The Changing Role for Endomyocardial Biopsy in the Diagnosis of Giant-Cell Myocarditis

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Abbreviations

2D, Two-dimensional; AV, Atrioventricular; BNP, Brain natriuretic peptide; BP, Blood pressure; CBC, Complete blood count; CGD, Chronic granulomatous disease; CI, Cardiac index; cMRI, Cardiac magnetic resonance imaging; cTnI, Cardiac troponin I; EF, Ejection fraction; EKG, Electrocardiogram; EMB, Endomyocardial biopsy; EP, electrophysiology, GCM, Giant-cell myocarditis; HCTZ, Hydrochlorothiazide; HR, Heart rate; HTN, Hypertension; IC, Intercostal; ICD, Implantable cardioverter-defibrillator; INR, International normalized ratio; IVS, Interventricular septum; JVP, Jugular venous pressure; LV, left ventricular; LVEDP, Left ventricular end-diastolic pressure; mPAP, Mean pulmonary artery pressure; NM, Normal myocardium; PA, Pulmonary artery; PCWP, Pulmonary capillary wedge pressure; PMH, Past medical history; PMI, Point of maximal impulse; PTT, Partial thromboplastin time; RA, Right atrium; RR, Respiratory rate; RV, Right ventricular; VT, Ventricular tachycardia.

Case Presentation

- 50-year-old male
- Progressive shortness of breath
- Functional limitation for 2 months
- Dizziness, light-headedness, palpitations, chest tightness
- 24-hour Holter: sustained monomorphic VT 106 beats per minute

Case Presentation

- 50-year-old male admitted to a local hospital in Minnesota, USA
  - cTnI 3.66 ng/mL (reference <0.034 ng/mL)
  - BNP 5,500 pg/mL (reference 4–40 pg/mL)
  - 2D echo
    - Globally-depressed LV systolic function
    - Marked septal dyssynchrony
    - EF 25–30%
    - Moderately reduced RV systolic function
    - Bi-atrial enlargement

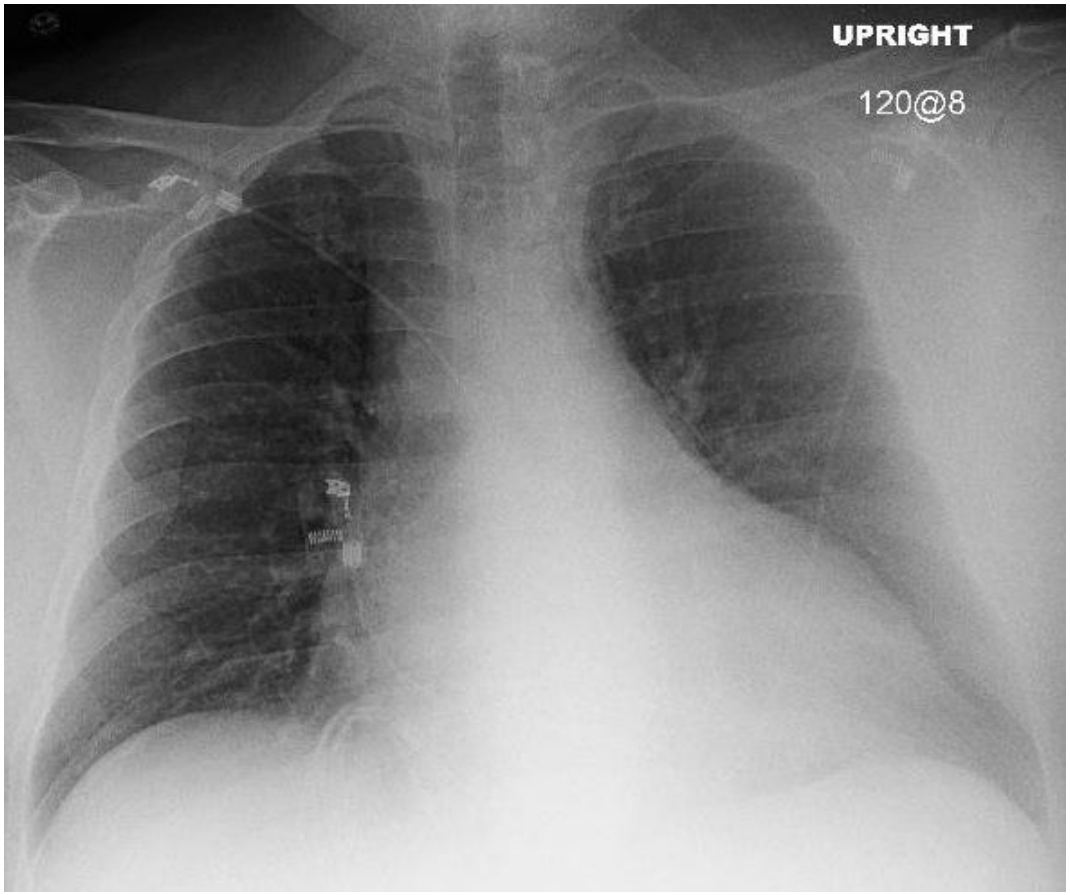
Case Presentation

- Initiated on intravenous heparin and amiodarone
- Transferred to our tertiary-care referral center
- PMH: HTN, hyperlipidemia
- Social history: Non-smoker; 1-2 beers/day; no illicit drugs
- Family history:
  - Brother with CGD deceased at 48 years (Staphylococcus sepsis)
  - Maternal grandparents deceased in early 40's (causes unknown)
- Never traveled outside the United States
- Medications: Atenolol, amlodipine, lisinopril-HCTZ, simvastatin

