. *Ann Rheum Dis* 59: 161-163.
2. Fayad ZA, Greenberg JD, Bucarius J (2012) Subclinical vasculitis as a potential mechanism to explain the heightened cardiovascular risk in rheumatoid arthritis. *Circulation* 126: 2449-2451.
3. Spira D, Kötter I, Ernemann U, Balletshofer B, Pfannenber CA, et al. (2010) Imaging of primary and secondary inflammatory diseases involving large and medium-sized vessels and their potential mimics: a multitechnique approach. *AJR Am J Roentgenol* 194: 848-856.
4. Pipitone N, Versari A, Salvarani C (2008) Role of imaging studies in the diagnosis and follow-up of large-vessel vasculitis: an update. *Rheumatology (Oxford)* 47: 403-408.
5. Mavrogeni S, Manoussakis MN, Karagiorga TC, Douskou M, Panagiotakos D, et al. (2009) Detection of coronary artery lesions and myocardial necrosis by magnetic resonance in systemic necrotizing vasculitides. *Arthritis Rheum* 61: 1121-1129.
6. Mavrogeni S, Vassilopoulos D (2011) Is there a place for cardiovascular magnetic resonance imaging in the evaluation of cardiovascular involvement in rheumatic diseases? *Semin Arthritis Rheum* 41: 488-496.
7. Mavrogeni S, Spargias K, Markussis V, Kolovou G, Demerouti E, et al. (2009) Myocardial inflammation in autoimmune diseases: investigation by cardiovascular magnetic resonance and endomyocardial biopsy. *Inflamm Allergy Drug Targets* 8: 390-397
8. Raman SV, Aneja A, Jarjour WN (2012) CMR in inflammatory vasculitis. *J Cardiovasc Magn Reson* 14: 82.
9. Mavrogeni S, Cantini F, Pohost GM (2013) Systemic vasculitis: an underestimated cause of heart failure - assessment by cardiovascular magnetic resonance. *Rev Cardiovasc Med* 14: 49-55.
10. Mavrogeni S, Sfikakis PP, Gialafos E, Karabela G, Stavropoulos E, et al. (2013) Diffuse, subendocardial vasculitis. A new entity identified by cardiovascular magnetic resonance and its clinical implications. *Int J Cardiol* 168: 2971-2972.
11. Campbell RT, Jhund PS, Dalzell JR, Cannon J, Mordi I, et al. (2015) Diagnosis and resolution of Löeffler endocarditis secondary to eosinophilic granulomatosis with polyangiitis demonstrated by cardiacmagnetic resonance-T2 mapping. *Circulation* 131: 114-117.
12. Kwong RY, Sattar H, Wu H, Vorobiof G, Gandla V, et al. (2008) Incidence and prognostic implication of unrecognized myocardial scar characterized by cardiac magnetic resonance in diabetic patients without clinical evidence of myocardial infarction. *Circulation* 118: 1011-1020.
13. Neilan TG, Coelho-Filho OR, Danik SB, Shah RV, Dodson JA, et al. (2013) CMR quantification of myocardial scar provides additive

- prognostic information in nonischemic cardiomyopathy. *JACC Cardiovasc Imaging* 6: 944-954.
14. Jeong HC, Kim KH, Cho JY, Song JE, Yoon HJ, et al. (2015) Cardiac involvement of churg-strauss syndrome as a reversible cause of dilated cardiomyopathy. *J Cardiovasc Ultrasound* 23: 40-43.
  15. Mavrogeni S, Bratis K, Sfrikakis PP (2013) Pleuro-pericarditis, vasculitis, subendocardial and nodular biventricular fibrosis. The multiple faces of systemic sclerosis detected by cardiac magnetic resonance in the same patient. *Int J Cardiol.* 163: e26-27.
  16. Bély M, Apáthy A (1996) Vasculitis in rheumatoid arthritis. *Orv Hetil* 137: 1571-1578.
  17. Yune S, Choi DC, Lee BJ, Lee JY, Jeon ES, et al. (2016) Detecting cardiac involvement with magnetic resonance in patients with active eosinophilic granulomatosis with polyangiitis. *Int J Cardiovasc Imaging* pp 1-8.
  18. Colin GC, Vancraeynest D, Hoton D, Jonard P, Gerber B (2015) Complete heart block caused by diffuse pseudotumoral cardiac involvement in granulomatosis with polyangiitis. *Circulation* 132: e207-210.
  19. Tung R, Rahimi AR, Gelfand EV, Kwaku KF (2009) Vasculitis presenting with ventricular tachycardia. *J Rheumatol* 36: 2127-2129.
  20. Post MC, Boomsma MF, van Heesewijk JP, Grutters JC, Van der Heyden J (2011) Cardiac magnetic resonance imaging showing complete resolution of subendocardial involvement in Churg-Strauss syndrome. *J Thorac Imaging* 26: 81-2.
  21. Hunt SA, Abraham WT, Chin MH, Feldman AM, Francis GS, et al. (2005) ACC/AHA 2005 Guideline Update for the Diagnosis and Management of Chronic Heart Failure in the Adult. *Circulation* 112: e154-e235.
  22. Dunogué B, Terrier B, Cohen P, Marmursztejn J, Legmann P, et al. (2015) Impact of cardiac magnetic resonance imaging on eosinophilic granulomatosis with polyangiitis outcomes: A long-term retrospective study on 42 patients. *Autoimmun Rev* 14: 774-780
  23. Ward EV, Nazari J, Edelman RR (2012) Coronary artery vasculitis as a presentation of cardiac sarcoidosis. *Circulation* 125: e344-346.
  24. Sung JM, Hsu SC, Chen FF, Huang JJ (2002) Systemic lupus erythematosus presented as non-inflammatory necrotizing vasculopathy-induced ischemic glomerulopathy and small vessels-related ischemic cardiomyopathy. *Lupus* 11: 458-462.
  25. Piette JC, Chapelon C, Boussen K, Mouthon JM, Guillevin L, et al. (1987) Lupus vasculitis. *Ann Med Interne (Paris)* 138: 425-436.
  26. Abu-Shakra M, Koh ET, Treger T, Lee P (1995) Pericardial effusion and vasculitis in a patient with systemic sclerosis. *J Rheumatol* 22: 1386-1388.
  27. Sharma A, Dhooria A, Aggarwal A, Rathi M, Chandran V, et al. (2016) Connective Tissue Disorder-Associated Vasculitis. *Curr Rheumatol Rep* 18: 31.
  28. Bély M, Apáthy A (1996) Vasculitis in rheumatoid arthritis. *Orv Hetil* 137: 1571-1578.