Clinical Case Presentation

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Case history

- 26 years old Finnish female, healthy, no regular medication, very sportive
- Initial of symptoms in July 2013: physical condition getting worse, presyncope symptoms, mental stress as a mixing factor
- Contact to Occupational Health: ECG showed LBBB, ECHO was made: EF 55-60%, basal septum was abnormal thin; Holter showed some extrasystolia

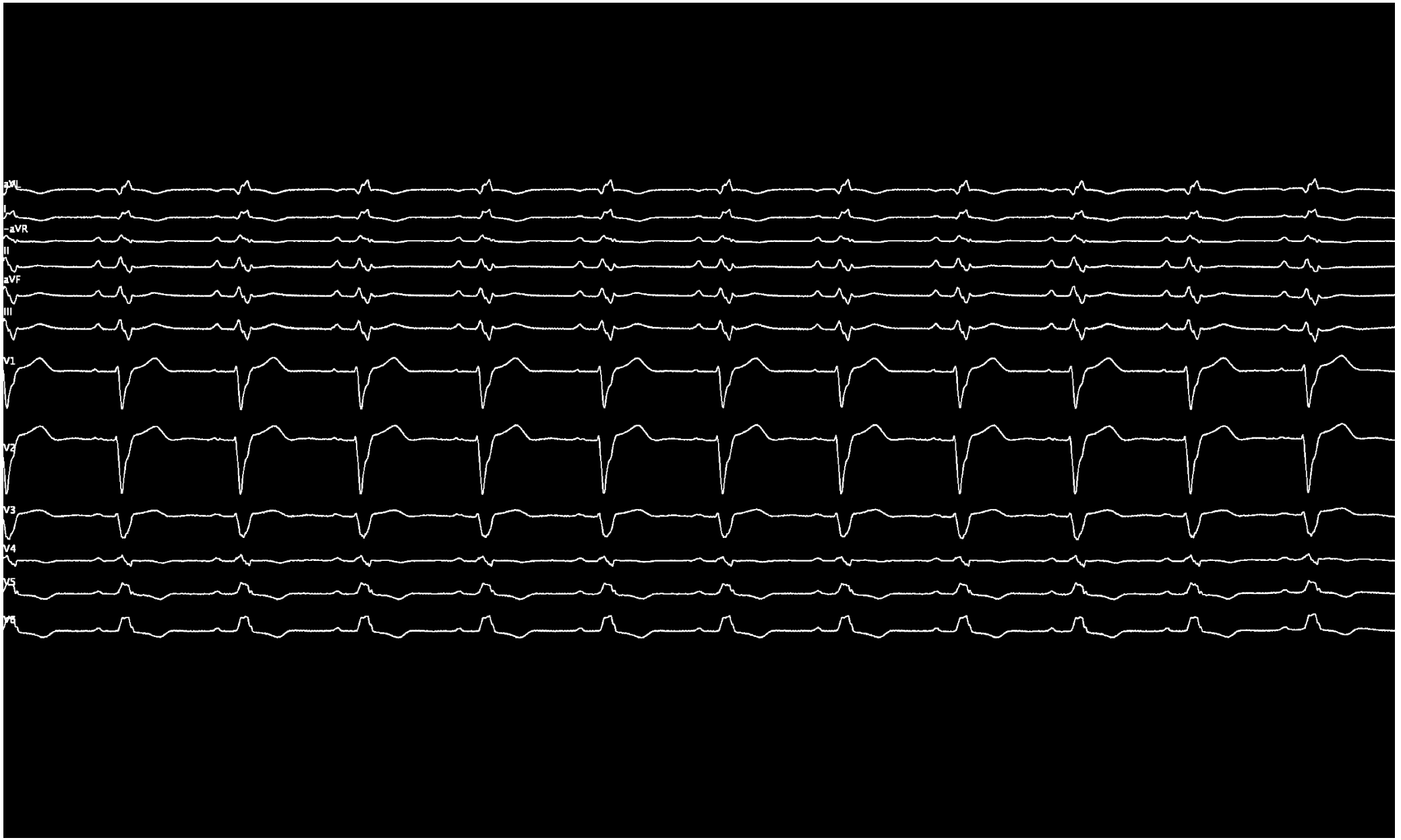
September 2013

- Transferred from home to hospital emergently: previous day after physical exercise started fast arythmia, then syncope
- In the emergency deparment of the central hospital: ECG showed ventricular tachycardia 185/min, which turned into sinus rhythm spontaneously before cardioversio was made
- Patient was sent to university hospital for further diagnostical tests 5 days later

Physical examination, ECHO etc.