Case Presentation - Physical Examination

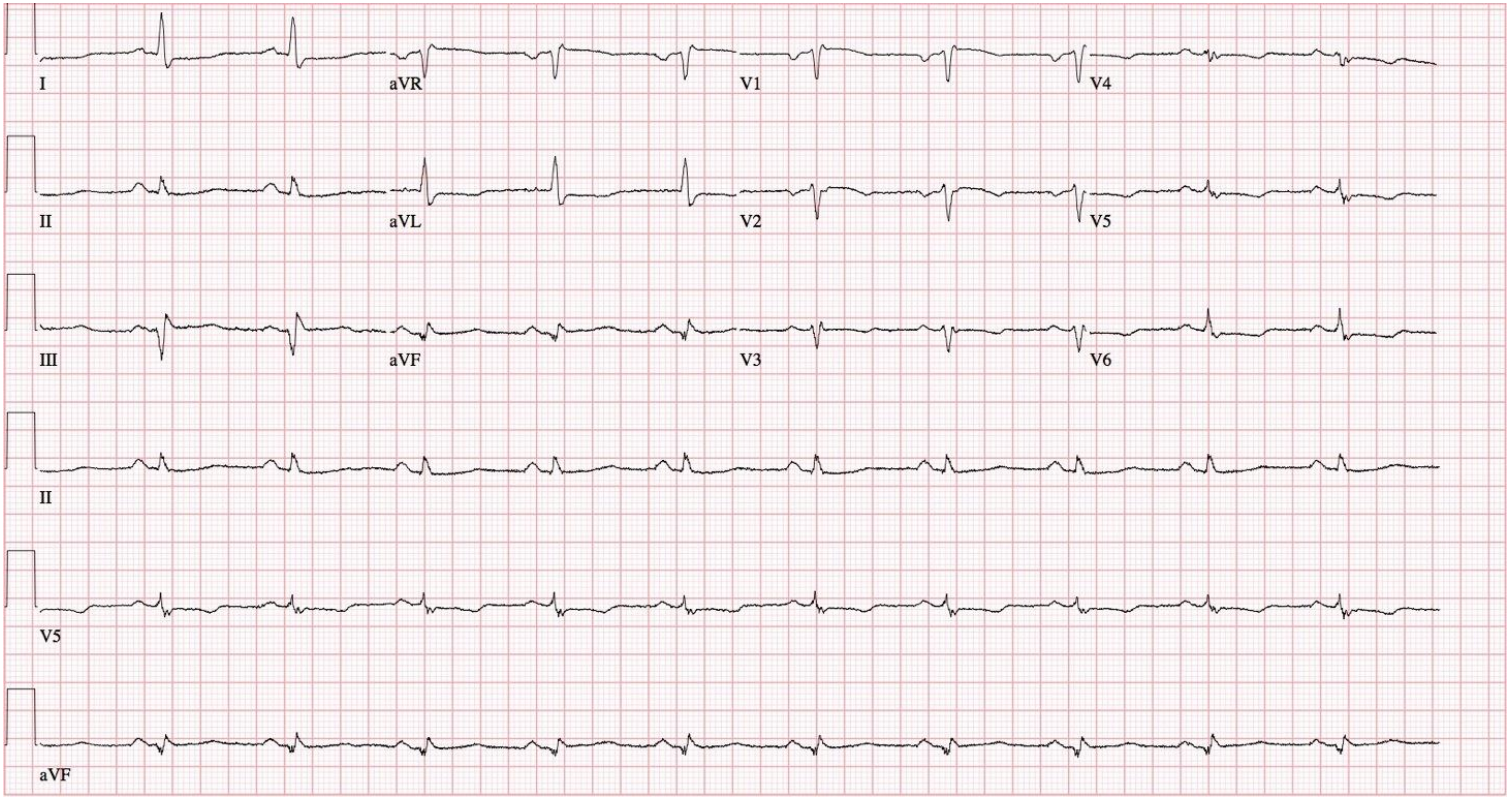
- Temperature: 37.3℃
- HR: 102/min (irregular)
- BP: 102/80 mmHg
- RR: 13/min, O_2 saturation: 97% (3 L/min inhalational oxygen via nasal cannula)
- Elevated JVP (15 cmH_2O)
- PMI 6th IC, 3 cm lateral to mid-clavicular line
- S3 gallop
- Bibasilar rales
- Hands and feet warm with intact peripheral pulses
- Trace peripheral edema

