Posture related symptoms in left ventricular outflow tract obstruction

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A 70 year old man presented with breathlessness on minimal exertion. He was previously diagnosed with asymmetric hypertrophic cardiomyopathy, small airway disease, dyslipidemia, hypertension and paroxysmal atrial fibrillation. BMI was >25 and he used to consume alcohol generously.

Clinical examination showed him in sinus rhythm with blood pressure of 120/80 mmHg. Heart sounds were normal and chest examination was clear. The patient was on Atenolol 50 mg, Digoxin 125 mcg, Ramipril 10 mg, Simvastatin 40 and Tamsulosin 400 mcg once a day.

A transthoracic echocardiogram showed hypertrophied interventricular septum, in particular mid to basal (22 mm) segments but with no cavity obliteration. The left ventricular (LV) cavity size was normal, left atrium enlarged with a diameter of 5.5 cm and there was trivial mitral and aortic regurgitation. Lung function test showed FEV₁ 2.76, FVC 3.74 and a ratio of 72.8.

A CT coronary angiogram showed extensive calcification at all right and left coronary branches with a total Agatston score of 1600. Cardiac MRI confirmed small LV volumes with preserved ejection fraction, asymmetrical septal hypertrophy (24 mm), chordal SAM, mid-wall inducible perfusion defects and prominent focal patches of fibrosis at mid-wall.

A conventional coronary angiogram showed 80% mid-LAD lesion and two circumflex lesions of 50% and 20% narrowing but myocardial scintigraphy showed normal perfusion.

A stress echocardiogram showed restrictive LV physiology, and outflow tract (OT) gradient of 20mmHg, at rest. With increase in heart rate from 67 to 140 bpm with dobutamine, BP dropped from 132/77 to 86/57 mmHg, and the patient developed severe LVOT obstruction with peak gradient of 145mmHg, moderate mitral regurgitation, atrial fibrillation with BBB. Interestingly, despite peak stress HR of 165, significant LVOT obstruction and drop of systolic blood pressure the patient did not experience breathlessness or other symptoms on supine position.

Discussion: LV outflow tract obstruction with exertion is a well known cause of drop of systolic blood pressure breathlessness and potentially syncope in the elderly. Our patient developed clear evidence of LVOT obstruction and a drop in BP, in addition to arrhythmia with conduction disturbances on supine position with no symptoms, suggesting the important role of posture in acute pathophysiological disturbances. These changes in cardiac function are likely to contribute to his breathlessness when vertical, due to the additional effect of gravity, compromising venous return and stroke volume, worse LVOT and hence symptoms. Such pathophysiology also explains the traditional use of Valsalva manoeuvre in patients with HCM to diagnose LVOT obstruction non-invasively. Ideally, most patients with such condition respond to heart rate control

Figure: Parasternal long axis view (left) and apical 4 chamber view (right) showing septal hypertrophy and SAM
medications and if not alcohol septal reduction or even surgical myectomy in severe conditions.

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Ebstein malformation associated with left atrium myxoma

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A 62 year old lady, diagnosed with Ebstein malformation 12 years ago, presented to clinic with severe fatigue, breathlessness, chest pain, palpitations, and loss of weight. She had clear evidence for irregular rhythm, rales at the basal lung fields and clinical signs of severe right sided failure. A 12 lead ECG showed atrial fibrillation with wide QRS complex (130 ms) and right bundle-branch block. Echocardiography showed septal tricuspid valve leaflet displacement of 1.6 cm into the right ventricle (Figure 1&2). The right atrium was enlarged, 70 x 110 cm in diameter, with moderate tricuspid regurgitation and estimated pulmonary hypertension of 90 mmHg. In addition, there was an echo dense mass (26 x 30 mm) attached to the left atrial side of the interatrial septum, with morphologic appearance consistent with atrial myxoma. According to our knowledge, this case of such combination seems the first reported in the literature.

The Ebstein’s anomaly, first prescribed by Wilhelm Ebstein in 1866, is a rare cardiac anomaly that accounts for less than 1% of all congenital heart disease and involves both genders equally. Ebstein anomaly consists of abnormal attachment of tricuspid valve leaflets to the annulus, with a downward displacement of the tricuspid valve into the right ventricle, dysplastic valve tissue associated with tricuspid regurgitation. Most patients with this anomaly remain asymptomatic for years. The predominant symptoms are fatigue, breathlessness on exertion and cyanosis. Palpitation in the form of paroxysmal atrial arrhythmia and premature ventricular beats are common, whereas ascites and peripheral edema are present in some of these patients. Ebstein anomaly may be associated with other congenital heart malformations: ventricular septal defect, interatrial septal defect or patent foramen ovale, aortic coarctation, pulmonary atresia with an intact ventricular septum, partial atrioventricular canal, hypoplastic pulmonary arteries, patent ductus arteriosus, pulmonary stenosis, tetralogy of Fallot, cleft anterior leaflet of the mitral valve or mitral valve prolapse. Surgical treatment of Ebstein anomaly includes valve replacement or the native valve repair.

Our patient remained silent for 50 years until presented with

Figure 1: Apical 4-chamber view showing tricuspid valve displacement into the right ventricle, giant right atrium and left atrial myxoma.

Figure 2: Respective images of color-Doppler showing the level of accelerated tricuspid flow, deep in the cavity of the right ventricle.