SUDDEN DEATH DUE TO HYPERTROPHIC CARDIOMYOPATHY- A CASE REPORT

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ABSTRACT

Hypertrophic cardiomyopathy (HCM) is an important cause of sudden death in individuals under 30 years of age often associated with exercise. It is a primary disease of cardiac muscle. The mechanism of sudden death is nearly always a ventricular arrhythmia. Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiovascular disease that is usually transmitted as an autosomal dominant trait. It is characterized by a hypertrophied left ventricular wall and normal to small sized ventricular cavities in the absence of another cardiac or systemic disease that might produce these alterations, such as aortic valve stenosis or systemic hypertension. The interventricular septum is usually 90% thicker than the left ventricular free wall and the ratio of septal to free wall thickness is usually \( \geq 1.3 \). The normal interventricular septum / left ventricular free wall (IVS/LVFW) ratio is \(< 1.3\). In some cases of hypertrophic cardiomyopathy (incidence varying between 2 to 20%) the hypertrophy is symmetric.

Hypertrophic cardiomyopathy may block blood flow out of the ventricle. When this happens, the condition is called obstructive hypertrophic cardiomyopathy. In some cases, the septum thickens and bulges into the left ventricle. In both cases, blood flow out of the left ventricle is blocked. As a result of the blockage, the ventricle must work harder to pump blood out to the body.

Hypertrophic cardiomyopathy also can affect the heart’s mitral valve, causing blood to leak backward through the valve.

INTRODUCTION

Death is said to be sudden or unexpected when a person not known to have been suffering from any dangerous disease, injury or poisoning is found dead or dies within 24 hours after the onset of terminal illness.\(^1\)\(^2\)

Sudden cardiac death is most commonly defined as unexpected death from cardiac causes either without symptoms, or within 1 to 24 hours of symptom onset (different authors use different criteria).\(^3\)

Hypertrophic cardiomyopathy (HCM) is an important cause of sudden death in individuals under 30 years of age often associated with exercise. The mechanism of sudden death is nearly always a ventricular arrhythmia. It is a primary disease of cardiac muscle that is usually transmitted as an autosomal dominant trait. It is characterized by a hypertrophied left ventricular wall and normal to small sized ventricular cavities in the absence of another cardiac or systemic disease that might produce these alterations, such as aortic valve stenosis or systemic hypertension. The interventricular septum is usually 90% thicker than the left ventricular free wall and the ratio of septal to free wall thickness is usually \( \geq 1.3 \). The normal interventricular septum / left ventricular free wall (IVS/LVFW) ratio is \(< 1.3\). In some cases of hypertrophic cardiomyopathy (incidence varying between 2 to 20%) the hypertrophy is symmetric.

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Sometimes the thickened heart muscle doesn’t block blood flow out of the left ventricle. This is called nonobstructive hypertrophic cardiomyopathy. The entire ventricle may become thicker, or the thickening may happen only at the bottom of the heart. The right ventricle also may be affected.\(^4\)^\(^5\)^\(^6\)

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The most important histologic features of HCM myocardium are massive myocyte hypertrophy, myofiber disarray and interstitial fibrosis.7

Case report

A 19 years old male who was a manual labourer, suddenly had chest pain and breathlessness at workplace. He collapsed after sometime. He was taken to the hospital where he was declared brought dead. He had habit of smoking bids and drinking alcohol. There was no history of hypertension or any other disease. There was no family history of any congenital heart disease. Autopsy was conducted at the mortuary of Victoria hospital, Bangalore Medical College and Research Institute, Bangalore. On external examination the dead body measured 166 cm in length, moderately built and nourished. Rigor mortis was present all over the body. Post mortem staining was seen over the back of the body. There were no external injuries on the body. On internal examination, Heart was massively enlarged and weighed 580 gms. The heart was thick walled with massive myocardial hypertrophy without dilatation of ventricles. The interventricular septum was thickened disproportionately (3.5 cm) in comparison to the free wall of left ventricle (2.6 cm). Right ventricle wall thickness measured 1.0 cm. Hyperaemic areas seen in the hypertrophied myocardium at places. Heart valves were normal. Left coronary artery, left anterior descending artery and left circumflex artery showed atherosclerotic changes but lumen of these coronaries were grossly patent. Right coronary artery also showed atherosclerotic changes and lumen was about 25% narrowed. Lungs showed congestion and edema. Other internal organs were congested. On histopathological examination of the heart, there was marked hypertrophy of myocytes with focal areas of disarray of hypertrophied myocardial fibers and enlarged nucleus in the septum, and areas of interstitial fibrosis along with inflammatory cells. Associated atherosclerotic coronary artery disease was also seen in this case and the person died suddenly due to myocardial infarction.