Case Presentation



Chest roentgenogram demonstrates mildly enlarged cardiac silhouette

Case Presentation



EKG shows normal sinus rhythm with low-voltage QRS complexes (134 ms), Q waves in leads III & aVF, poor R-wave progression and evidence of left atrial enlargement

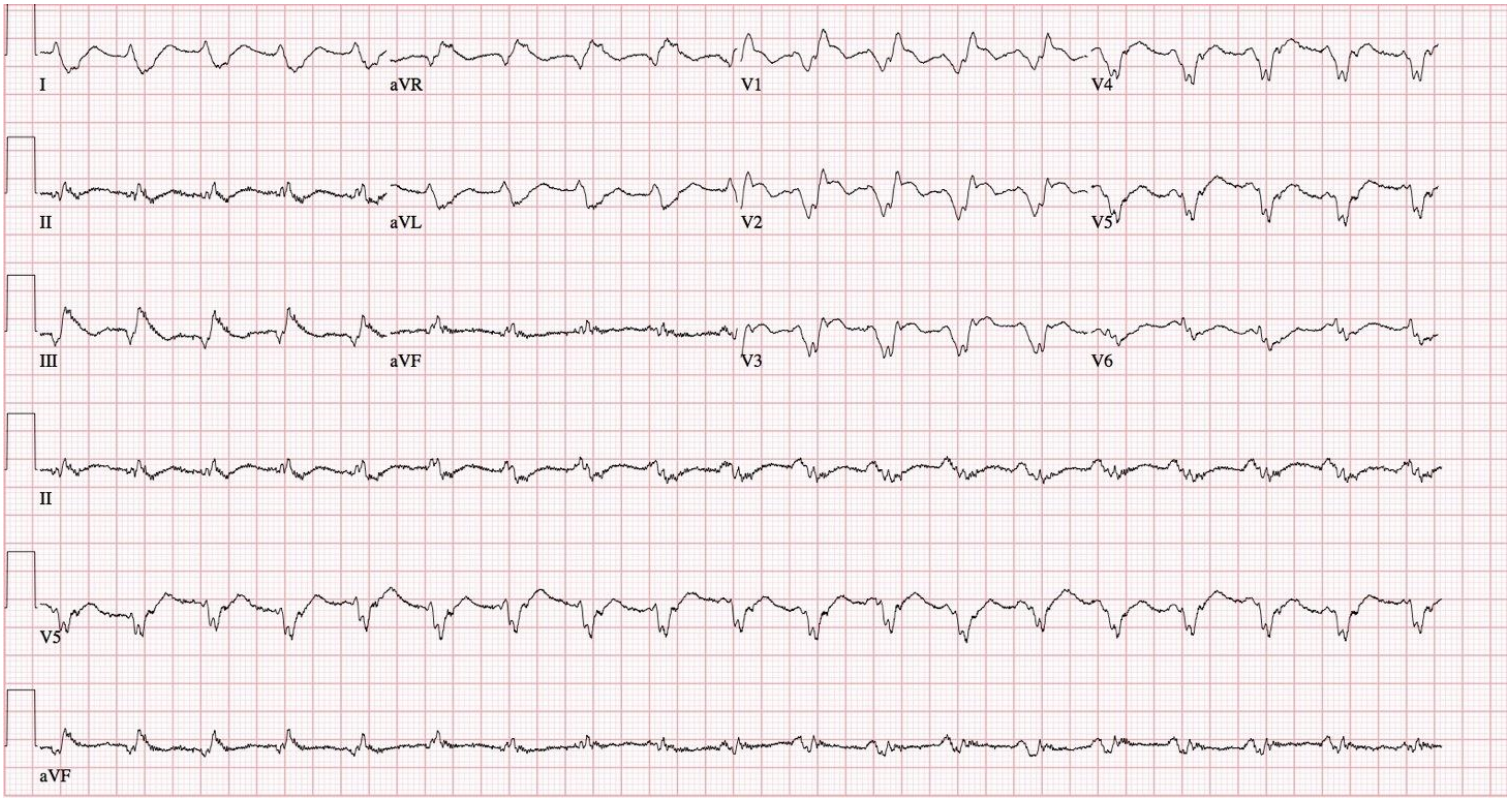
Case Presentation

- Normal CBC, basic metabolic panel, INR, PTT
- cTnI 3.66 > 3.351 > 2.915 ng/mL
- Normal coronary angiogram
- LVEDP 36 mmHg

