Expect the Unexpected: A Case Report of Giant Cell Myocarditis

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Case Introduction

- 66-year-old female with pertinent history of MI and active tobacco smoking.
- Presented to PCP with CC of SOB on exertion and worsening epigastric pain.
- Initial treatment plan: albuterol for SOB, omeprazole trial for GERD, outpatient referrals for EKG, imaging for CAD workup, and labs including CBC with differential and CMP.
- Sought further care at ED due to persistent symptoms and was ultimately diagnosed with NSTEMI and new worsening systolic heart failure.
- Presentation complicated by cardiogenic shock requiring inotropic and vasopressor support with intra-aortic balloon pump (IABP) placement.
- Cardiac biopsy confirmed a diagnosis of Giant Cell Myocarditis (GCM).

Hospital Course

- Vital signs notable for HR 114 BPM.
- Unremarkable pulmonary and neurologic exams, S4 with no JVD, palpable distal pulses and nonedematous LEs.
- Arrived on IV milrinone 0.375 mcg/kg/min, IV furosemide 20 mg/hr, IV vasopressin 0.03 units/hr, and heparin drip.
- Swan-Ganz catheter measurements: CI 1.7L/min, CVP 16mmHg, PAP 51/31mmHg.
- PO prednisone 40mg BID and CellCept 1000mg BID were initiated for GCM treatment.
- Impella 5.5 device placement and IABP removal with successful weaning of inotropic support on HD 3.
- Repeat echo 3 weeks later: EF 35%-40%, demonstrating cardiac recovery.
- Weaned from Impella support over the following week.
- Repeat echo: EF 45% with mild anteroseptal and anterior wall hypokinesis.
- After one-month hospital course, discharged on CellCept 1000mg BID, prednisone taper, Entresto 24mg-26mg BID, empagliflozin 10mg daily with outpatient cardiology follow-up.

Discussion

- GCM is a rare, rapidly fatal disease most often affecting previously healthy young to middle-aged populations.
- Etiology thought to be related to immune dysregulation, specifically T-cell dysfunction.
  - Inflammatory cytokines, (i.e. IL-2, interferon-gamma) result in giant cell characteristic.
- Associated with autoimmune conditions like IBD, Hashimoto's, tumors of immune cells (i.e. thymoma and lymphoma[1]).
- Potential association with checkpoint inhibitor immunotherapy.[2]
- Study of 63 GCM patients, 19% had prior diagnosis of AI disorder (Hashimoto's, RA, myasthenia gravis[3]).
- Early symptoms include CHF, arrhythmias, cardiac conduction abnormalities.
- Usually rapidly progresses to cardiogenic shock and/or chronic HF.
- Myocardial biopsy remains gold standard for diagnosis.
  - Myocyte necrosis + multinucleated giant cells.
- Early detection and accurate diagnosis significantly impact treatment options and prognosis.
- Current standard of care is immunosuppressive therapy.
  - Corticosteroid + immunomodulatory drug.
- Transplant-free 5-year survival increased if immunosuppression is continued long term.

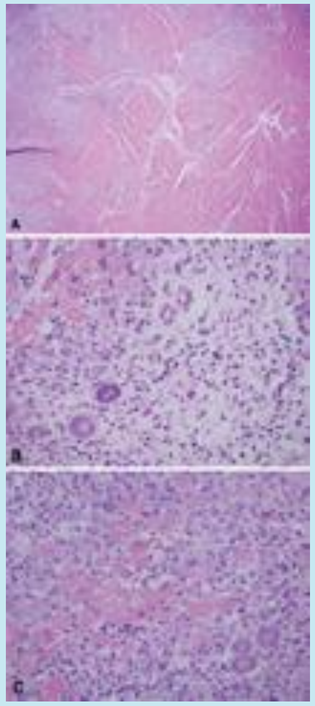

Figure Legend[1]: Morphologic features of giant cell myocarditis. A, Multifocal serpiginous inflammatory infiltrate. B, The infiltrate is composed of lymphocytes, eosinophils, plasma cells, and prominent multinucleated giant cells. Discrete granulomas are not evident. C, There is extensive myocyte necrosis associated with giant cell infiltrate (hematoxylin-eosin, original magnifications ×4 [A] and ×40 [B and C]).

Arch Pathol Lab Med. 2016;140(12):1429-1434. doi:10.5858/arpa.2016-0068-RS

- Mechanical circulatory (MC) support (IABP, ECMO) utilized if failed immunosuppression or hemodynamic instability.
- MC support + immunosuppression shown to increase survival.[2]
- Cardiac transplant is often ultimate management in GCM.
- Studies show poor prognosis without transplant.
  - Death of 31% of patients with 69% surviving after undergoing successful heart transplants.[4]
  - 63 patients with GCM: 89% died or underwent cardiac transplant.[3]

Conclusions

- Despite immunosuppression, rare for patient survival without transplant.
- Even with transplant, GCM can reoccur in up to 25% of patients.
- Highlights importance of aggressive immunosuppression and routine follow-up.[2]

References

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