Idiopathic Constrictive Pericarditis

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• JP is a 61 year old white male seen at BRMC on 8/25/09 for complaints of chest pain radiating to the neck and dyspnea.
• NKA
• Medications: metformin, crestor
• Past Medical History: Type 2 DM, HTN, Hyperlipidemia

• Past Surgical History: Left hand reconstructive surgery, repair of a crush injury of the right knee and thigh, left tibia ORIF, and lumbar spine surgery
• Family History: Dad died of MI age 77, Mom died of MI age 64
• Social History: no alcohol, quit tobacco 35 years ago, works as a farmer
• Review of Systems: otherwise negative

• Physical Examination: Temp 96.7, pulse 87, Blood Pressure 106/70 RR 16
• HEENT: normal
• Chest: clear
• Cardiovascular: normal S1 and S2, no rubs, gallops, or murmurs
• Abdomen: normal

• Extremities: 1+ lower extremity edema
• Neurologic: mildly sedated, no lateralizing findings

• Troponin I < 0.05
• Na 134, K 4.0, glucose 248, creatinine 1.09
• Hgb 14.5, WBC 14.4
• CRP 6.23
• Cholesterol 166, HDL 46, Triglycerides 139 LDL 92
• BNP 14.9
EKG
- Mild sinus tachycardia
- T wave flattening in the inferior leads

CXR PA and Lateral
- Atelectasis in the lung bases

Lexiscan Stress Test
- Chest pain occurred
- EKG: no evidence of ischemia or arrhythmias
- Ejection Fraction 59%
- No perfusion defects noted

- The patient was discharged from BRMC to follow up with his PCP
• On 11/05/09 the patient presented to HVHMC with a chief complaint of chest pain and dyspnea
• HPI: The patient had progressive exertional dyspnea, dry cough, 3 pillow orthopnea and a 20 pound weight gain over one to two weeks

• Physical Examination: Temp. 98, BP 133/76, pulse 97, RR 21, poxim 96% on 2 liters O2
• HEENT: JVD
• Chest: decreased breath sounds at the lung bases
• Cardiovascular: decreased heart tones
• Abdomen: normal
• Extremities: 1+ UE and 3+ LE edema

Laboratory
• Na 136, K 4.2, BUN 18, creatinine 1.34 glucose 123
• Hgb 13.5, WBC 7.3
• Tl 0.05
• D-dimer 2.37
• BNP 220

EKG
• Low voltage
• Non specific T wave abnormalities
CXR PA and Lateral
• Bilateral pleural effusions

CT Scan of the Chest
• No PE
• Large right and moderate left pleural effusions
• Small pericardial effusion
• Right lower lobe atelectasis
• Ascending aorta 4.9 cm

Chest MRA
• 4.9 aneurysmal dilatation of the ascending thoracic aorta without dissection
• 2.7 proximal descending thoracic aorta

Echocardiogram (TTE)
• EF 55%
• RV: normal
• Trace TR
• Thickened, calcified, bicuspid aortic valve
• Normal pericardium
Left and Right Heart Catheterization
Hemodynamics

- Cardiac output 2.4
- Cardiac Index 1.1
- RA 17
- RV 38/14/20
- PA 34/20/26
- PCW 19
- LV 107/15/25

Coronary Arteriograms

- Left Main: normal
- LAD: normal
- LCX: mid 30-40% stenosis
- RCA: normal

The patient underwent a therapeutic thoracentesis and clinically improved
He was scheduled to have an aortic valve and root replacement and discharged 11/11/09.

CV Surgery 11/16/09 Findings

- Diffusely thickened (1 cm) pericardium with dense adhesions to the epicardium which prevented addressing the AV and aortic root
- Pericardium was resected as much as possible from the interventricular septum to the lateral right atrium
- 700 cc right pleural fluid was drained
Perioperative TEE

- EF: 55-60%
- RV: normal
- Atria: normal
- MV: normal
- TV: trace TR
- AV: Bicuspid (left and right coronary leaflets fused), no aortic stenosis, mild to moderate regurgitation

TEE continued

- PV: normal
- Ascending aorta 48mm
- Pericardium: thickened (4mm) very small effusion

Pathology

- Gross: rubbery yellow to red tissue
- Microscopy: cytologically bland spindle cells separated by abundant fibrous stroma
- Cytokeratin strongly positive (positive cells are regular suggestive of a reactive process and not mesothelioma)

Pathological Diagnosis

- Chronic fibrous pericarditis
Tissue Cultures

- No organisms or WBCs noted on gram stain
- Viral cultures negative
- Fungal cultures negative
- AFB cultures negative
- Routine cultures negative

Postscript

- The patient returned to HVHMC 11/25/09 with severe dyspnea on exertion and right pleural effusion.
- He was symptomatically improved after a thoracentesis removed 2200 cc of fluid.