Case Presentation

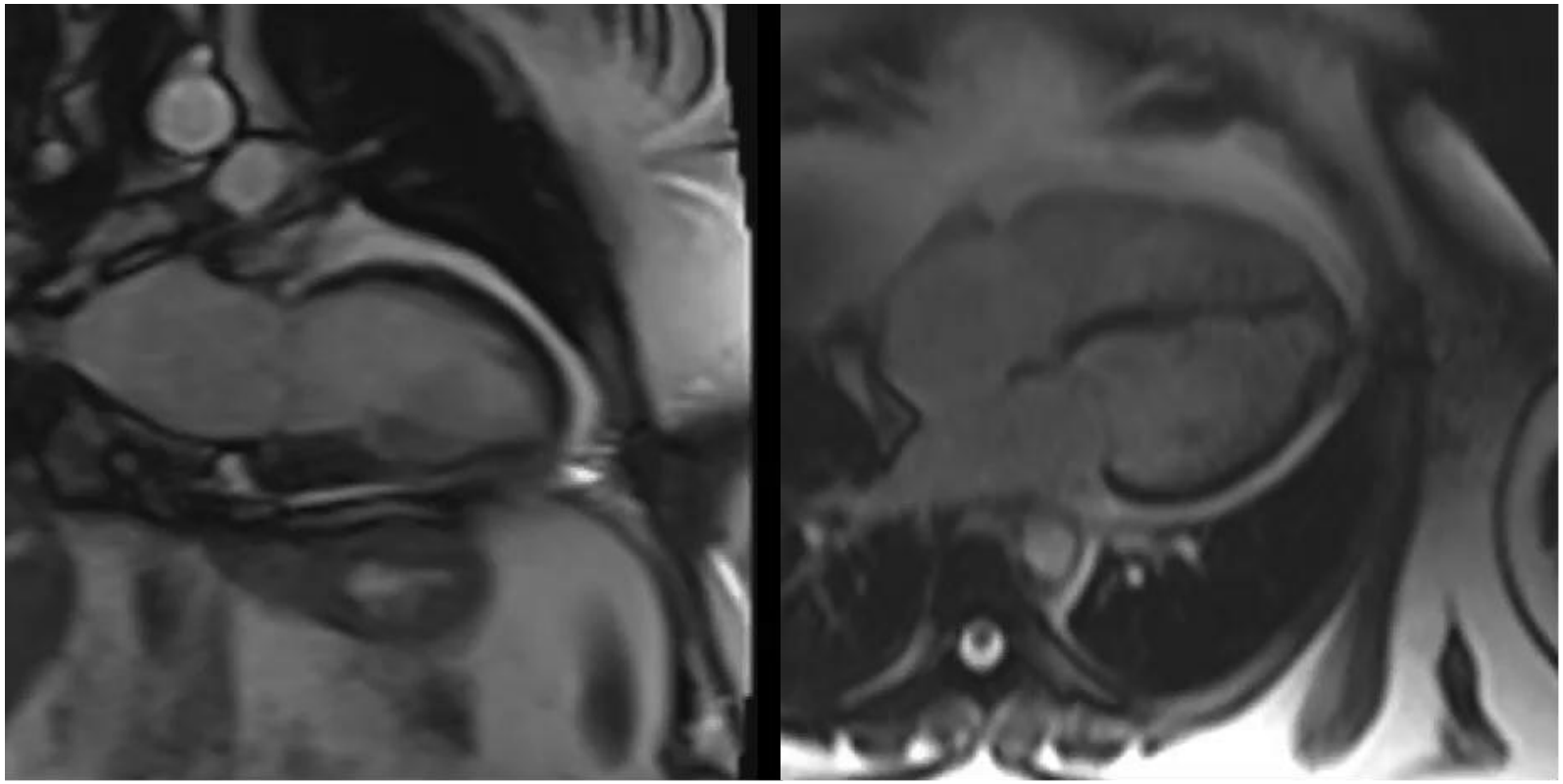
- RA 23 mmHg
- RV 33/20 mmHg
- PA 42/34 mmHg
- mPAP 38 mmHg
- PCWP 23 mmHg
- CI 1.4 L/min/m^2

