A Case Report of Idiopathic Giant Cell Myocarditis

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Case Report

A 32 year old African American male was admitted to an outside hospital in July 2007 with symptoms of severe heart failure that required implantation of a short-term left ventricular assist device (LVAD). He was subsequently transferred to our facility due to worsening left ventricular heart failure, episodes of Torsades de Pointes, and monomorphic ventricular tachycardia. His device was replaced with a longer-term LVAD and he was discharged home in October. Fortunately, he underwent successful orthotopic heart transplant in November 2007.

Surgical pathology of the explanted heart revealed widespread cardiomyocyte necrosis, marked polymorphous inflammation, and giant cell formation consistent with idiopathic giant cell myocarditis (figures). Follow-up post transplant endomyocardial biopsies did not reveal recurrence of giant cell myocarditis or acute rejection.

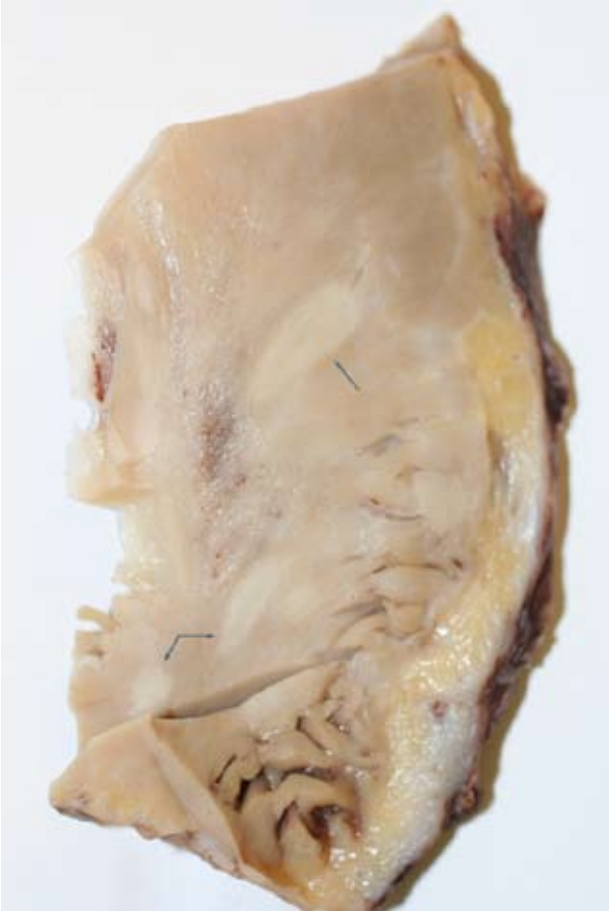

Figure 1. *Gross image of left ventricular myocardium showing nodular, ovoid tan-white areas (indicated by arrows) that microscopically corresponded to the areas of inflammatory cell infiltration and myocyte necrosis.*

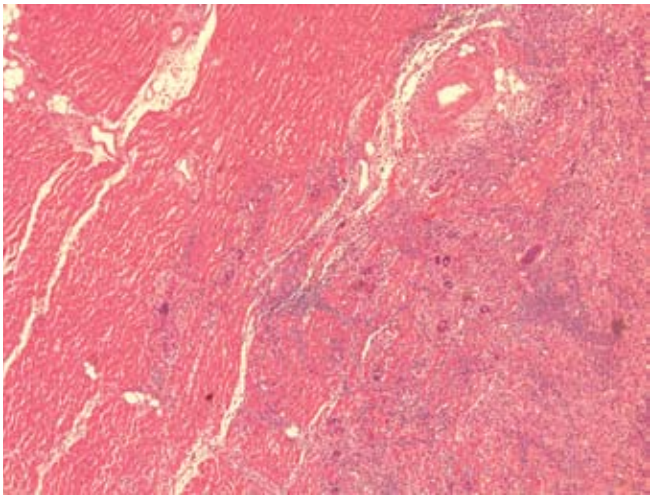

Figure 2. *Low power image (40x, H&E) showing clear delineation of involved areas with inflammatory cell infiltration, giant cells, and myocyte damage compared to relatively unaffected myocardium on the left.*

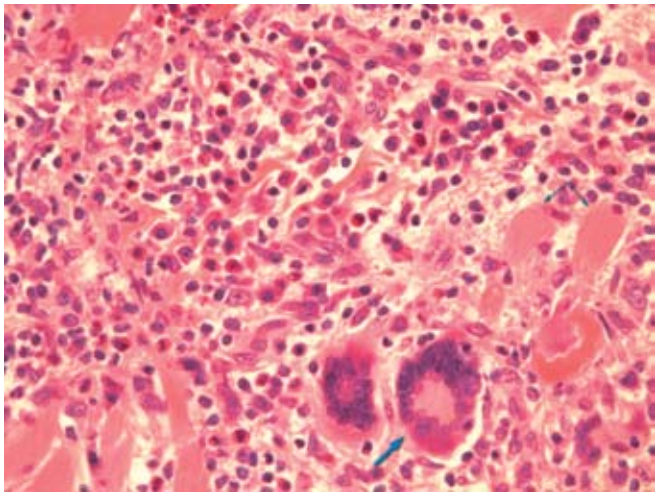

Figure 3. *High power image (400x, H&E) showing a mixed inflammatory cell infiltrate composed of eosinophils, plasma cells, small lymphocytes, and multinucleated giant cells. Prototypical multinucleated giant cell indicated by large thick arrow. Thinner arrow shows associated myocyte damage.*

Discussion

Idiopathic Giant Cell Myocarditis (IGCM) is a rare and generally fatal entity that has been largely confined to scattered case reports and observational studies, most notably a multi-center registry created by Cooper et al in 1997. While IGCM has historically been a histological diagnosis at autopsy, the current gold standard method of diagnosis remains endomyocardial biopsy. This

disorder typically afflicts younger adults with mean age of forty-three.[2] It is most commonly characterized by rapid and progressive congestive heart failure generally refractory to conventional heart failure treatments. Other common clinical presentations include ventricular arrhythmias, heart block, and less commonly as early symptoms of acute myocardial infarction.

While no uniform guidelines yet exist for the treatment of IGCM, animal models suggest involvement of CD4 T-lymphocytes[5,9] with reported success with combined steroid and immunosuppressive therapy.[2,3,7] Reported associations with autoimmune disorders are also consistent with this theory.[1,6]

Patients in the IGCM registry that received immunosuppression lived longer than those that did not.[2] Heart transplantation also appeared to be efficacious with 5-year survival data reported at 71% despite recurrence of disease in 20-42% of the newly transplanted hearts.[2,4,8] Our case of IGCM illustrates the importance of surveillance biopsies and the growing evidence that transplantation and immunosuppression may be effective therapy for an otherwise rapidly fatal disease.

References

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