Coexistence of Sick Sinus Syndrome and Hypertrophic Cardiomyopathy in Patient with Syncope

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ABSTRACT

Introduction: Both hypertrophic cardiomyopathy (HCM) and sick sinus syndrome (SSS) can cause syncope. However HCM and SSS different enitity, rarely seen together.

Case report: 76 years old female patient admitted to hospital with a complaint of syncope due to sinus bradycardia.Since the drug intoxication, electrolyte imbalance and coronary ischemia was ruled out she was accepted as sick sinus syndrome. During follow up in hospital the patient had also experienced ventricular tachycardia. Transtorasic echocardiograpy revealed hypertrophic cardiomyopathy. She underwent DDD-ICD (Implantable cardi-overter defibrillator) implantation due to having high risk for sudden cardiac death

Conclusion: Patients with HCM may also have SSS together or SSS may develop in later years. Antiarrhythmic drug therapy should be given carefully and 24 hour ambulatory electrocardiogram monitoring should be donein certain periods.

Keywords: Hypertrophic cardiomyopathy, sick sinus syndrome, syncope

INTRODUCTION

Hypertrophic cardiomyopathy (HCMP) is an autosomal dominant inherited genetical diseae which is characterized by myocardial hypertrophy.The prevalence of HCMP is 0.01-0.1% in generalpopulation.[1] One of the most fearful complication of HCMP is sudden cardiac death (SCD). The risk factors for SCD in HCMP are unexplained syncope, ventricular tachycardia attacks (without continuous ambulatory ECG), abnormal blood pressure response in exercise test, left ventricular hypertrophy and SCD in family history. Sudden cardiac death rate is up to 4-6% in a year in high-risk patients.[2-3] Sick sinus syndrome (SSS) due to sinus node degeneration, is characterized by arrhythmias such as sinus bradycardia, sinus arrest, atrial tachycardia and atrial fibrillation.The usual symptoms in SSS are dizziness, presyncope and syncope. The prevalence of SSS is 0.3 % in general population.. Sick sinus syndrome is usually diagnosed with clinically and electrocardiography.[4] Simultaneous detection of SSS and HCMP is very rare entity. In this paper we present a case with syncope and cardiac arrest who had concomittant SSS and HCMP.

CASE REPORT

Seventy-six years old woman admitted to the emergency department with a complaint of dizziness and syncope. In her history she had a hypertension for 10 years and using angiotensin converting enzyme inhibitor. She had no known other systemic disease. A year ago she had experienced syncope withoutneeding further investigation. On admission her blood pressure was 60/30 mmHg, heart rate was 35 beats/min. There was a 2/6 degree systolic murmur in aortic area and no murmur detected in the carotid artery. Physical examination of other systems was normal. Electrocardiogram revealed sinus bradycardia with a 35 beats/min rate (Figure-1). During first physical examination patient had experinced ventricular tachycardia which was defibrilated with 300 joule. Following defibrilation, the patient remained in sinus bradycardia and developed respiratory arrested. Therefore she was intubated immediately and implanted temporary pacemaker.During follow-up in hospital patient developed new onset atrial fibrillation with rapid ventricular response. Diltiazem (25 mg) was given intravenously to decrease the ventricular rate of atrial fibrillation. The rhythm was turned to sinus rhythm on first day andshe was extubated on second day. However patient's rhythm was remained in sinus bradycardia and she was dependent on temporary pacemaker. The seconadry causes for sinus bradycardia such as electrolyte disturbances, drug intoxication were ruled out, for this reason patient was diagnosed with SSS. When becoming clinically stable she had underwent echocardiography which revealed 50-55% ejection fraction, mild mitral and aortic regurgitation, left atrial enlargement and impaired diastolic filling. Interventricular septum and left ventricular posterior wall thickness was 22 mm and 11 mm respectively. Septum posterior wall ratio was 2. Left ventricular dimensions were in normal range.Echocardiographic findings demonstrated HCMP. In order to rule out myocardial ischemia she underwent coronary angiography which showed no coronary artery stenosis. Left heart catheterization revealed 30 mmHg gradient in the left ventricular outflow tract (Figure-2). DDD-ICD (Implantable cardioverter defibrillator) was implanted due to coexistence of SSS-HCMP and ventricular tachycardia.

DISCUSSION

The most fearful symptom and complicationof HCM is SCD. Therefore risk factors for SCD in HCMPshould be identified and appropriate treatment must be planned. Familial history of sudden death, unexplained syncope, ventricular tachycardia, abnormal blood pressure response in exercise test, and left ventricular hypertrophy are the main risk factors for SCD.[3]

Syncope is an another symptom and clinical finding of HCMP. The main reasons of syncope in HCM are ventricular arrhythmias and left ventricular outflow tract obstruction.

