Isolated Right Ventricular Hypertrophic Obstructive Cardiomyopathy - A Case Report

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Abstract:
Hypertrophic cardiomyopathy of the left ventricle is the most common genetically determined cardiac disease with a prevalence of 1 in 500. But right ventricular involvement is uncommon and isolated right ventricular hypertrophic cardiomyopathy is rare. A 19 year old male patient presented with complaints of breathlessness, palpitation and chest pain. He was found to have a holosystolic murmur in 3rd intercostals space along left sternal border. On echocardiography, predominant right ventricular outflow tract obstruction with right ventricular free wall and interventricular septal hypertrophy was observed without left ventricular involvement.

Keyword:
Hypertrophic cardiomyopathy, isolated, right ventricular, echocardiography.

INTRODUCTION:
Hypertrophic cardiomyopathy (HCM) is classically thought of as a disease of the left ventricle, but right ventricular (RV) abnormalities have also been reported. Unlike left ventricular pathology, involvement of the right ventricle is seen in only about 15% of patients and isolated involvement of right ventricle is rare. Here we report a case of isolated right ventricular obstructive cardiomyopathy without left ventricular involvement.

CASE REPORT
A 19 year old upholstery worker presented with complaints of exertional breathlessness, NYHA grade II. He also had intermittent chest pain and palpitations. There was no history of syncope. He was told to have ventricular septal defect clinically at 1 ½ yrs of age. Regarding family history, he is the 3rd of 4 siblings born of a non-consanguinous marriage his mother was diagnosed to have acquired mitral stenosis, one sister has MVP of anterior mitral leaflet and a brother was found to have non-compaction of left ventricle. There is no family history of sudden death. On examination there was no clubbing, cyanosis or markers congenital heart disease. His Pulse was regular and rate was 68/min,
Blood pressure was 116/80 mm Hg. Jugular venous pulse was normal. Apical impulse was in the Left 5th intercostal space in mid-clavicular line. Supraclavicular pulsations and a grade 2 parasternal heave were present. A grade 3/6 holosystolic murmur was heard in the left sternal border 3rd intercostal space, which increased during inspiration, and on valsalva as well as standing and decreased on squatting. There was no ejection click and second sound was normally split with normal intensity of both components. ECG showed Right axis deviation, Right atrial enlargement and biventricular hypertrophy with strain.

Chest X ray showed right atrial enlargement with right ventricular apex. Echocardiography revealed gross hypertrophy of the right ventricular free wall and interventricular septum (RV free wall thickness 2.04 cm and IVS thickness 2.27 cm) and RV outflow tract peak systolic gradient was 113 mm Hg. TAPSE was 2.3 cm. LVEF was 53%, and LV systolic function, valvular structure and function were normal.

DISCUSSION:
HCM is characterized by a thickened but non dilated left ventricle, in the absence of other cardiac or systemic conditions (e.g., aortic valve

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stenosis, systemic hypertension, and physiologic athlete’s heart) capable of producing the magnitude of left ventricular (LV) hypertrophy evident.\textsuperscript{1} Mutated sarcomeric genes are associated, most commonly beta-myosin heavy chain and myosin-binding protein C. There is an increased transverse diameter of myocytes, chaotic disorganised arrangement and abnormal intramural coronary arteries (wall thickened and narrow lumen) which leads to silent myocardial ischemia, myocyte death and replacement scarring by fibrosis. Disturbed electrophysiology leads to arrhythmias and sudden death. Longstanding LV outflow tract obstruction (basal gradient 30 mm Hg) is a major determinant of HCM-related progressive heart failure symptoms. Subaortic obstruction in HCM is produced in the proximal left ventricle by systolic anterior motion (SAM) of mitral valve and midsystolic ventricular septal contact due to asymmetric septal hypertrophy (ASH). SAM is produced by drag and venturi effect. Mitral regurgitation is a secondary consequence of SAM, with the jet (usually mild to moderate in degree) directed posteriorly. There may be associated diastolic dysfunction. Clinical features include exertional dyspnoea, chest pain, syncope and palpitations. The pulse is characteristically described as jerky. There may be double or triple LV apical impulse, and a medium-pitched systolic ejection murmur along the lower left sternal border and at the apex, which varies in intensity with the magnitude of the subaortic gradient, either at rest (supine or standing), with the Valsalva maneuver, or during and immediately following exercise. Echocardiographic criteria for diagnosis include Ø LV wall thickness of 13 mm in the anterior septum or posterior wall or 15 mm in the posterior septum or free wall, in the absence of LV dilatation or other cardiac and systemic causes of increased mass. Ø Asymmetric septal hypertrophy is defined as a ratio of septal thickness to posterior wall thickness of at least 1.3-1.5. Ø Narrowing or obstruction of the LV outflow tract caused by IVS and the anterior leaflet of the mitral valve. Ø Severe SAM (drag effect, venturi effect) with septal-leaflet contact. Other investigative modalities include cardiac MRI, cardiac catheterization and electrophysiology. Although the genetics of RV HCM have not been exemplified well, histologic features appear to be similar to those of the LV, suggesting similar pathogenesis. Major RV involvement may cause RV outflow obstruction and decreased RV diastolic filling, with increased risk of severe dyspnea, arrhythmias, and pulmonary thromboembolism. Isolated right ventricular hypertrophic obstructive cardiomyopathy presents as systolic murmur in left upper parasternal region with clinical and electrocardiographic evidence of right ventricular and right atrial strain. Normal intensity and splitting of second sound and absence of pulmonary valvular click differentiate it from pulmonary valve stenosis while the inspiratory increase in the systolic murmur differentiates it from ventricular septal defect. Echocardiography with colour flow imaging excludes other possibilities and shows hypertrophy of right ventricular outflow tract muscle with systolic gradient across the obstruction. Like classical hypertrophic obstructive cardiomyopathy, patients may have partial respite of symptoms with beta-blockers. Several medical and surgical therapies have been tried with variable results and newer techniques such as percutaneous catheter ablation are under study but the ideal treatment for patients with RV disease is still unknown. Further characterization of RV HCM is necessary to expand the clinical features and
treatments of this type of HCM more clearly

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ABBREVIATIONS

HCM -Hypertrophic Cardiomyopathy
RV - Right Ventricle
LV -Left Ventricle
NYHA -New York Heart Association
ECG

TAPSE -Tricuspid Annular Plane Systolic Excursion
LVEF -Left Ventricular Ejection Fraction
IVS -Interventricular Septum
SAM -Systolic Anterior Motion