Case Presentation



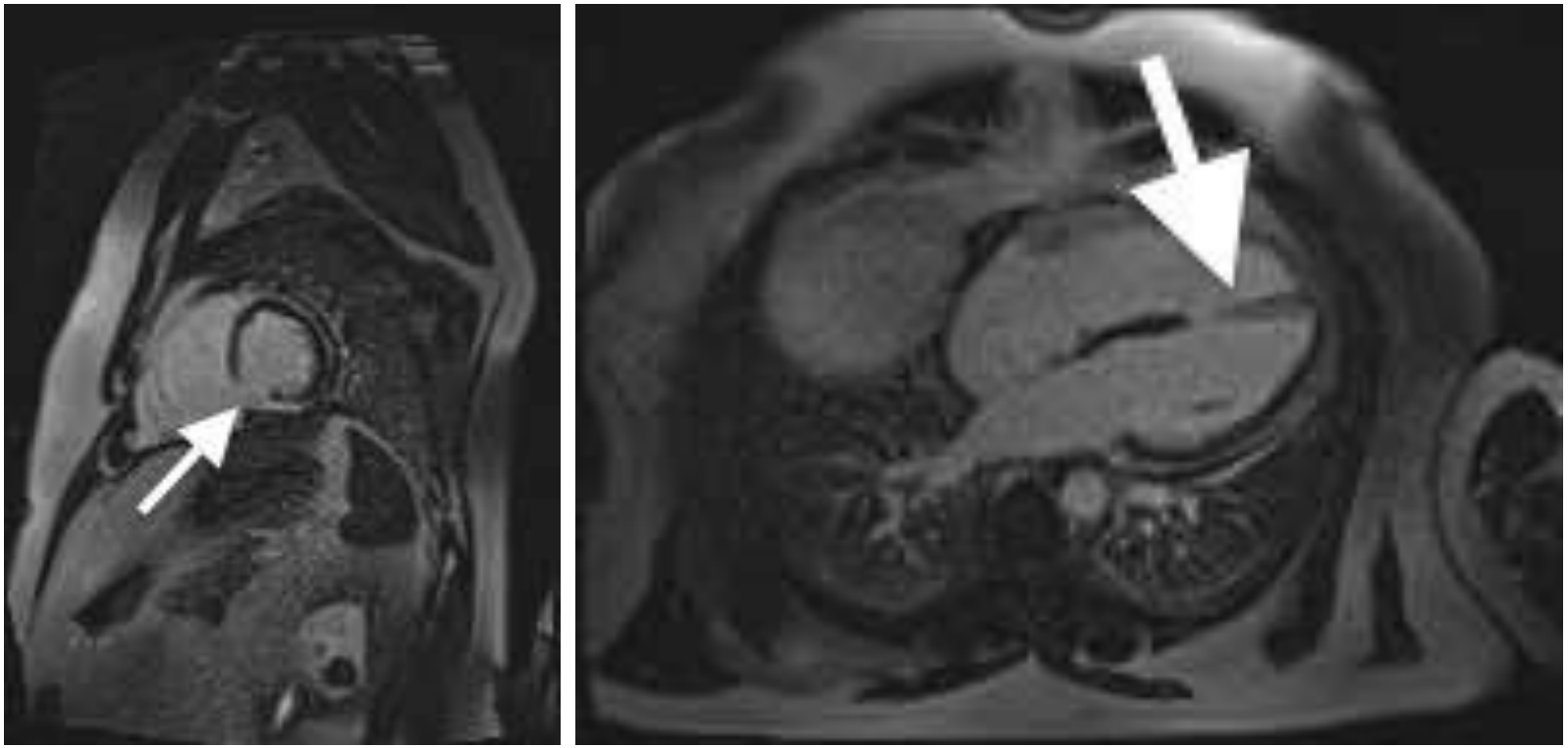
12-lead EKG during sustained slow monomorphic VT of right bundle branch block morphology (QRS = 194 ms)

Case Presentation



cMRI with gadolinium demonstrates mildly enlarged and severely hypokinetic left ventricle with prominent septal dyssynchrony

Case Presentation



cMRI with gadolinium demonstrates delayed hyperenhancement on T1-weighted imaging in the mid-inferior wall and mid-inferior septum

Case Presentation - Summary

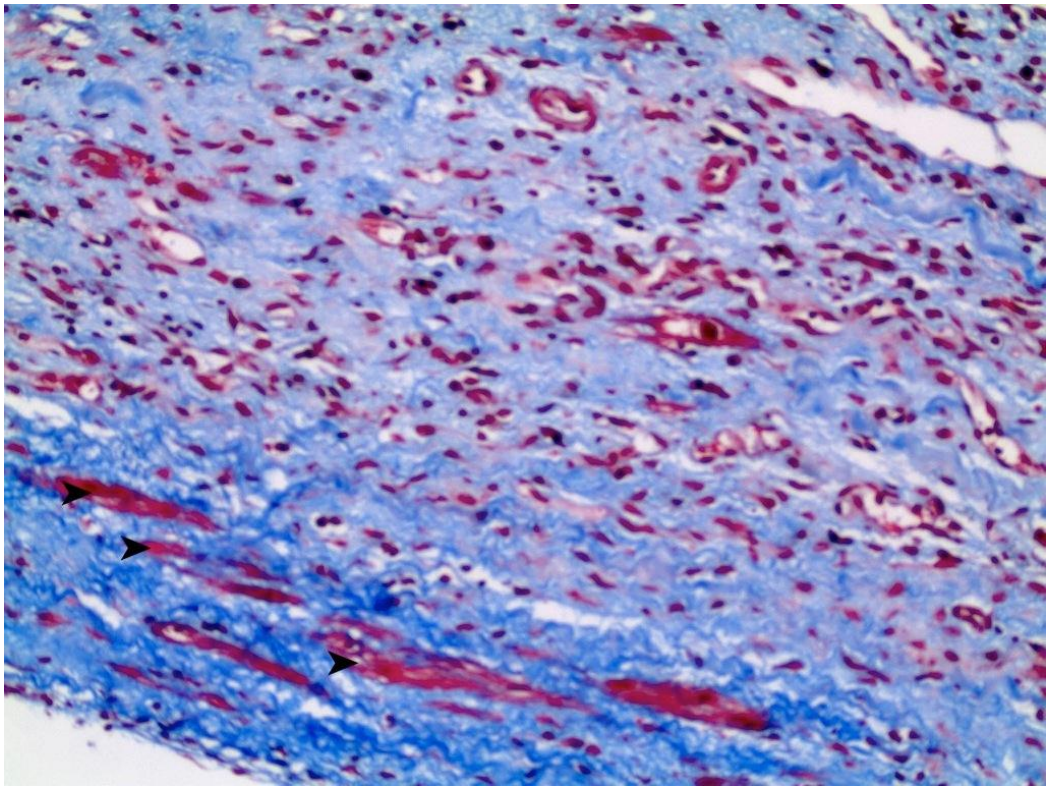
- Unexplained new-onset heart failure for 2 months
- Dilated left ventricle with regional wall-motion abnormalities
- New ventricular arrhythmia
- Hemodynamic compromise
- Pathologic Q waves
- Persistently elevated cTnI

