CASE REPORT

Hypertrophic cardiomyopathy masquerading as infiltrative restrictive cardiomyopathy and refractory congestive failure-resolution with catheter ablation of atrial flutter – A case report

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We report a case of a young male presenting as Restrictive cardiomyopathy, refractory heart failure and syncope due to typical right atrial flutter complicating hypertrophic cardiomyopathy.

Successful catheter ablation of the flutter promptly ameliorated the congestive failure with resolution of restrictive physiology.

Keywords: Hypertrophic cardiomyopathy, Atrial flutter, Catheter ablation

Hypertrophic cardiomyopathy (HCM) is a genetically mediated disease with variable presentation. Common arrhythmias in HCM are ventricular tachycardia and atrial fibrillation, both of which are poorly tolerated and are known causes of sudden cardiac arrest (SCA) [1].

A 39-year-old male presented with recurrent palpitations occurring over a period of six months. He had significant effort intolerance with fatigue, recurrent presyncope and one recent episode of resuscitated SCA. At admission, his heart rate was 120 beats per minute, blood pressure 100/74 mmHg, jugular venous pressure elevated beyond the angle of mandible, varying S1, and clear lungs. Baseline ECG revealed atrial flutter (AFl) with 2:1 conduction (Fig. 1). Echo revealed dilated atria, asymmetrically thickened ventricular septum, restrictive filling pattern (E/A > 2, short E deceleration time, indicative of severely elevated filling pressures) of both ventricles and normal biventricular systolic function. There was dense spontaneous echo contrast (SEC) in the left atrium but no thrombus either in the left atrium or left atrial appendage on transesophageal echo. He

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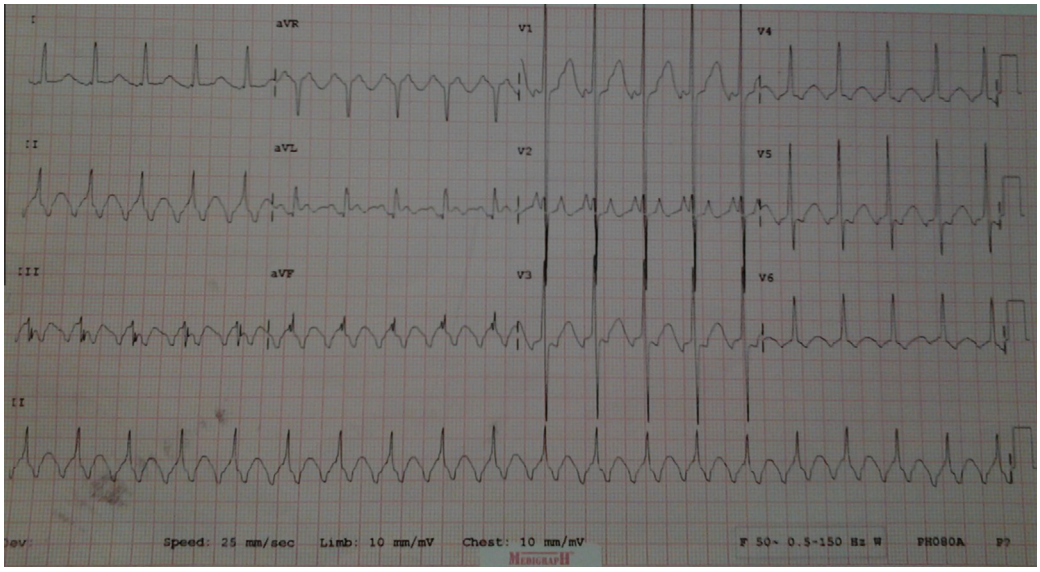

Figure 1. Baseline ECG showing typical counterclockwise atrial flutter with 2:1 conduction.

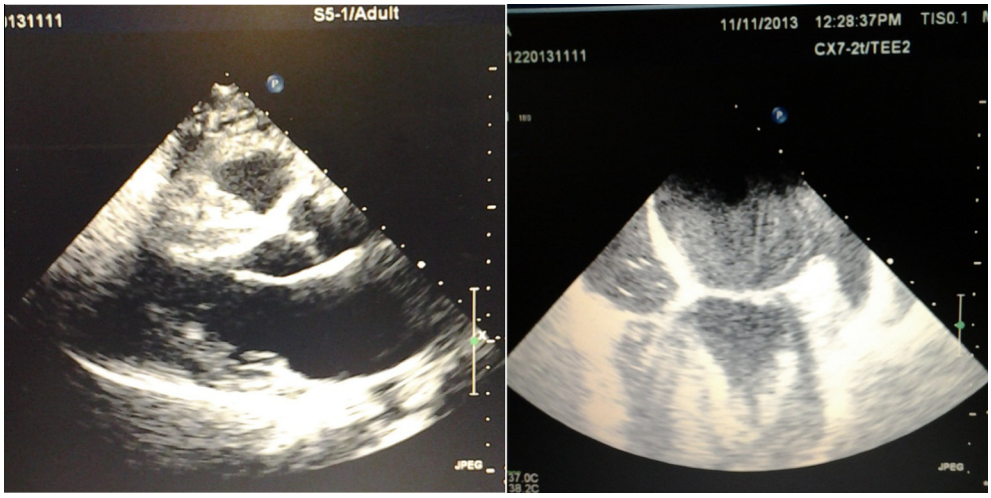

Figure 2. Echocardiogram showing 'speckled' thickened interventricular septum and dilated left atrium with swirling 'spontaneous echo contrast'.

had been previously treated with metoprolol, digoxin and amiodarone with no improvement in symptoms, and on coumarin anticoagulation to maintain International Normalized Ratio (INR) 2.5–3. In view of recurrent presyncope and recent resuscitated SCA with intermittent fast ventricular rate during AFl, the patient was presented with the treatment options, including implantable cardioverter-defibrillator (ICD) and radio frequency (RF) ablation. ICD was declined as non-affordable, and he was taken up for 3D mapping guided electrophysiological (EP) study and RF ablation of the persistent AFl.