- BP 101/68, HR 91, SaO2 100%, Tax 36.7
- Cardio-respiratory auscultation was normal
- Chest x-ray was normal
- Laboratory test results: Hb 137, Leuc 6.5, Tromb 254, La 2, CRP < 3, K 3.9, Na 141, Krea 73, Alb 45, TnI **0.10** (<0.04), BNP **205** (<100)
- **ECHO**: basal septum was hypokinetic and thin, LVEF 30% M-mode, global EF 40% by Simpson´s method, cardiac valves were normal
- Diagnostic suggestions?

ECG: LBBB



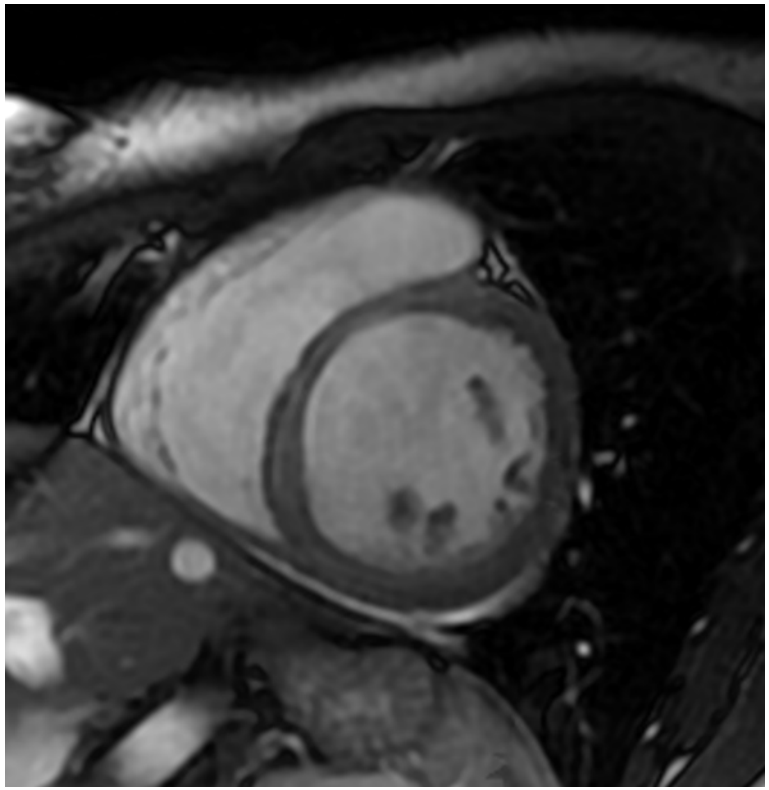
Differential diagnosis

- LBBB, ventricular tachycardia and basal septal findings in cardiac ECHO in young previously healthy patient ->

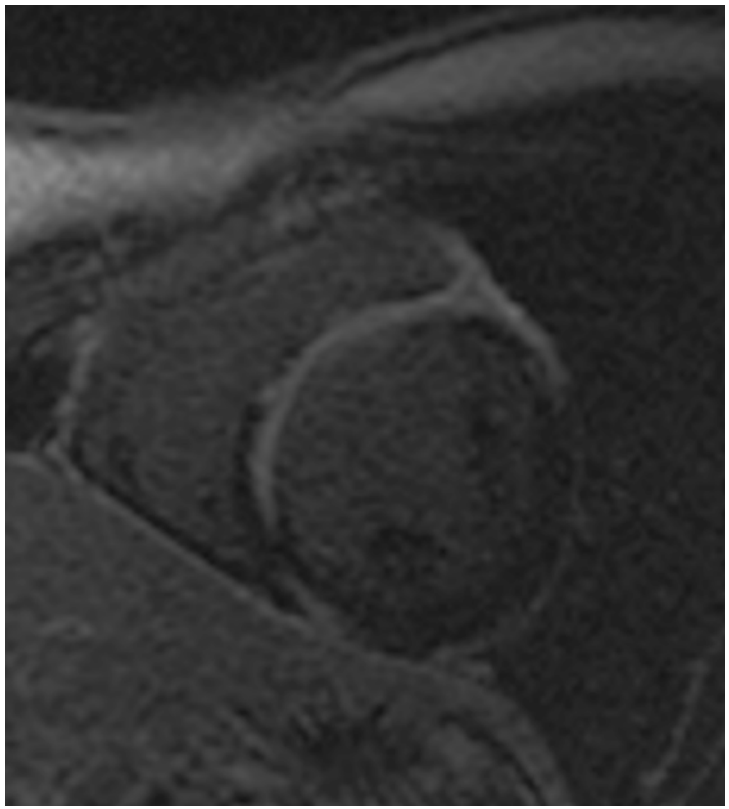
  -Viral myocarditis or inflammatory disease:
  Sarcoidosis/Giant-cell myocarditis?