Fig 1 The photograph of the Formalin fixed heart showing Asymmetric hypertrophy of interventricular septum in case of Hypertrophic Cardiomyopathy.

Fig 2 The photomicrograph of Hypertrophic Cardiomyopathy showing disarray of hypertrophied myocardial fibers and enlarged nucleus (H & E, 10X).

DISCUSSION

The sudden death in apparently healthy young individuals is always a devastating and shocking event. The incidence of sudden deaths in young due to various cardiac pathologies has wide variations in different parts of the world. Now because of change in lifestyle and habits, many cardiac pathologies are noticed in comparatively younger age group even in a developing country like India and some of these cardiac pathologies are inheritable.

The causes of sudden cardiac death differ greatly among various age groups. In individuals > 40 years old, atherosclerotic coronary heart disease is the most common cause. Between 1 to 40 years of age, the causes of sudden cardiac death are commonly hypertrophic cardiomyopathy, myocarditis, congenital heart disease, arrhythmogenic right ventricular dysplasia/ cardiomyopathy etc.8

Hypertrophic cardiomyopathy (HCM) is the most common genetic cardiovascular disease9 and is associated with sudden death, especially in young adults.10,11

In a study done by Ogeng’o et al in Nairobi from December 2005 to November 2009, they found that cardiovascular causes comprised 13.2% of all autopsy cases. Among various cardiac pathologies most common was myocardial infarction (18.7%) and next common was cardiomyopathy (17.2%).12

Alessandra Doolan et al did a study in Australia between 1994 and 2002 to know the cause of sudden cardiac deaths in young Australians aged ≤ 35 years. She found hypertrophic cardiomyopathy/ unexplained left ventricular hypertrophy in 15% cases.13

In a study by Eckart RE et al, in America, over a period of 10 years, in 902 cases of adjudicated unanticipated sudden cardiac death, they found that out of total 298 cases of <35 years of age hypertrophic cardiomyopathy was seen in 38 (12.8%) cases while out of 604 cases of age ≥35 years HCM was seen in 19 (3.1%) cases.14

Udayashankar Y, in 2014, reported a case of sudden death of a newly married husband during sexual intercourse due to hypertrophic cardiomyopathy.15 Durakovic Z et al in a retrospective study for a period of 27 years from 1984 to 2010
analysed 69 cases of sudden cardiac death during physical exercise in males in Croatia. Out of total 5 cases of hypertrophic cardiomyopathy, three suddenly died during training and two of them died during recreational physical exercise, probably because of malignant ventricular arrhythmia due to hypertrophic cardiomyopathy. One had an obstructive form of hypertrophic cardiomyopathy and four had non-obstructive form of hypertrophic cardiomyopathy.16

In the present case, hypertrophic cardiomyopathy (HCM) was found in a 19 year old male. He was labourer by occupation and had habit of smoking beedis and drinking alcohol. There was asymmetric hypertrophy of interventricular septum. There was no history of hypertension and valvular pathology in this case. Associated atherosclerotic coronary artery disease was also seen in this case and the person died suddenly due to myocardial infarction. There was no family history of any congenital heart disease.

CONCLUSION

In this case report a young male who was a manual labourer, had chest pain and breathlessness. He died suddenly at workplace. He had habit of smoking beedis and drinking alcohol. During autopsy the heart was massively enlarged. The heart was thick walled with massive myocardial hypertrophy without dilatation of ventricles. There was asymmetric hypertrophy of interventricular septum. It was diagnosed as Hypertrophic cardiomyopathy on histopathological examination of the heart. There was no history of hypertension and valvular pathology in this case. Associated atherosclerotic coronary artery disease was also seen. There was no family history of any congenital heart disease. This case emphasises the requirement of essential investigations and preventive measures to prevent sudden cardiac death in young individuals and their family members due to inheritable cardiac pathologies like this. It also highlights the role of meticulous autopsy and histopathological examination to detect this condition.

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