中文題目：肥厚性心肌病變致右心流出道阻塞之病例報告

英文題目：A case of hypertrophic cardiomyopathy related right ventricular outflow tract obstruction

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Introduction:

We demonstrated a case with hypertrophic cardiomyopathy (HCM) related right ventricular outflow tract obstruction (RVOTO). We also reviewed previous literation and tried to established current consensus about prevalence, diagnosis and treatment.

Case Presentation:

We presented a 45 years old man with past history of hypertension. He didn't follow at outpatient department (OPD) regularly and presented with hypertension urgency on February, 2017. Though denied chest tightness, dyspnea or headache, he presented with blood pressure 217/136mmHg. Physical examination showed a systolic murmur over left upper sternum border, grade 3/6. We added hypertensive medication and suggested admission for further survey. Electrocardiogram (ECG) showed left ventricular hypertrophy and left axis deviation. Cardiac echo showed both left and right ventricular hypertrophy. (Figure 1 and 2) Also, mild tricuspid regurgitation and severe pulmonary stenosis was found. (Figure 3) Right heart catheterization was done, showing suspected pulmonary stenosis with pressure gradient around 52mmHg. Coronary artery angiography was done, demonstrating coronary artery disease, 1-vessel disease with 70% stenosis over right coronary artery. We then arranged chest computed tomography angiography (CTA), however no obvious stenosis over pulmonary trunk, embolus or specific lung disease was found. We suggested further investigation but patient refused and lost follow up.

He then presented with chest tightness 5 years later. Chest tightness was dull pain without radiation and especially happened on exertion. Associated symptoms included dyspnea on exertion. No peripheral edema, orthopnea was noticed. ECG showed T wave insertion over lead III and aVF. Coronary artery angiogram showed coronary artery disease, 2-vessels disease with 99% occlusion over right coronary artery (RCA) and left anterior descending artery (LAD). Plain old balloon angioplasty was done over RCA and LAD. Cardiac echo still showed elevated pulmonary pressure gradient. We arranged trans-esophagus echocardiography (TEE) this time, showing obstruction over sub-valvular level at right ventricular outflow tract. (RVOT) (Figure 4 and 5) The hypertrophic right ventricular free wall and septum had compressed the outflow tract. M-mode showing completely obstruction of RVOT. (Figure 6) No obvious obstruction at pulmonary valve level through M mode. (Figure 7) We kept medication with Ticagrelor, Atorvastatin, Valsartan, carvedilol and suggested regular OPD follow.

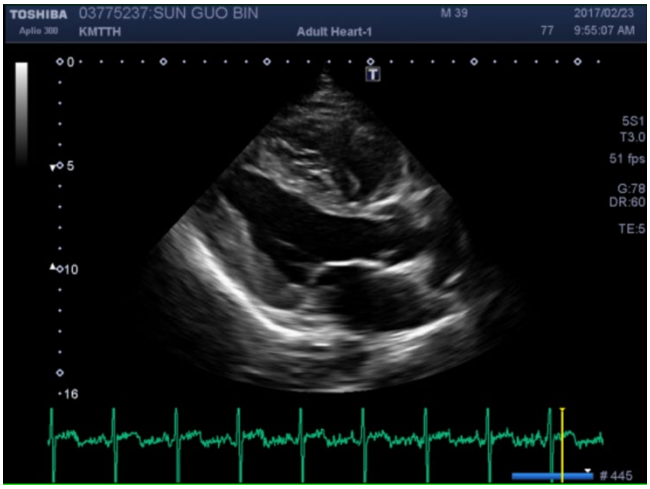

Figure 1.
Parasternal long axis view of TTE, showing LV and RV hypertrophy.

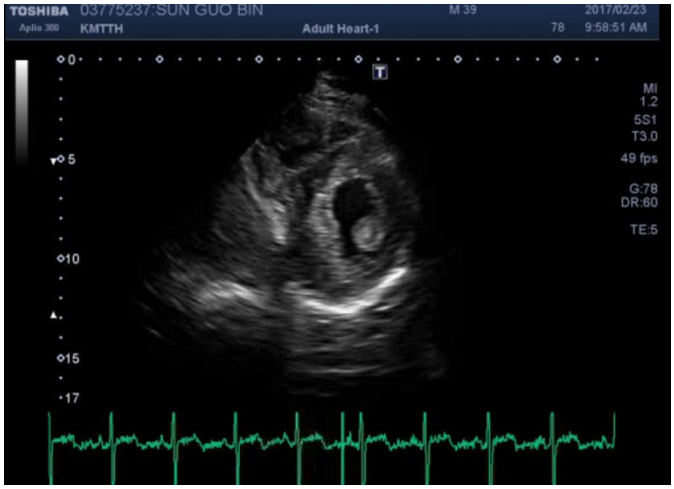

Figure 2.
Parasternal short axis view of TTE, showing LV and RV hypertrophy.