Constrictive Pericarditis

- Scarring and loss of elasticity of pericardial sac
- Usually chronic, but can be subacute and transient
- The pericardium is thicker than normal in 80% of cases
- Cardiac filling is impeded by external force
- Total cardiac volume cannot change

- There is ventricular interdependence
- Cardiac compression occurs in mid through late diastole
- The bimodal pattern of venous return is maintained
- Venous return to the right heart does not increase during inspiration
- Respiratory variation in intrathoracic pressure with inspiration is not transmitted to the heart
• Early diastolic filling is more rapid than normal
• Neither ventricle fills in mid through end diastole

Cardiac Tamponade VS Constrictive Pericarditis

Common Features
• Diastolic dysfunction
• Preserved ventricular EF
• Increased ventricular interaction
• Elevated central venous, pulmonary venous, and ventricular diastolic pressures
• Pulmonary hypertension (systolic 35-50 mmHg)

Distinctive Features
• In tamponade the pericardium transmits respiratory variation in thoracic pressure to the heart
• In tamponade venous return increases enlarging the right heart which encroaches on the left
• In constrictive pericarditis there is impaired left ventricular filling due to a decreased pressure gradient from the pulmonary vessels
• Unlike in tamponade equalization of right atrial, pulmonary venous, and ventricular diastolic pressure is not present throughout the respiratory cycle
• Right atrial pressure is not changed by inspiration

Etiology of Constrictive pericarditis

• Idiopathic or viral: 42-44%
• Following cardiac surgery: 11-37%
• Following radiation therapy: 9-31%
• Connective tissue disease: 3-7%
• Post-infectious (TB): 3-6%
• Miscellaneous: 1-10%

Miscellaneous Causes

• Malignancy
• Trauma
• Drug induced
• Asbestosis
• Sarcoidosis
• Uremia

• Tuberculosis accounted for 49% of constrictive pericarditis in 1962
• This is rare now but the incidence may increase with immigrants and those infected with HIV
History

- Fluid overload
- Diminished cardiac output with exertion (fatiguability, DOE)
- Unexplained elevation in jugular venous pressure

Physical Examination

- Elevated jugular venous pressure (93%)
- Peripheral edema
- Ascites
- Pulsatile hepatomegaly
- Pleural effusions

- Pulsus paradoxus is uncommon
- Kussmaul’s sign (lack of inspiratory decline in jugular pressure) is noted in 13-21%
- Kussmaul’s sign does not distinguish from severe TR or right heart failure

- Pericardial knock (third heart sound) in 47%
- Pericardial friction rub in 16%
• Profound cachexia may be noted in late stage disease

EKG
• Non specific ST-T wave changes
• Low voltage (27%)
• Atrial fibrillation (22%)

Chest X-ray
• Calcification around the heart (lateral or anterior oblique views) in 27%
• These patients were more likely to have idiopathic disease, and longer duration of disease
• Pleural effusions

Echocardiography
• Transthoracic echo is insensitive
• TEE correlates with CT scan of the chest
• Abrupt posterior motion of the septum in early diastole with inspiration is noted
<table>
<thead>
<tr>
<th>TEE</th>
<th>Doppler echocardiography</th>
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<tbody>
<tr>
<td>• Increased pericardial thickening (37%)</td>
<td>• High E velocity of right and left ventricular inflow due to rapid early diastolic filling</td>
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<tr>
<td>• Abnormal septal motion (49%)</td>
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<td>• Atrial enlargement (61%)</td>
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<tr>
<th>CT Scan</th>
<th>MRI</th>
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<tr>
<td>• Thickened pericardium (&gt;4mm) in 72%</td>
<td>• Pericardial thickening</td>
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<td>• Pericardial calcification in 25%</td>
<td>• Dilatation of the inferior vena cava</td>
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<td>• Therapeutic procedure of choice?</td>
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<tr>
<td>Differential Diagnosis</td>
<td>Restrictive Cardiomyopathy</td>
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<tr>
<td>• Restrictive cardiomyopathy</td>
<td>• LVEDP is higher than RVEDP</td>
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<td>• Cirrhosis with ascites</td>
<td>• LVEDP and RVEDP are nearly equal in CP</td>
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<td>• Endomyocardial biopsy may be helpful in RC</td>
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<th>Plasma BNP</th>
<th>Treatment</th>
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<td>• Elevations in Constrictive pericarditis should be much less than in restrictive cardiomyopathy</td>
<td>• Pericardectomy (should be as complete as possible)</td>
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<td>• Operative mortality: 12% (1970-1885)</td>
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<tr>
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<td>• Operative mortality: 6% (1977-2000 at Mayo Clinic, Cleveland Clinic, and Johns Hopkins)</td>
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At the Mayo Clinic 83% of 90 long term survivors were free of symptoms
Patients with mild constriction, advanced disease, or mixed constrictive-restrictive disease may not benefit from surgery

Survival at the Mayo Clinic
- 5 year: 78%
- 10 year: 57%

Predictors of Adverse Outcome
- Older age
- Renal dysfunction
- Pulmonary hypertension
- Left ventricular dysfunction
- Hyponatremia

Etiology of CP and Seven Year Postop Survival Rates
- Idiopathic: 88%
- Post surgical: 66%
- Radiation induced: 27%
## End stage Disease

- Cachexia
- Atrial fibrillation
- Low cardiac output
- Hypoalbuminemia
- Impaired hepatic function due to chronic congestion

## Cirrhosis with Ascites

- Jugular venous pressure falls rapidly with removal of ascitic fluid