

Rheumatology: Current Research

Clinical Image Article

Matched Perfusion Defects and Abnormal Delayed Enhancement at Cardiac MR Imaging in Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss Syndrome)

Christina L. Luong¹, Hardeep Mahal^{1*}, Darra T. Murphy² and David A. Wood³

¹Department of Medicine, University of British Columbia, Vancouver General Hospital, Vancouver, British Columbia, Canada

²Department of Radiology, University of British Columbia, Vancouver General Hospital, Vancouver, British Columbia, Canada

³Division of Cardiology, University of British Columbia, Vancouver General Hospital, Vancouver, British Columbia, Canada

*Corresponding author: Dr. Hardeep Mahal, Vancouver General Hospital, University of British Columbia, 6049 Bryant St Burnaby BC Canada V5H1X8, Canada, Tel: 604-614-1247; E-mail: hmahal.11@gmail.com

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Clinical Image

A 67-year-old man with a history of asthma and sinusitis presented with severe chest pain associated with constitutional symptoms and parasthesias. Blood tests showed peripheral eosinophilia of 17.6×10^9 /l and a troponin of 13.7μ mol/l. ECG revealed nonspecific ST-segment changes. Computed tomography of the head demonstrated pansinusitis. He underwent urgent coronary angiography, which revealed non-obstructive disease. Transthoracic echocardiogram demonstrated interval decrease of left ventricular function at 45% with a thickened apex. Further investigations yielded a normal CCP, ANA, ENA, IgM/IgA titers and negative Hepatitis B/C, ANCA serology. Additionally, elevated RF 81 kIU/L, IgG 18.7 μ g/L, IgE 5558 μ g/L, and CRP 54.4 mg/L were noted. A diagnosis of eosinophilic granulomatosis with polyangiitis (EGPA) was made based on ACR criteria.



Figure 1: Showing the utility of cardiac MRI (CMR) in confirmation of cardiac involvement in eosinophilic granulomatosis with polyangiitis (EGPA), formerly Churg-Strauss syndrome.

Cardiac involvement was confirmed by CMR with abnormal subendocardial delayed enhancement; a finding usually attributed to fibrosis due to eosinophilic infiltration (Figure 1a and 1c). However, this was also associated with a matched perfusion defect on first pass imaging (Figure 1b and 1d); a finding that is felt to represent inflammatory myocardial edema and microvascular dysfunction [1]. This was supported by subtle increased signal in the same distribution on a T2 fat-saturation sequence (not shown). Although subendocardial delayed enhancement is a well-established modality for identification of cardiac inflammation or fibrosis, CMR perfusion provides a means of differentiating the two processes. Cardiac manifestations of EGPA are variable and can affect virtually every cardiac structure. EGPA cardiomyopathy is associated with significant morbidity and mortality with 5-year survival of 78.2% compared to 91.6% for those without cardiomyopathy [2]. It remains unclear whether therapy should be titrated to CMR evidence of inflammation. In this case, the CMR perfusion finding was essential for identification of cardiac involvement and provided initial direction for therapy. Perfusion CMR serves as a useful adjunct to myocardial delayed enhancement at CMR in evaluation of acute inflammatory cardiomyopathies.

References

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