- No history of previous respiratory infection
- Basal septal involvement seen in cardiac ECHO is typical for sarcoidosis
- Further diagnostic work-up?

Cardiac MRI



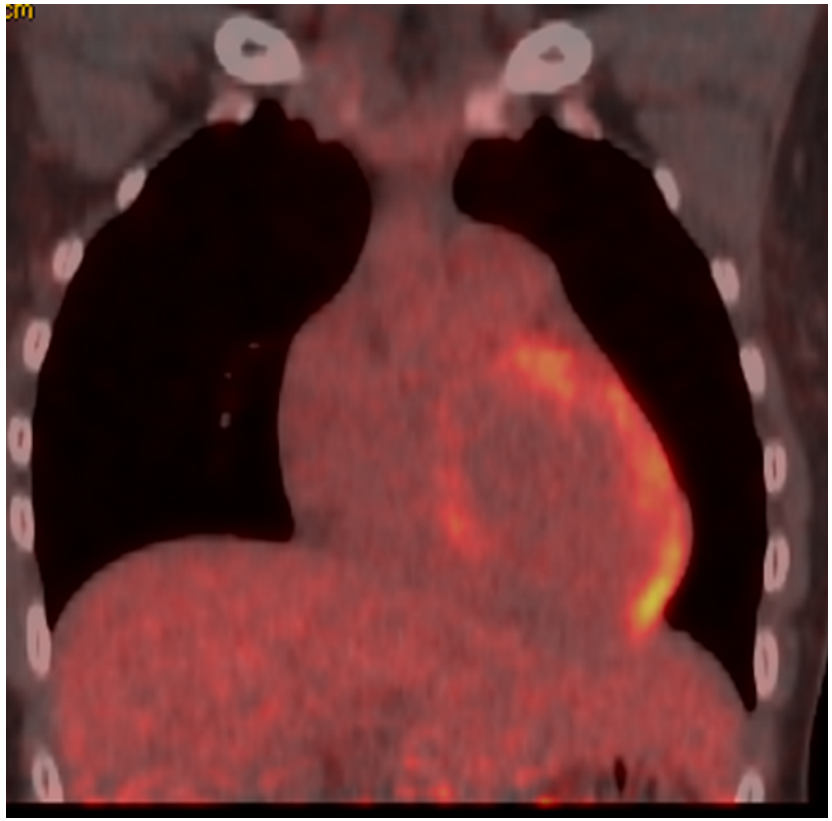
Thinned upper part of septum

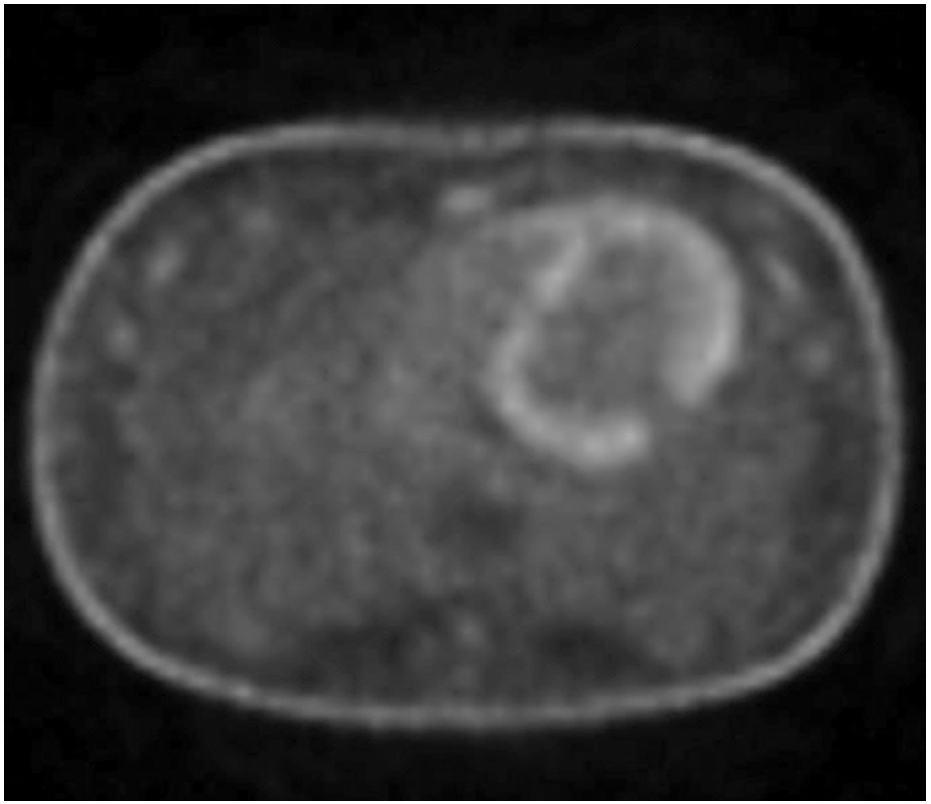


Wide abnormal bright effect in after phase picture

FDG-PET combined with a resting myocardial perfusion study

Increased FDG uptake in the myocardial perfusion defect areas, typical findings for inflammatoric myocardial disease





Endomyocardial biopsy

- Biopsies were taken from septum of the right ventricle (transvenous ultrasound guided endocardial biopsy by cardiologist), seven samples were taken
- Biopsy findings: eosinophilia, Giant cells as single and groups, partly normal and partly abnormal inflammatoric heart muscle. NO amyloid nor iron accumulations
- Patological diagnosis: **Giant-cell myocarditis**

Treatment

- The combined immunosuppressive treatment was started immediately after histological diagnosis (3 days after getting into the university hospital):
  - Prednisolon 60 mg x 1 p.o.
  - Atzatioprin 50 mg x 2 p.o.
  - Cyklosporin 100 mg x 2 p.o.
