

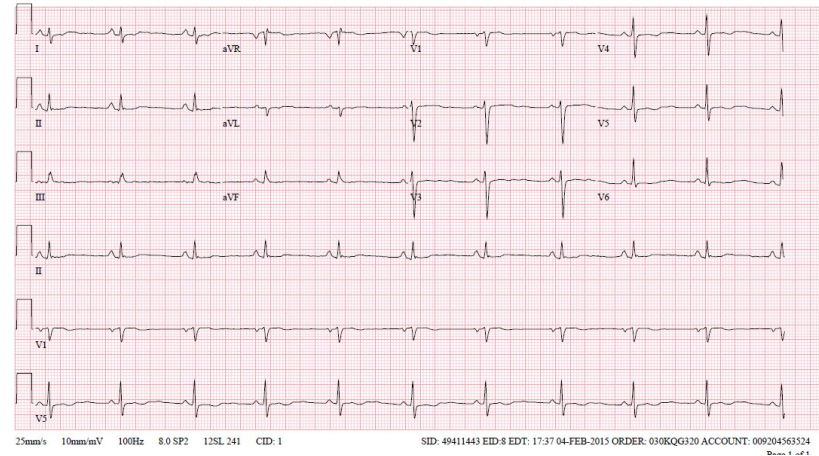
Puzzling Cases in Cardiac Function: Read with the Experts January 17, 2022

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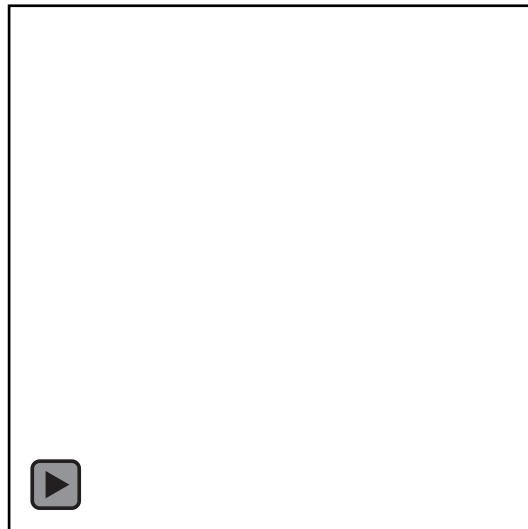
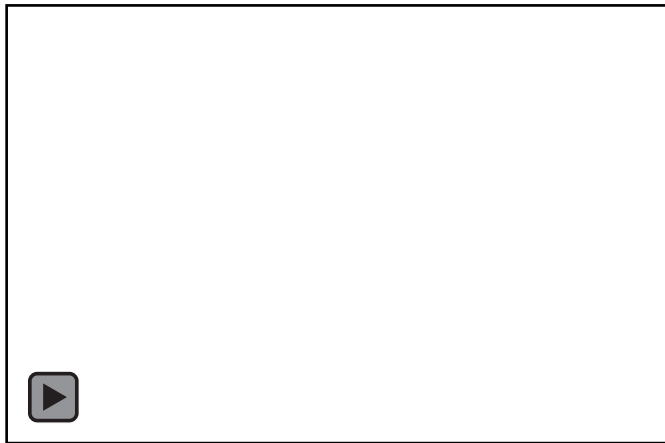


31 yo previously healthy woman presented to the ER with syncope while taking a shower.

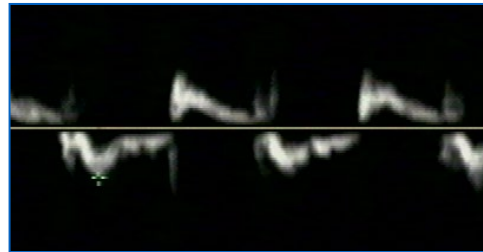
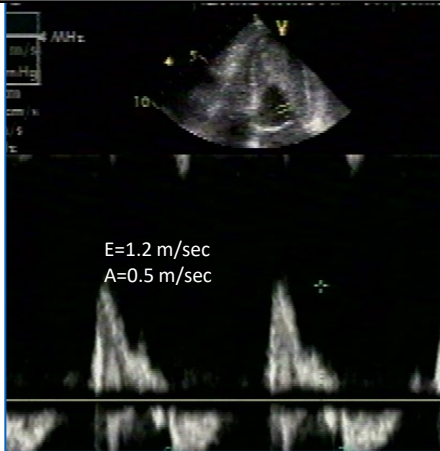
- 4-week history of headache, pallor and dyspnea with orthopnea.
- NT Pro-BNP 22,000 pg/ml; troponin 0.435 ng/ml
- CXR – pulmonary vascular congestion, bilateral pleural effusions c/w mild CHF
- ECG – NSR, nonspecific T wave changes



Echocardiogram



- Thick, hyperechoic, homogeneous material layering on the endocardium, extending from mid-ventricle to the apex; filling distal half of the ventricle
- Hyperdynamic base to mid segments of all walls with severe HK of distal segments; LVEF 35%
- Normal RV size w/ moderate RV dysfunction. Homogeneous layering of the material in the RV apex
- Diastolic dysfunction with elevated LV filling pressure



Mitral E/A = 2.4
E' septal = 5.1 cm/sec
E/E' = 24

Which test would be most helpful to make the diagnosis?

- A. Cardiac MRI**
- B. TEE**
- C. CBC with differential**
- D. Head CT**



CBC: WBC $15.7 \times 10^9/L$: 51% eos
Absolute eosinophil count 8.0×10^9

What is the most likely diagnosis?

