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Patient with Churg Strauss Syndrome and Myocarditis Treated with Cyclophosphamide

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ABSTRACT

Cardiac involvement in Churg Strauss Syndrome is common and a poor prognostic indicator. Myocarditis in Churg Strauss Syndrome can present in different ways. It has been shown that basic cardiac investigations including echocardiography can be normal even in symptomatic patients. More recently cardiac magnetic resonance imaging (MRI) has been shown to be more sensitive in its diagnosis. Our case report describes a 45 year old male who presented with palpitations and breathlessness. Echocardiography was normal but cardiac MRI demonstrated abnormalities consistent with Myocarditis. He was treated with Cyclophosphamide and follow up MRI imaging demonstrated complete resolution of these abnormalities which was accompanied by resolution of symptoms. This case therefore supports the use of cardiac MRI in Churg Strauss Syndrome as a sensitive diagnostic tool and as a means of monitoring response to therapy. It also supports the therapeutic effectiveness of Cyclophosphamide therapy in Churg Strauss related Myocarditis, something that has yet to be assessed on a large scale.

Keywords: Cardiac MRI; Cyclophosphamide; Myocarditis; Churg Strauss Syndrome

1. Introduction

Cardiac involvement in Churg Strauss Syndrome is common, with a prevalence of up to 62% [1]. It is the leading cause of mortality in this disease [2]. The necrotising granulomatous Myocarditis associated with Churg Strauss can manifest in different ways. Clear diagnosis is difficult without myocardial biopsy. Unlike patients with renal disease, patients with cardiac involvement are usually ANCA (perinuclear type) negative [3,4]. Furthermore traditional cardiac investigations including echocardiography can be normal even in symptomatic patients. More recently cardiac MRI has been proposed as a useful diagnostic and monitoring tool in this condition. Here we present a case of Churg Strauss Myocarditis in which cardiac MRI was utilised to good effect.

2. Case Report and Methods

A 45 year old man, recently diagnosed with Churg Strauss Syndrome, presented with a two week history of palpitations, presyncope and breathlessness on exertion. He had been taking Prednisolone since 1999 when he had been initially diagnosed with chronic eosinophilic pneumonia, and was currently prescribed 30mg daily.

He was admitted in January 2010 for investigation of his symptoms. Electrocardiography and serum Troponin *T* were normal. A 24 hour tape showed sinus rhythm with 22 isolated ventricular ectopics and 962 episodes of ventricular bigeminy. An echocardiogram demonstrated normal biventricular size and function, and no significant abnormalities.

He was incidentally prescribed Myfortic (Mycophenolic Acid) 720 mg twice a day in February 2010, in addition to Prednisolone, on the basis of persistent sinus symptoms.

Based on his ongoing cardiac symptoms he subsequently underwent cardiac MRI in April. This demonstrated a mildly reduced left ventricular ejection fraction (LVEF) of 46%. There was mild hypokinesia of the basal septum with late contrast hyper-enhancement; consistent with Myocarditis (see **Figure 1**). There was no evidence of endocardial disease or myocardial hypertrophy.

Based on the MRI findings he was commenced on Cyclophosphamide in May and Myfortic therapy was withdrawn. He received 12 doses (at 15 mg/Kg body weight) as an intravenous infusion every 2 weeks.

A cardiac MRI was repeated in August after 6 doses of Cyclophosphamide. This demonstrated clear improvement with a LVEF of 62%. There was very subtle hyper-enhancement in the basal septum but significantly less than previously. He was also now asymptomatic.

He received his last Cyclophsophamide dose in October 2010 and was then commenced on Azathioprine therapy at 150 mg a day. A third MRI was performed two months

after conversion to maintenance immunosuppression in December 2010. He remained symptom free at this time. This demonstrated preserved LV function, with an ejection fraction of 61%. There was now no evidence of the previously noted abnormalities (see **Figure 2**).

In summary; a 45 year old man with Churg Strauss syndrome, on long term oral corticosteroid therapy, developed symptoms of palpitations and breathlessness. Echocardiography was normal but cardiac MRI demonstrated mildly impaired LVEF and basal hypokinesia; consistent with Myocarditis. His symptoms were successfully treated with Cyclophosphamide and repeat cardiac MRI was able to demonstrate resolution of the abnormalities.