  - Others: Omepratzol 40 mg x 1, Calcium-D-vitamin 500mg/10ug 1 x 2, Ramipril 5 mg x 2, Bisoprolol 2,5 mg x 2, Spironolacton 12.5 mg x 1

Treatment and follow-up

- In the follow-up during the hospitalization there was no renewable ventricular tachycardias or fibrillations
- The ICD was installed because of the high risk of new ventricular arythmias
- The follow-up in the cardiological policlinic (the frequent controls):
  - 3 months after diagnosis the situation is stabile, there haven ´t been remarkable arythmias, the patient hasn´t had any dyspnea, is doing mild exercise like 2 km walkings, in control-ECHO EF is 40%, in left ventricle there is a large septal aneurysmatic scar area, Prednisolon dose has been reduced slowly (5mg/2 weeks)

Giant-cell myocarditis

- Rare, rapidly progressive and frequently fatal myocardial disease
- Patients are often young adults
- T lymphocyte mediated inflammation of heart muscle
- ≈ 20 % of patients has associated autoimmune disorders

Clinical presentation

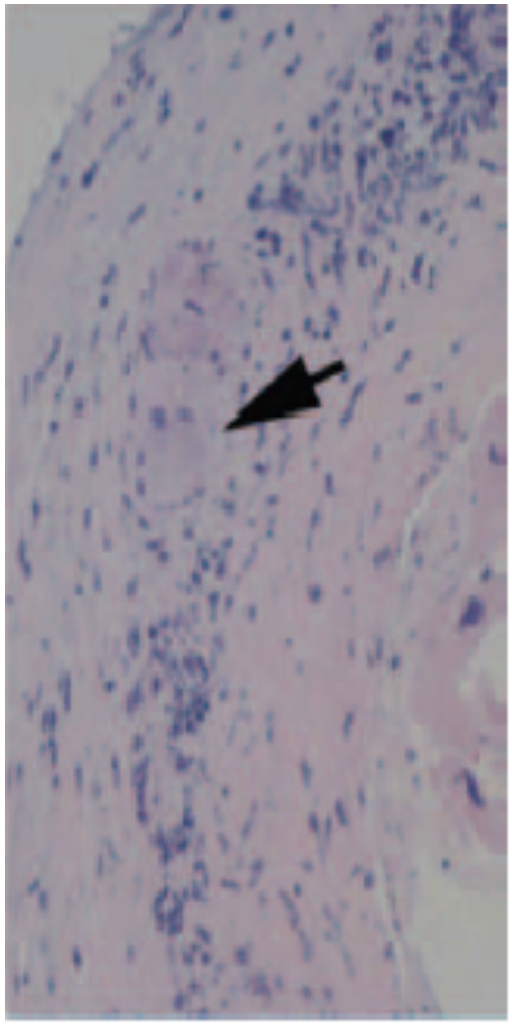
- Congestive heart failure
- Distal atrioventricular block
- Ventricular tachycardia
- Syndrome mimicking acute myocardial infarction