Case Presentation

- What is the next best step?
  - EMB: Class I recommendation[2]
    - Unexplained, new-onset heart failure of <2-weeks duration with normal-size/dilated LV and hemodynamic compromise
    - Unexplained, new-onset heart failure of 2-weeks to 3-months duration with dilated LV and new ventricular arrhythmia or Mobitz type II or third-degree AV block

[2]Circulation. 2007; 116: 2216-2233.

Case Presentation



EMB (trichrome stain) from the RV septum demonstrating extensive fibrotic replacement (stained blue) with residual cardiac myocytes (arrowheads)

Case Presentation

- EP study/ablation:
  - Multiple inducible VT at variable cycle lengths
- Dual chamber ICD placement

Case Presentation

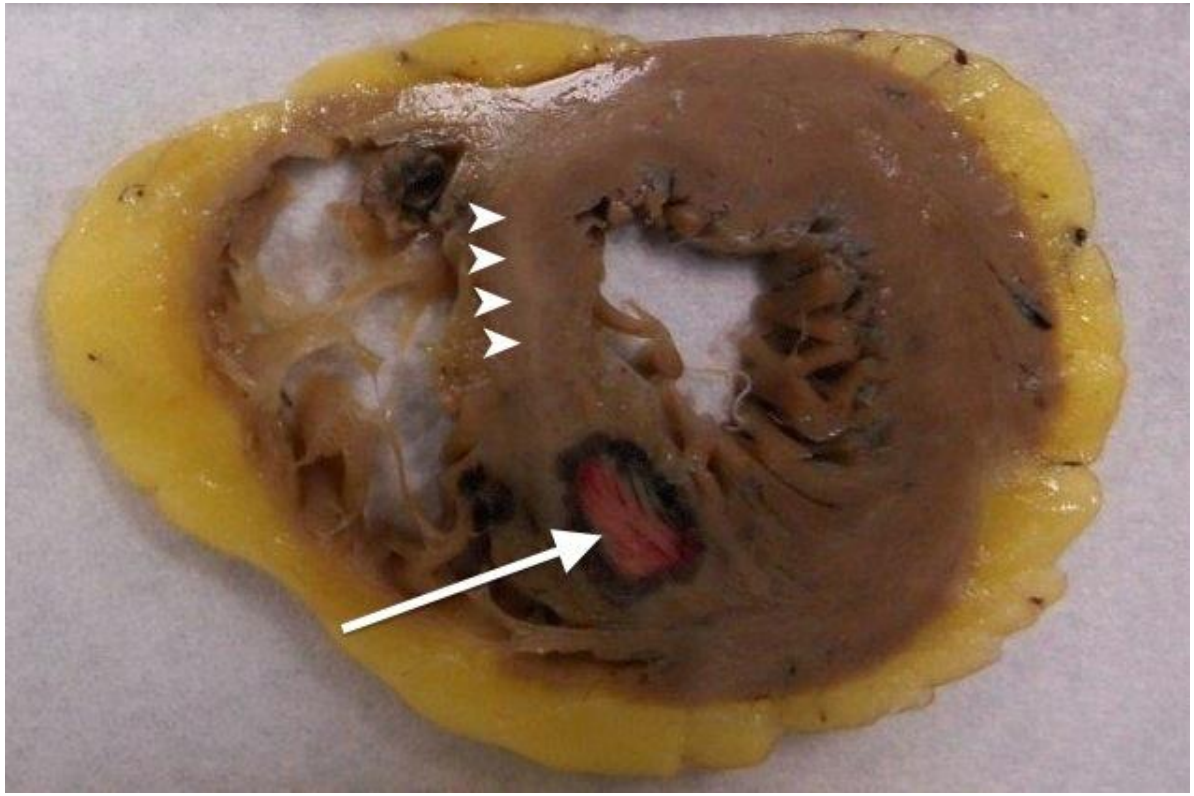
- Received immunosuppression with methylprednisolone & mycophenolate mofetil
- Planned for emergent 1A cardiac transplantation
- Orthotopic heart transplantation

[2]Circulation. 2007; 116: 2216-2233.