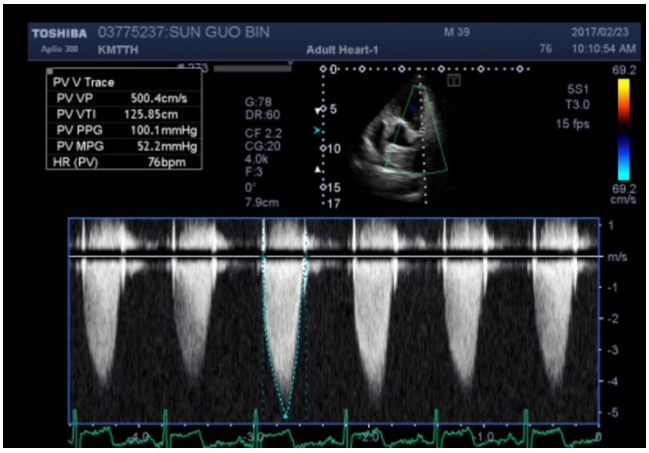

Figure 3.
Continues wave doppler over pulmonary valve through TTE showing velocity of 5m/s, with estimated pressure gradient 100mmHg.

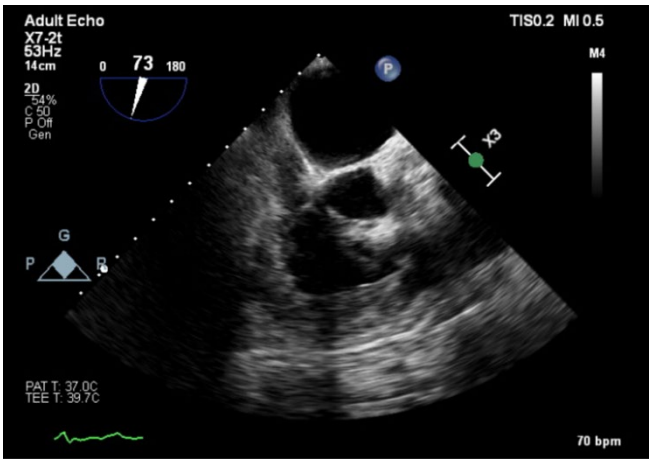

Figure 4.
TEE showing obstruction of RVOT while systole.

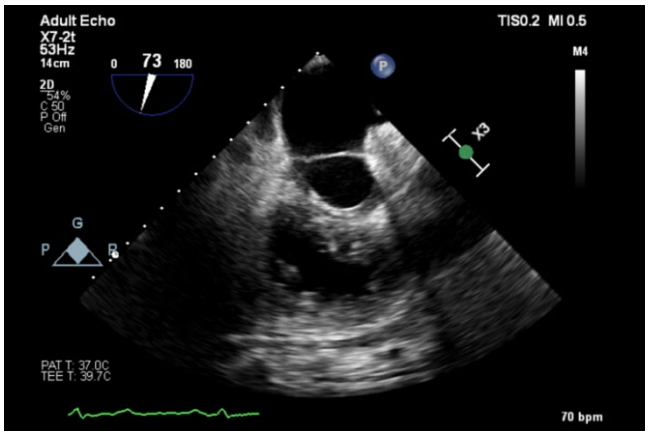

Figure 5.
TEE showed patent RVOT while diastole.

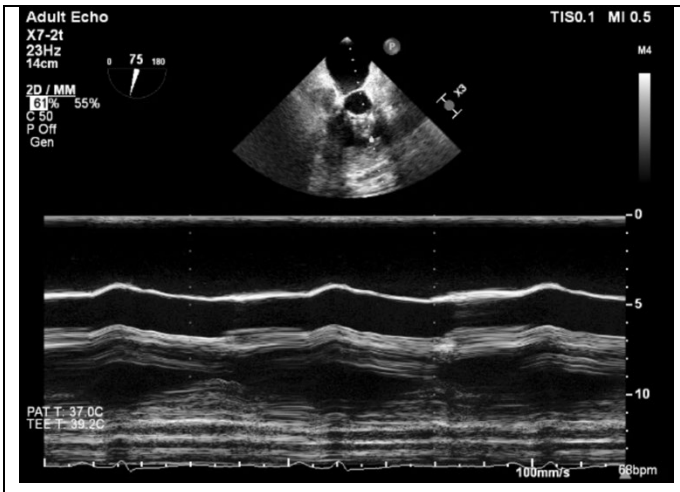

Figure 6.
M mode of TEE at sub-valvular level, showing dynamic collapse.

