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Review

Impact of cardiac magnetic resonance imaging on eosinophilic granulomatosis with polyangiitis outcomes: A long-term retrospective study on 42 patients

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ABSTRACT

Objective: To determine the diagnostic and prognostic significance of cardiac magnetic resonance imaging (CMRI) in a cohort of patients with eosinophilic granulomatosis with polyangiitis (EGPA).
Methods: We conducted a monocentric retrospective study including 42 EGPA patients who had consecutively undergone CMRI at diagnosis or during follow-up, independently of signs of cardiac involvement.
Results: Forty-two patients (male 59.5%, mean age at diagnosis 46.5 years) were included. ANCA was positive in 26.2%, and median EGPA duration before the 1st CMRI screening was 5 months. Seventeen (40.5%) were diagnosed with cardiomyopathy, independently of CMRI findings. CMRI showed myocardial late gadolinium enhancement (LGE) in 82.4% patients with cardiomyopathy vs. 44% without cardiomyopathy ($P = 0.024$). Using LGE as the sole criterion, CMRI sensitivity and specificity for diagnosing cardiomyopathy were 82.4% and 56%, respectively. Among the 15 patients with cardiomyopathy who underwent additional CMRI during follow-up, CMRI-detected cardiac lesions had improved in 7 patients, while those of 8 patients worsened or stabilized despite treatment. These latter patients presented with significantly more cardiac events during follow-up ($P = 0.026$). No differences were found between non-cardiomyopathic patients with or without CMRI anomalies concerning EGPA cardiac manifestations and outcomes.
Conclusion: The diagnostic significance of myocardial LGE in EGPA patients remains uncertain and should not be the only criterion for cardiomyopathy diagnosis. For patients with no other signs of cardiomyopathy, CMRI-detected anomalies do not seem to adversely affect prognosis or outcome. For patients with cardiomyopathy, CMRI reassessment seems promising in detecting patients with a less favorable cardiac outcome.

Contents

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1. Introduction

Cardiac involvement is a major concern in eosinophilic granulomatosis with polyangiitis (EGPA, formerly called Churg–Strauss syndrome), both frequent (described in 13.5% of patients) and severe [1], especially among anti-neutrophil cytoplasmic antibody (ANCA)-negative patients in which it is reported in 22.4% [2] to 49% [3]. Cardiomyopathy is the main cause of first-year and overall mortality in EGPA [4], accounting for almost one third of deaths [5], due to acute or chronic heart failure and severe arrhythmias [1,4,6,7]. EGPA cardiomyopathy increases the mortality risk by a four-fold ratio, with 5-year survival rates dropping from 91.6% to 78.2% when the myocardium is involved [5]. Cardiomyopathy has therefore long been recognized as a poor-prognosis factor for all vasculitides, as defined by the 1996-Five Factor Score (FFS) [8], as well as by the "revisited" FFS in 2011 [9].

Despite the severity of EGPA cardiomyopathy, there is some evidence that electrocardiogram (ECG) and clinical manifestations may be reversible under treatment [1,10,11], thus stressing the importance of diagnosing and treating cardiac lesions of EGPA early and effectively. Cardiac magnetic resonance imaging (CMRI) has been well studied in sarcoidosis, in which myocardial late gadolinium enhancement (LGE) is the main sign of granulomatous inflammation [12,13]. This anomaly has been described in other conditions, including post-infarct lesions [14], acute viral myocarditis [15], and more recently in EGPA [16–20]. In EGPA, myocardial LGE may be detected in 83% [18] to 100%[19] of patients with symptomatic cardiomyopathy, but also in up to a third of asymptomatic patients [19,20], raising the issues of both the diagnostic and prognostic significances of such CMRI anomalies. Results drawn from previous prospective studies from our group showed that LGE in non-cardiomyopathic patients may not be a risk for further cardiac complications [19], but may nevertheless be accessible to immunosuppressive treatment [21].

In order to better understand the diagnostic and prognostic significance of CMRI anomalies in EGPA, we conducted a retrospective analysis of 42 EGPA patients who had undergone CMRI at diagnosis or during follow-up, independently of cardiac symptoms.

2. Patients and methods

2.1. Patients

This retrospective, descriptive and monocentric study included 42 EGPA patients followed at our Department of Internal Medicine (French national referral center for systemic necrotizing vasculitides), who had consecutively undergone CMRI at diagnosis or during follow-up, independently of signs of cardiac involvement. Twenty of these patients had already been enrolled in a previous observational and prospective CMRI study [19], and eight of these 20 patients had also been included in a second therapeutic and open-labeled prospective study [21]. The other patients underwent CMRI as part of their follow-up or diagnostic work-up. All patients had a diagnosis of EGPA as defined by the Lanham diagnostic criteria [6] and/or American College of Rheumatology classification criteria [22].

2.2. Cardiac magnetic resonance imaging assessment

The initial CMRI was performed for all patients between February 2004 and June 2008, in the Radiology Department of Cochin hospital. Subsequent CMRI was performed as part of a follow-up procedure for 17 patients. CMRI was performed at our center with a 1.5-T imager Avanto 76 × 32 SQ (Siemens Medical Solutions, Erlangen) using a dedicated cardiac, ECG-triggered, phased-array coil, as previously described [23].

2.3. Clinical, biological and other imaging data

Clinical and extra-clinical data were collected by BD from medical files in a standardized case report form, and included: age, gender, date of EGPA diagnosis, date of asthma diagnosis, date of first vasculitis symptoms, manifestations of EGPA, cardio-vascular risk factors (family history, tobacco use, hypercholesterolemia, diabetes, obesity, hypertension), personal history of cardio-vascular events unrelated to EGPA, ANCA status and type (as defined by immunofluorescence and ELISA techniques), eosinophil counts at diagnosis and at time of CMRI, presence of cardiac clinical signs or symptoms (chronic dyspnea, thoracic pain, palpitations, syncope, signs of heart failure), ECG results, levels of troponin and BNP or NT-proBNP, results of echocardiography, when performed. CMRI results were collected using the full radiological report. Results of chest X-ray, thoracic CT-scan, myocardial scintigraphy, and coronography were also collected. Follow-up data included CMRI results, when performed, as well as the occurrence of an EGPA relapse, death, or a major cardiac event (as defined by acute heart failure, arrhythmia, heart surgery, coronary event, conductive block). Data concerning the evolution of clinical cardiac symptoms, ECG anomalies, and echocardiographic results were also collected.

2.4. Definition of EGPA-related cardiomyopathy

Patients were considered having EGPA-related cardiomyopathy when clinical or extra-clinical signs of patent cardiopathy were present, and no other causes of cardiopathy could be found, despite extensive extra-clinical work-up. CMRI results were usually not taken into account for diagnosing EGPA-related cardiomyopathy. However, considering the retrospective character of the analysis, this possibility may not be totally ruled out.

2.5. Statistics

Statistical analyses were performed using the R software (version R.2.15.1). Characteristics of patients were compared according to their ANCA or cardiomyopathic status using non-parametric tests: respectively Fisher's and Mann–Whitney tests when the studied variables were either qualitative or quantitative. Statistical significance was considered when the α risk was <0.05.