HOCM, Or NO-CM? A Curious Case of Recurrent Syncope

Syed Rahman, Paul Bergl

Medical College of Wisconsin, Milwaukee, WI

Learning Objectives
1. Recognize dynamic left ventricular outflow tract obstruction (LVOTO) predisposing one to preload dependence and consequently, syncope.
2. Identify therapeutic challenges including maintaining adequate preload and balancing the need for antihypertensive therapy against the propensity for orthostatic hypotension.

Case
Our patient is a 62 year-old female with a history of alcohol abuse who presented to the emergency room for a syncopal episode of 15 minutes duration after voiding on the toilet. The patient denied any prodome of vertigo, aura, dyspnea, chest pain, or shortness of breath. No post-ictal confusion or urinary/bowel incontinence. Her fiancé reported episodes of effort-related syncope over the preceding week.

Examination:
The patient had a harsh, late-peaking, grade III systolic murmur along the left sternal border and diminished carotid upstrokes. The patient appears hypovolemic with flat neck veins, orthostatic hypotension, acute kidney injury, and hyponatremia.

Pertinent studies:
Na: 129
Cr: 1.56
Lactate acid 1.4
Magnesium 1.0
White cell count 12.3
Cardiac Enzymes: Within Normal Limits
EKG: prolonged QTc
Renal ultrasound: normal

Hospital Course:
Given concern for aortic stenosis, we obtained an echocardiogram which demonstrated an ejection fraction of 65-70% with no valvular abnormalities. However, left ventricular hypertrophy and a substantial outflow tract resting gradient of 60 mmHg were found increasing to 87 mmHg with Valsalva maneuver: interpreted as evidence of hypertrophic cardiomyopathy (HOCM). Given the unusual age of onset and lack of family history for HOCM, we ordered cardiac resonance imaging (CMR) which revealed asymmetric left ventricular basal septal hypertrophy but no significant outflow gradient. The patient was treated with hydration and titration of lisinopril and metoprolol as tolerated. Patient was discharged in improved condition.

Discussion
- Syncope accompanied by a systolic murmur often suggests a fixed mechanical obstruction. However, LVOTO after exercise may occur in the absence of left ventricular hypertrophy.
- Predictors of hemodynamically significant LVOTO include chordal SAM at peak, small hyperdynamic left ventricles, increased septal wall thickness, and younger age.
- In this patient, LVOTO lead to preload dependence augmented by Valsalva maneuver. This physiological change is manifested by syncopal episodes.
- Therapeutic challenge
  - Maintaining adequate preload.
  - Balancing her propensity for orthostatic hypotension against the need for antihypertensive therapy particularly angiotensin II antagonists.
- Treatment
  - Hydration.
  - Reverse remodeling with lisinopril and metoprolol.
  - No additional syncopal events noted.

References