Diagnosis

- ECHO: EF < 50 % , locally thickened or thinned interventricular septum, dilated left ventricle
- Contrast enhanced cardiac MRI
- FDG-PET combined with resting myocardial perfusion imaging

Diagnosis

- Endomyocardial biopsy (repeated if necessary)
  - Inflammatory infiltrate including lymphocytes, histiocytes and multinucleated giant cells
  - Myocyte necrosis and eosinophils



Kandolin et al. Circ Heart Fail. 2013;6;15-22

Treatment

- Combined immunosuppression (steroid, azathioprine, cyclosporine)
- ICD
- Heart transplantation

Prognosis

- 2013 Kandolin & al: 26 patients with combined immunosuppression therapy
- Transplant free survival 77 % at 1 year and 63 % at 2 years

Kaplan-Meier curves for survival free of major adverse cardiac event (death, transplant) from the time of diagnosis in 26 giant-cell myocarditis (GCM) patients diagnosed by endomyocardial or surgical biopsies and treated with immunosuppressive medications.

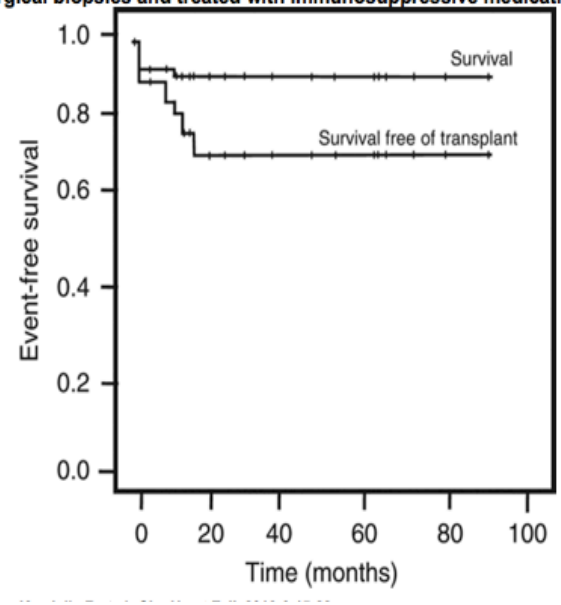


Table 1. Immunosuppressive Treatment of the 26 Patients With Biopsy-Diagnosed GCM

Corticosteroid + Azathioprine + Cyclosporine 17 (65%)
Corticosteroid + Azathioprine 4 (15%)
Corticosteroid + Azathioprine + Muromonab + Gammaglobulin 1 (4%)
Corticosteroid + Azathioprine + Myocophenolate mofetil 1 (4%)
Corticosteroid + Cyclosporine+ Myocophenolate mofetil 2 (8%)
Corticosteroid + Cyclosporine + Azathioprine/Methotrexate* 1 (4%)

GCM indicates Giant-cell myocarditis.
*Azathioprine was replaced with methotrexate after 3 wk of treatment because of pancreatic irritation.

Kandolin et al.Circ Heart Fail. 2013;6:15-22

Conlusions

- Consider diagnosis of giant cell myocarditis in patient with left ventricular failure of new onset who decline clinically despite usually care and with refractory ventricular tachycardia or atrioventricular block
- Use cardiac MRI and FDG-PET to identify and localize inflammatory processes
- Take repeated EMB
- Make the diagnosis quickly and start treatment with combined immunosuppression therapy