Case Presentation

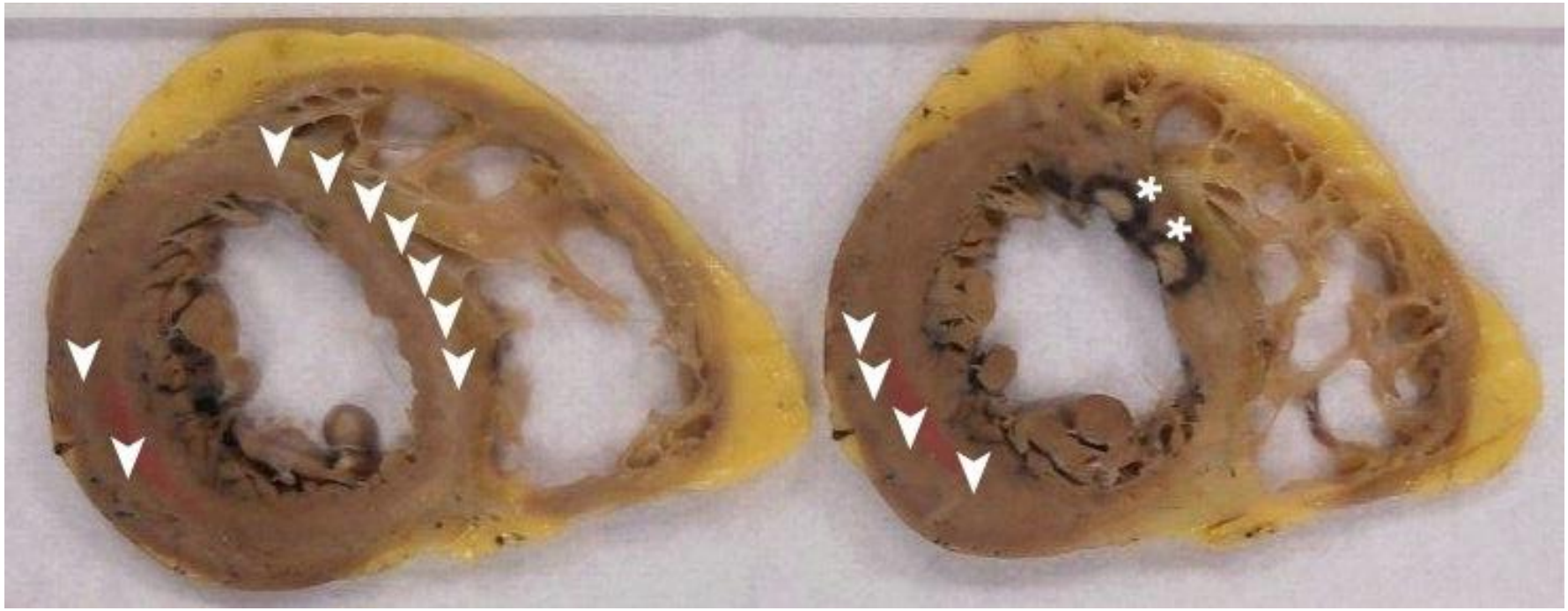
Extensive replacement fibrosis of the IVS (arrowheads) with areas of old hemorrhage (asterisk)