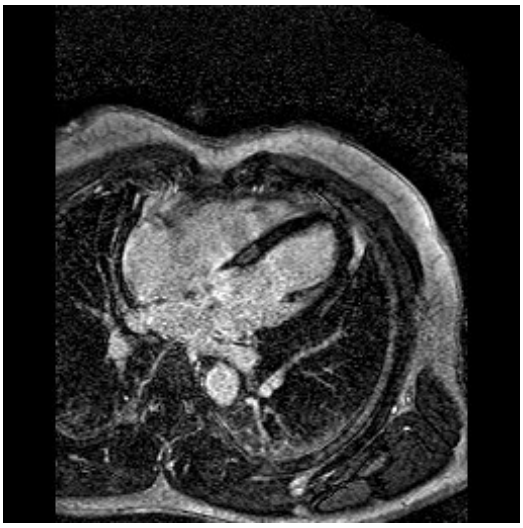

Figure 1. MRI April 2010. 2D Horizontal Long Axis view demonstrating hyper-enhancement in basal septum after gadolinium administration.

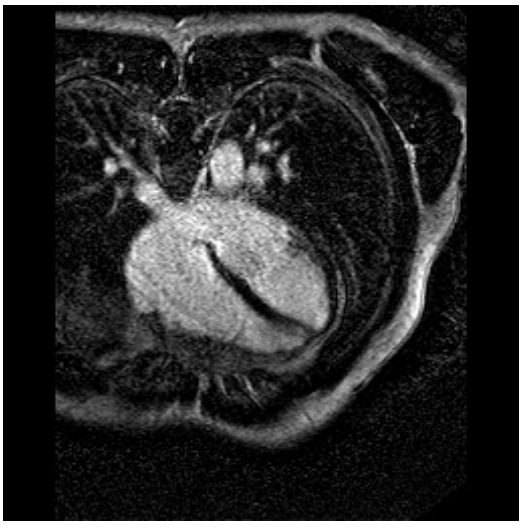

Figure 2. MRI December 2010. 2D Horizontal Long Axis view showing no hyper-enhancement in basal septum after gadolinium administration.

3. Discussion

Cardiac MRI has been shown to be more sensitive than basic echocardiography in diagnosing Myocarditis [5]. MRI can demonstrate delayed enhancement in affected myocardial segments which has been shown to correlate with histologically proven myocardial inflammation or fibrosis [6]. One study showed such abnormalities to occur even in some asymptomatic patients [5]. Cardiac MRI has also been advocated for monitoring in patients who receive immunosuppressive treatment [7]. Follow up cardiac MRI in some of these patients showed resolution of abnormalities after treatment which correlated with clinical improvement. Our case report further supports the use of cardiac MRI as a diagnostic tool. Without evidence of myocardial disease on MRI it would have been difficult to justify the increment in immunosuppressive therapy. Cardiac MRI therefore appears to be very useful in Churg Strauss Syndrome patients presenting with cardiac symptoms, particularly when other investigations are normal. This case also supports its use in monitoring for expected resolution after immunosuppressive therapy. The resolution of MRI abnormalities indicates they were due to myocardial inflammation, rather than fibrosis. The potential role of cardiac MRI as a screening tool for cardiac involvement in asymptomatic patients is less clear and requires further research. Nonetheless it could be argued that baseline imaging is useful given the high lifetime prevalence and mortality of cardiac disease in Churg Strauss Syndrome. It would allow for easier diagnosis in the event of the future development of symptoms.

This case also supports the therapeutic effectiveness of Cyclophosphamide in Churg Strauss Myocarditis. To our knowledge there have, as of yet, been no large scale trials to support its use. Its prescription is largely on an empirical basis but case reports have shown benefit. One of the largest studies of its kind to date showed a clinical or radiological improvement (demonstrated on cardiac MRI) in 6 of 8 patients with Myocarditis treated with Cyclophosphamide [8]. The patient in our case report showed radiological and clinical resolution of his Myocarditis with Cyclophosphamide. He developed cardiac disease despite long term steroid treatment and his symptoms did not improve with Myfortic therapy. Our case would therefore support the use of Cyclophosphamide in Churg Strauss Myocarditis above these other therapies.

4. Conclusion

In conclusion, we would advocate the use of cardiac MRI in Churg Strauss Syndrome Myocarditis; both as a sensitive diagnostic tool and as a means of monitoring response to immunosuppressive therapy. Our case also supports the therapeutic effectiveness of Cyclophos-

phamide in this condition.

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