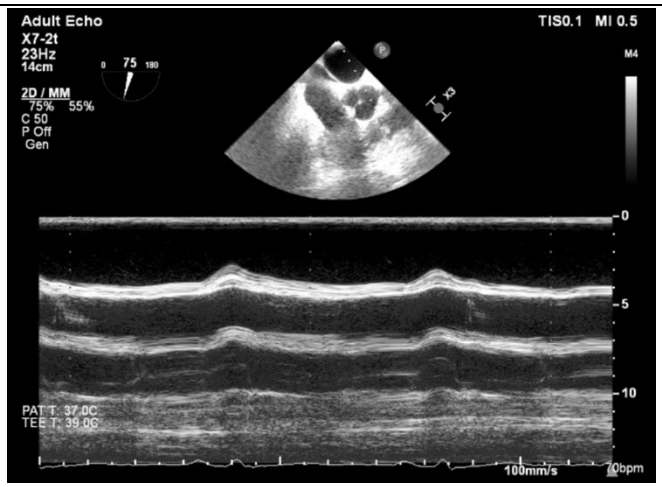

Figure 7.
M mode of TEE at pulmonary valve level, showing no obstruction.

Discussion:

Hypertrophic cardiomyopathy (HCM) is characterized by the presence of increased ventricular wall thickness that is not explained by abnormal loading condition (hypertension, valvular disease).[1] The prevalence is about 0.2% in general and is found mostly related to multiple sacromeric proteins mutation.[1] Patient with HCM frequently have systolic anterior motion of mitral valve, causing dynamic obstruction of LVOT.[1] Though RVOTO was rarely mentioned in previous text book, it was

actually not a rare finding in non-sydromic HCM patients and had been described in up to 15% when examined carefully with a diagnostic criterion of peak velocity > 2.0m/sec.[2]

Some cases of HCM causing RVOTO had been reported back around 1970s.[3-6] According to Shimizu, M., et al. study, they evaluated data from 91 HCM patients. 14 of 91 patients met RVOTO criteria defined by peak velocity and 6 of 14 patients had coincident LVOTO as well.[2] More recent cases had demonstrated HCM with isolated RVOTO. [7-10] More recently, Guo, X., et al. had recruited 2650 HCM patients from 1996 to 2013.[11] 34(1.3%) patients had demonstrated with severe right ventricular hypertrophy (SRVH) with right ventricle thickness > 10mm in end diastole. 11 out of 34 patients had demonstrated RVOTO at rest with a peak pressure gradient of 52mmHg and mostly of them increased after provocation test. 14 Patients with SRVH had related with increased mortality. However, the presence of RVOTO was not related to further increased mortality[11]

After reviewing previous literature, mechanism of RVOTO in HCM patients still varied. Some demonstrated hypertrophy of crista supraventricularis, moderator band and trabeculae.[12] Some demonstrated more variable anatomic mechanism included hypertrophic septum bulging in to RVOT, septal muscle bundle or even septal hypertrophy extending to infundibulum level.[13] More recently, Jagdish C. Mohan had demonstrated a case with isolated RVOTO.[14] Hypertrophy of parietal and septal muscle causing a fixed obstruction around infundibulum was considered the key anatomic change which was similar to the TEE findings in our case.

Unlike LVOTO, few consensuses regarding definition, treatment was made. Few cases reported myomectomy trough conal part of right ventricle.[15] However those cases included was biventricular outflow tract obstruction. The indication, surgical approach for HCM related isolated RVOTO remained uncertain.

Conclusion:

We presented a case with hypertrophic cardiomyopathy related right ventricular outflow tract obstruction. HCM induced RVOTO may be underdiagnosed and may coincident with LVOTO. Mostly severe right ventricular hypertrophy carried a poor prognosis but it was not related to the presence of RVOTO. Currently the standard diagnosis and treatment of HCM induced RVOTO was still under discussion.

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