EP study was performed under fasting conditions and under conscious sedation with patient in tachycardia. Adequate heparinization was maintained throughout the procedure to keep the activated clotting time >300 s. CARTO 3D mapping was performed with Cordis-Webster NaviStar 8F orange catheter, and tachycardia was confirmed to be typical isthmus dependent macro-reentrant right atrial flutter (Fig. 2). RF ablation of the cavotricuspid isthmus was performed with open saline irrigation at 30 W/40 °C. Atrial flutter terminated during RF ablation and further consolidating lesions were given in sinus

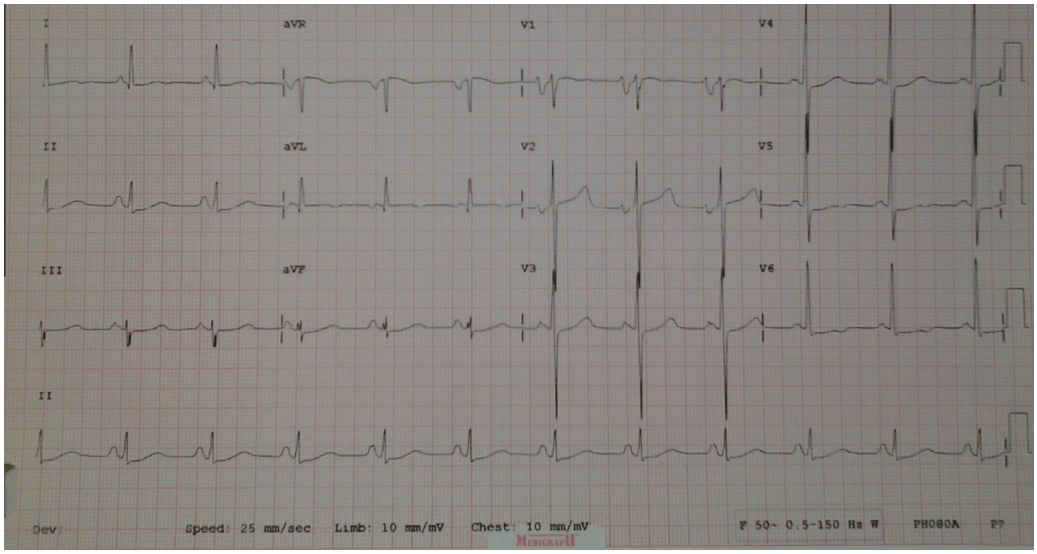

Figure 3. ECG-post ablation.

rhythm to obtain bidirectional block, confirming non-conduction of impulses across the ablation line in isthmus from both septal and lateral sides, and indicating successful ablation and cure of propensity for flutter.

Patient was discharged without any antiarrhythmic medication. At short-term follow-up, he had good effort tolerance, was in sinus rhythm (Fig. 3), and free from congestive heart failure. Echo revealed improved diastolic function (prolonged relaxation) and no SEC.

Discussion

Although asymmetrical septal hypertrophy is the most common type of morphological pattern, HCM can present with concentric, apical, or free wall LV hypertrophy. While ECG may show evidence of ventricular hypertrophy and atrial enlargement, 2D echocardiography is the diagnostic method of choice. In our patient, the thickened septum with ''speckled'' appearance was similar to that seen in amyloid infiltrative cardiomyopathy, as the diastolic LV filling was restrictive. Typical atrial flutter is a macro-reentry using cavotricuspid isthmus in right atrium as a critical slow-conducting zone to perpetuate the tachycardia. Commonly (90%), it is counterclockwise as in our patient, with flutter waves negative in inferior leads. Atrial flutter is relatively uncommon compared to atrial fibrillation in HCM, but all forms of atrial tachyarrhythmias are known to be poorly tolerated in HCM where cardiac output is critically dependent on the atrial support for ventricular filling and adequate diastolic filling time. In this situation of hypertrophied stiff ventricles with baseline impaired relaxation, tachycardia led to severe elevation in filling pressures as manifested by restrictive filling pattern, leading to a mistaken diagnosis of restrictive cardiomyopathy. Ablation of the tachycardia led to prompt improvement in hemodynamics with improved relaxation, longer diastolic filling time and better atrial kick, supporting ventricular filling.

In this patient, AFl was resistant to multiple antiarrhythmic drugs, caused poor hemodynamics, severe effort intolerance, refractory congestive heart failure, thrombotic milieu in left atrium, and when associated with fast ventricular rate led to sudden cardiac arrest from which the patient was resuscitated. Ablation for atrial fibrillation and atrial tachycardia have been reported in HCM but most reports of atrial flutter have been with pharmacological management [2,3].

In conclusion, atrial flutter worsened the hemodynamics of a well-tolerated hypertrophic cardiomyopathy, mimicking restrictive cardiomyopathy with refractory congestive failure and resuscitated sudden cardiac arrest. Catheter-based RF ablation of the atrial flutter was successful in achieving prompt clinical improvement.

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