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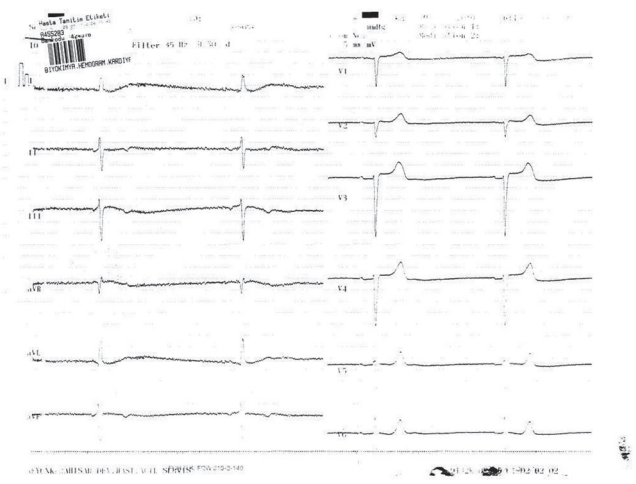

Figure-1: Electrocardiogram shows deep sinus bradycardia

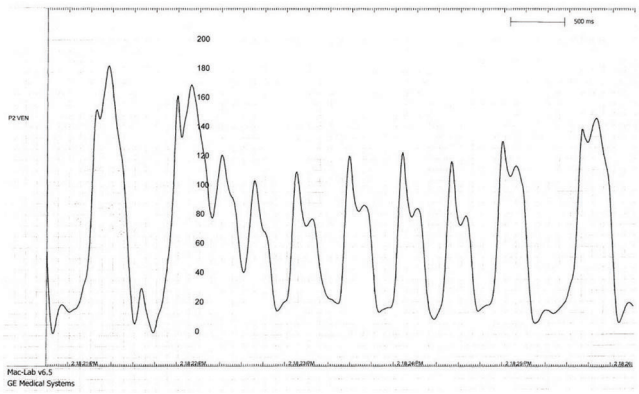

Figure-2: Left heart catheterization revealed 30 mmHg gradient in the left ventricular outflow tract

Severe left ventricular outflow tract obstruction reduces stroke volume which leads to reducecerebral perfusion and evantuallyresulted in syncope. Reduced stroke volumeespecially occurs during exercise. HCM without left ventricular outflow tract obstructioncalled as nonobstructive HCMP may also cause syncope due to arrhytmic reasons which is not uncommon.[2]

Treatment of HCMP includes pharmacological, interventional and surgical methods. Currently ICD is the most effective treatment for the primary and secondary prevention of sudden death in patients with HCM.[5] However, the data about ICD treatment implantation for the primary prevention of SCD in patients with hypertrophic cardiomyopathy is limited.

SSS is an arrhtymic pathology which is characterized by degenerative changes in sinus node.However degenerative changes can be seen not only in sinus node but also in atrial wall, AV node or bundles. Sinus bradycardia, sinus arrest, sinus tachycardia, and slow ventricular response or rapid ventricular response of atrial fibrillationcould be the main findings of SSS.[6] The most frequently symptoms of SSS are dizziness, presyncope and syncope.The conditions affecting the sinus nodeactivity such as drugs, myocardial ischemia, electrolyte imbalance should be ruled out before making a definitive diagnosis of SSS. In our case we didn't find any secondary causes that effect sinus not activity.

Treatment of SSS depends on the underlying rhythm problems. Antiarrhythmic and rate control drugs such as digitals, beta blockers, verapamil, diltazem, amiodarone, should be avoided since those compromise SSS bydeeping bradycardia. The cases who need antiarrhythmic drugs for their tachyarrhythmia should be fitted with pacemaker to prevent from bradyarrhythmia associated symptoms.[6]

The coexistence of SSS and HCMP is very rare. Ten patients have been reported so far. The age of the cases were ranging 5 to 84.[7-9] Yoshika etal reported a case with HCMP-SSS also associated with Klinefelter's Syndrome.[7] The other case having HCMP-SSS has also Moyamoya disease.[8] Both of these cases strengthens the possibility of the genetic origin. We believe that the coexistence of HCMP and SSS, may be, perhaps, a new sub-group of HCMP.

We assumed that the present case having both HCMP and SSS was accepted in high risk group for sudden death due to experincing ventricular tachycardia attacks, havinga history of previous syncope and severe left ventricular hypertrophy and also having symptomatic bradycardia. Because of the coexistence of HCMP and SSS, we have implanted DDD-ICD in our patient.

It has been reported that SSS could be developed a few years later after diagnosing of HCM. Therefore, while deciding an ICD treatment in HCMP the clinician should consider that patient with HCMmay also have aconcomittant SSS in that period or develop SSS in the later times.

CONCLUSION

In conclusion, patients with HCM may also have SSS together or SSS may develop in later years. Antiarrhythmic drug therapy should be given carefully and 24 hour ambulatory electrocardiogram monitoring should be donein certain periods. Otherwise, asymptomatic SSS patients may become symptomatic because of antiarrhythmic drugs.

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