Hypertrophic cardiomyopathy

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HPI

- 37 yo Male
- Hypertrophic cardiomyopathy
- biological father passed away at age 46 from a heart attack
- Diagnosed in 2010
- HTN, SVT, Paroxysmal Afib, smoker
- 11/2014 - Started Bisoprolol 10 mg daily --> cardizem 240 daily and metoprolol 25 bid --> dyspnea no improvement
- 12/2014 seen in THC
- NYHA Class III, stage C
- INTERMACS 5
12/2014 – EKG

Holter 12/2014

Sinus rhythm with sinus bradycardia, junctional tachycardia and SVT/ Aflutter at 150 bpm.
• 12/2014- TTE for HCM protocol eval LVOT gradient
  EF – 67%
  Highest max gradient at narrowest point in LVOT is 35 mmHG
12/2014 cMRI r/o infiltrative ds
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• SEVERE ASYMMETRIC LEFT VENTRICULAR HYPERTROPHY WITH THE BASAL SEPTUM MEASURING 42-44 mm (LV MASS 308 gm/m2)

• NO EVIDENCE OF HOCM (LVOT PEAK SYSTOLIC VELOCITY 1.38 M/S

• LVEF 56%

• RVEF 34%

• MODERATE RV FREE WALL HYPERTROPHY AT 13 mm.

• DIFFUSE PATCHY LATE GADOLINIUM ENHANCEMENT OF THE LEFT VENTRICULAR SEPTUM, ANTERIOR AND INFERIOR WALL SEGMENTS AND RV FREE WALL. THIS IS CONSISTENT WITH FIBROSIS SEEN IN HCM. NO EVIDENCE TO SUGGEST AMYLOIDOSIS.
01/2015

- THC: Increase verapamil to 360 mg daily and Lopressor 50 bid
- Admission: pre-syncope/lightheadedness after taking the medication → bradycardia and hypotension → AICD implant by EP; Verapamil decreased to 120mg daily
- 04/2015 THC: verapamil 180mg daily and Lopressor 50 bid

02/2015 – Genetic clinic

- mutations in at least 18 genes have been identified in association with HCM: MYH7, TNNT2, MYBPC3, TNNI3, TPM1, ACTC, MYL3, MYL2, LAMP2, PRKAG2, GLA, CAV3, MTTG, MTTI, MTTK, MTTQ, TNNC1 and TTR.
- 04/2015 : (+) MYBPC3 gene
Discussion: Role of genetic testing in hypertrophic cardiomyopathy?
10/2016 Cardiology Clinic

- progressive exertional dyspnea, increased fatigue
- NYHA class III, stage C
- INTERMACS 6
- Consider myomectomy
- Start Midodrine 10mg tid
- verapamil 180mg daily and Lopressor 50 bid
11/2016 Echocardiogram

- LVEF=70%
- HCM, severe
- Very diminished LV cavity
- IV septum 3.8 cm
- PW 2.4 cm
- SAM present
- Trivial MR
- Restrictive physiology
- LVOT MG 7 mm
- LVOT PG 24 mm
- CVP 10 cm
12/2017 THC

- Obtain Stress Echocardiogram to assess LVOT gradients with exertion
- Metoprolol 50mg PO BID, Verapamil 120mg po BID
- Previously referred for myectomy evaluation – was told “too high risk”
02/2018 THC

• Proceed heart transplant evaluation
• RHC
• CPx
• Verapamil 120mg po BID and Metoprolol 50mg po BID
Echo Stress Test 2/2018

- The patient exercised according to the modified Bruce protocol for 4:28 mins, achieving a work level of max. METS:3.40. The resting HR of 64 bpm rose to a maximal HR of 142 which represents 77% of maximal age-predicted heart rate. The resting blood pressure 108/73, dropped to 78/43 at peak exercise and then increased to 129/73 during recovery.
- No EKG evidence of ischemia
- LVEF=70%
- LVOT/mid cavity gradients were 15 mmHg at rest and increased to 76 mmHg during exercise
- Mild dynamic left ventricular outflow tract (LVOT) obstruction.
03-04/2018

- RHC
  - RA 14
  - RV 30/7/12
  - PA 25/18/21
  - PCWP 17
  - TPG 4
  - MAP 82
  - PA sat 37%
  - AO sat 95%
  - Fick CO/CI  2.2/1.2
  - PVR 1.9 WU
  - SVR 2500 dsc

07/2018

- RHC
  - RA 11
  - RV 68/9
  - PA 24/6/15
  - PCWP 14
  - TPG 1
  - MAP 93
  - PA sat 45.4%
  - Ao Sat 100%
  - Fick CO/CI  2.11/1.10
  - PVR 0.47 WU
  - SVR 3109 dsc
Discussion: Treatment options for this patient?
09/2018

- Worsening exercise capacity, orthopnea, abdominal bloating
- Chronic HFpEF; NYHA Class IV, Stage D
- Start Lasix 20mg
- Not a candidate for LVAD due to small cavity
- Listed 1B status with UNOS
- Cr beginning to increase
11/2018

• Orthotopic heart transplantation
• Malignant Hyperthermia, Lactic acidosis, Rhabdomyolysis
• Post-transplant echo

• RHC
  – RA 11
  – PA 34/16/24
  – PCWP 21
  – MAP 86
  – PA sat 57.5%
  – Ao Sat 99%
  – Fick CO/CI 5.3/2.7
  – TPG 3
  – PVR <1 WU
  – SVR 1132 dsc
  – RVSWI 390