- A. Primary vs. secondary cardiac tumor**
- B. Hypereosinophilic endomyocardial disease/Loeffler endocarditis**
- C. Apical hypertrophic cardiomyopathy**
- D. Chaga's disease**



Hypereosinophilic Syndromes

- **Definition:**
 - Persistent eosinophilia with ≥ 1500 eosinophils/mm³ for at least 6 months with evidence of organ involvement
- **Etiologies include:**
 - Primary (or neoplastic - underlying stem cell, myeloid or eosinophilic neoplasm)
 - Secondary (or reactive - parasitic infections, allergic disorders, drug hypersensitivity, infection, immunologic disorders)
 - Idiopathic – unknown despite thorough etiologic work-up
- **Common target organs include the skin, lung, GI tract, cardiovascular system and brain.**

Pathogenesis

Hypereosinophilia due to primary, secondary or idiopathic etiologies



Overproduction of cytotoxic eosinophils



Degranulation of eosinophilic granules



Heart damage evolves thru 3 stages



Acute pericarditis, myocarditis or endocarditis



Necrotic phase



Formation of intramural thrombi adjacent to the injured endocardium



Thrombotic phase



Localized or extensive replacement fibrosis

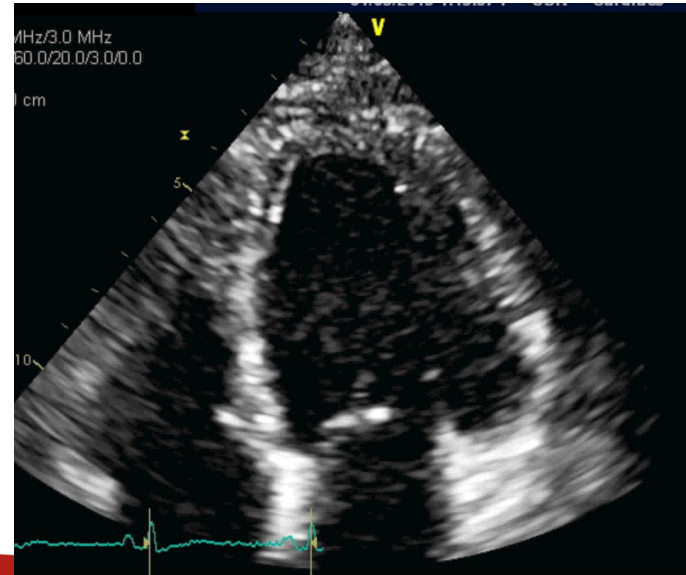
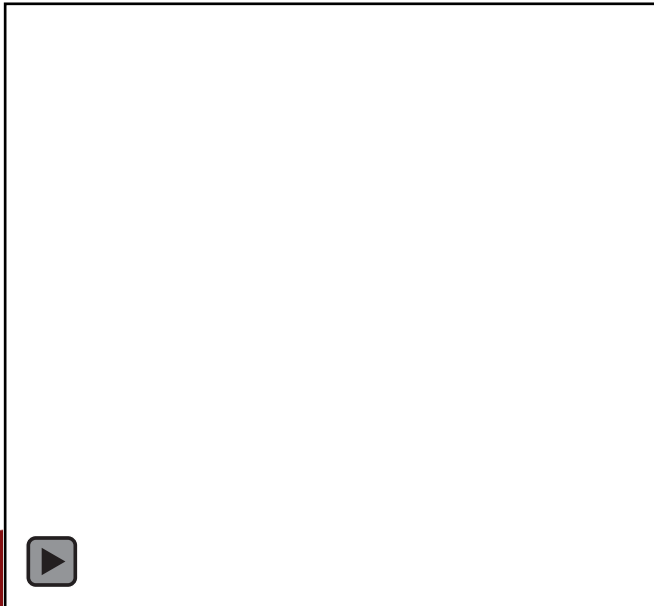


Fibrotic phase

Treatment

- **High dose steroid**
- **Hydroxurea**
- **Imatinib**
- **Anticoagulation if thrombus present**

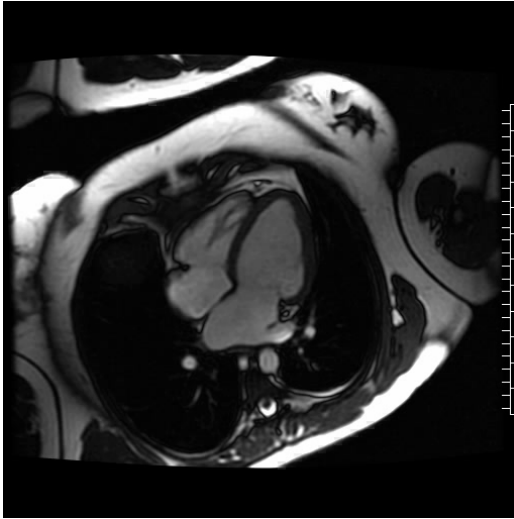
- Pt diagnosed with idiopathic HES
- After 6 mos of treatment w/ steroid, hydroxurea, lovenox, imatinib:
 - Normalizing of eosinophil count
 - Improvement of symptoms
 - Repeat echo - marked improvement of LVEF; mild thickening of the apical anterior wall and apex; resolution of thrombus



Pre-TX

Post- TX

Cardiac MRI post - treatment



- Normal LV size and systolic function. No residual LV thrombus
- Subendocardial hyperenhancement involving the mid-distal anterior wall, mid anteroseptum, distal septum and apex suggestive of a low degree of fibrotic changes

Thank You!