Case Presentation



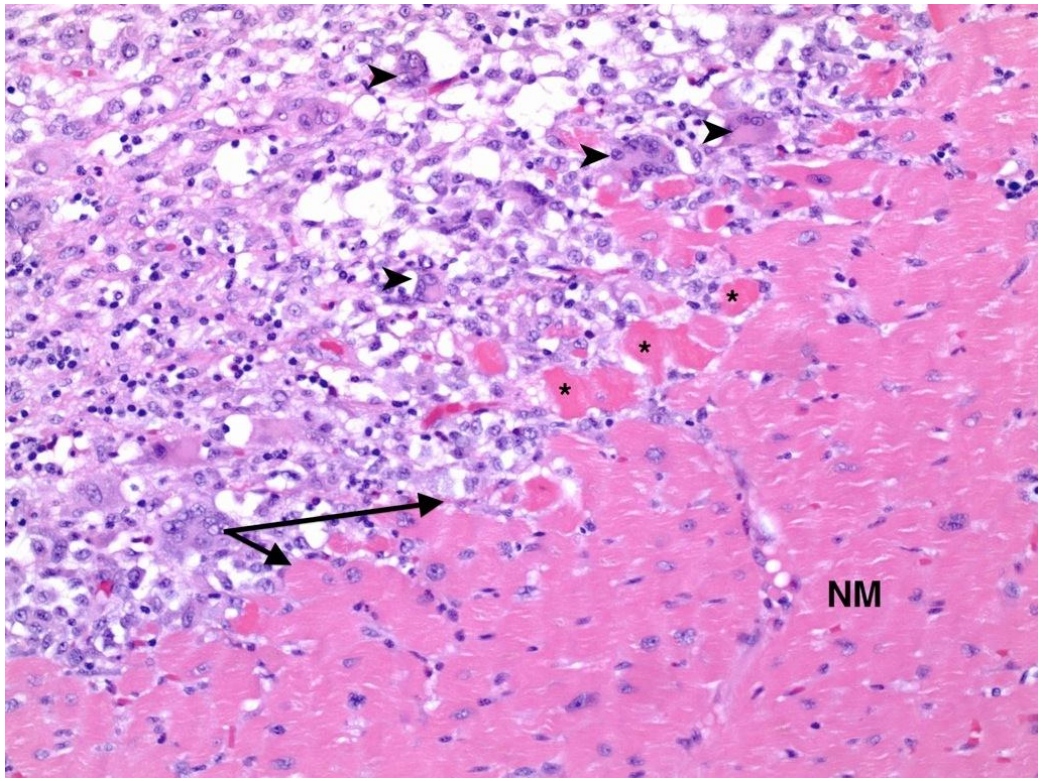
Extensive replacement fibrosis of the IVS (arrowheads); there is acute hemorrhagic infarction of the IVS (arrow)

Case Presentation



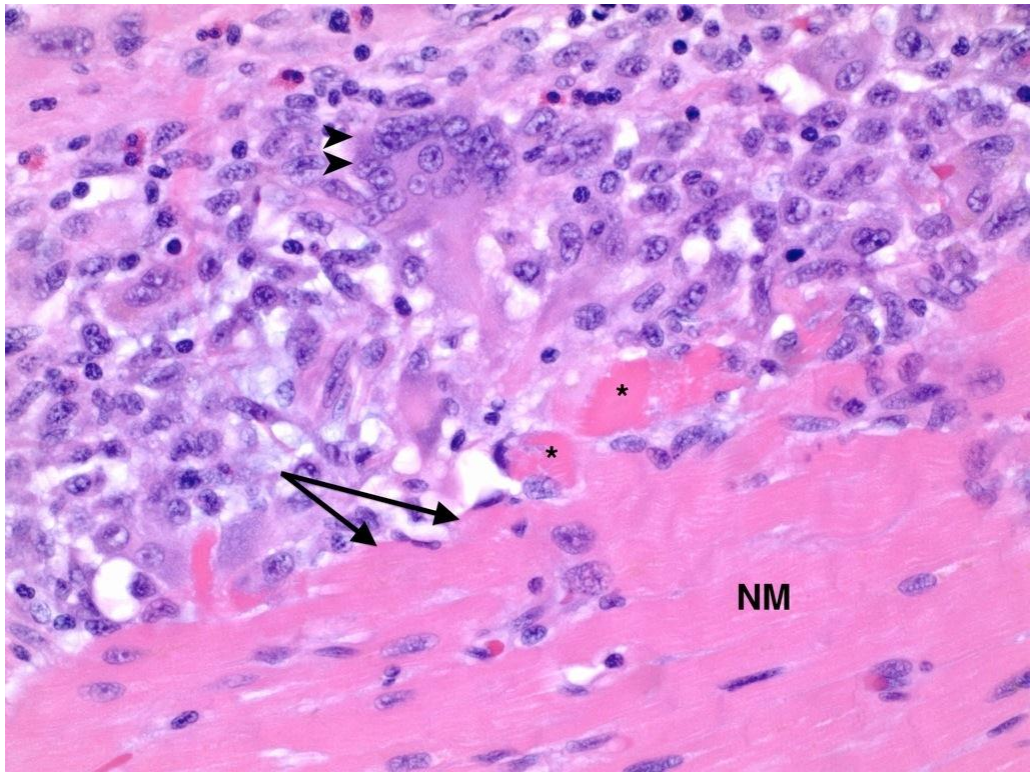
Extensive replacement fibrosis with involvement of the LV free wall in addition to the IVS (arrowheads) with areas of old hemorrhage (asterisk)

Case Presentation



Interface (arrows) between active GCM and viable NM with hypereosinophilic and necrotic cardiac myocytes (asterisk). Multi-nucleated giant cells (arrowheads) are seen invading into NM

Case Presentation



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Conflict of interest. Ankur Kalra, Rachel Kneeland, Michael A. Samara, and Leslie T. Cooper Jr declare no conflict of interest.

Compliance with ethics guidelines. Informed consent was obtained from this patient for being included in the paper.

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