



SILENT STORM WITH ECHOES OF TRAGEDY UNRAVELING SUDDEN DEATH IN YOUTH – AN IN-DEPTH AUTOPSY CASE STUDY OF HYPERTROPHIC OBSTRUCTIVE CARDIOMYOPATHY

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ABSTRACT

Hypertrophic obstructive cardiomyopathy (HOCM) is a rare but potentially fatal cardiac condition, particularly in young individuals, often presenting with sudden death. This case report presents the autopsy findings of a young adult who succumbed to sudden death, with subsequent diagnosis of HOCM. The deceased, a 19-year-old male, experienced no prior significant cardiac symptoms, making the sudden demise unexpected. Autopsy revealed left ventricular hypertrophy with asymmetric septal thickening, confirming the diagnosis of HOCM. Histological examination demonstrated myocardial disarray and fibrosis, typical of HOCM pathology. Notably, the absence of a family history of cardiac disease complicated the initial diagnosis. This case highlights the importance of considering HOCM in the differential diagnosis of sudden cardiac death, even in individuals without apparent symptoms or family history. Early recognition and appropriate management of HOCM are critical to prevent potentially fatal outcomes in young individuals. Increased awareness among healthcare professionals, particularly in the context of sudden cardiac death, is imperative for prompt diagnosis and intervention.

KEYWORDS : Hypertrophic Obstructive Cardiomyopathy, Sudden Death, Cardiac Pathology.

INTRODUCTION

Hypertrophic Obstructive Cardiomyopathy (HOCM) stands as a complex and multifaceted cardiovascular disorder characterized by the abnormal thickening of the myocardium, most notably within the left ventricle, leading to dynamic obstruction of the left ventricular outflow tract¹. This condition, although relatively rare, presents a significant clinical challenge due to its varied clinical manifestations and propensity for sudden and catastrophic outcomes, such as sudden cardiac death (SCD), particularly in young individuals.

The sudden and unexpected nature of SCD in individuals with HOCM underscores the urgent need for heightened awareness among healthcare professionals regarding the clinical red flags and risk factors associated with this condition. Moreover, it emphasizes the critical role of autopsy examination in elucidating the underlying pathological mechanisms and providing closure for the bereaved family members. Recent studies indicate that the disease is familial in 50% to 60% of cases and Sporadic in the remainder.

The case under examination is of tragic incidence of the sudden demise of 19 years old boy, the cause of which was ultimately determined to be hocm upon comprehensive postmortem examination.

Through a meticulous examination of this case, we aim to not only elucidate the clinical and pathological features of HOCM but also to underscore the importance of proactive screening, risk stratification, and timely intervention in at-risk individuals. By enhancing our understanding of the diagnostic challenges and therapeutic strategies associated with HOCM-related sudden death, we can strive towards more effective prevention and management strategies, ultimately mitigating the devastating impact of this condition on affected individuals and their families

Case report

A 19 years old healthy young boy suddenly fell down while jogging in the morning at about 5:30 am and was suddenly rushed to hospital where he declared dead by Doctors after applying all the resuscitation measures The case was brought to the Mortuary for Medico legal autopsy.

As per history given by relatives, the boy was absolutely

normal since birth.

There was no history of any cardiac disease or medication. Autopsy Findings : Body was average built. Rigor mortis was present all over body and hypostasis was present on back and fixed. No significant marks of injury were present on the body. Other Findings were unremarkable.

Internal examination revealed enlarged globular Heart. Ventricles were hypertrophied (figure 1). Thickness of the right ventricular wall was 1 cm, Left was 3 cm and inter-ventricular septum was irregularly thickened and gritty and hard on cut section.

Few pale, hard and fibrous areas were evident at places. The left ventricular cavity was small. Coronaries were patent and heart valves were normal and healthy. All other internal organs were congested. Microscopic examination of heart revealed extensive myocyte hypertrophy (Figure 2) with hyperchromatic big nucleus. There was haphazard disarray of bundle of myocytes (myofiber disarray). There were interstitial and fibrosis at places. All the findings were consistent with Asymmetric Hypertrophic Cardiomyopathy.

The final cause of death was given as Death due to cardiac failure as a result of Hypertrophic cardiomyopathy



Figure 1 Showing Left Ventricular Hypertrophy

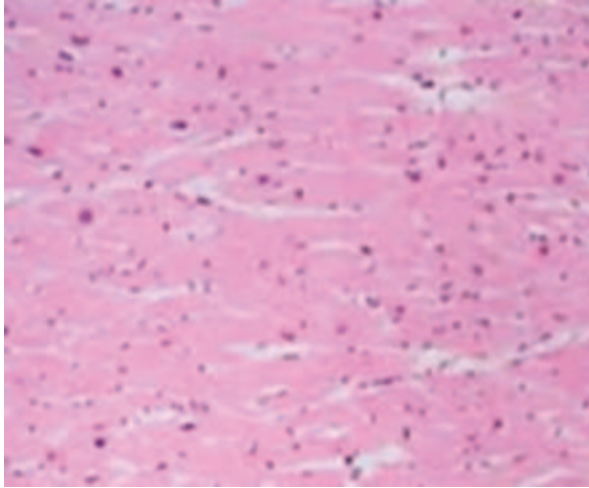


Figure 2 Showing Hypertrophy Of Cardiac Myocyte

DISCUSSION

Numerous names have been given to the illness process over time. This illness has been referred to as hypertrophic obstructive cardiomyopathy, muscular subaortic stenosis, idiopathic hypertrophic subaortic stenosis, and asymmetric septal hypertrophy (ASH). The World Health Organization has named this distinct form of primary muscle hypertrophy—which can occur with or without a dynamic left ventricular outflow tract gradient—hypertrophic cardiomyopathy¹.

A section of the myocardium becomes hypertrophied in HOCM, a primary myocardial disease, for no apparent reason^{2,5}. In Heart muscle hypertrophy is brought on by the growth of the sarcomere, a contractile component of muscles. It also interferes with the heart muscles' electrical activity. Mutations in one of the nine sarcomere proteins are thought to be the cause of familial HOCM, which is inherited as an autosomal dominant characteristic.

The deceased was only sixteen years old. Because of the enlarged myocardium at such a young age, we ruled out other pathologies.

The histology report, which showed widespread myocyte enlargement and a hyperchromatic huge nucleus, was satisfying. There was intermittent and replacement fibrosis along with haphazard disorder of the myocyte bundle (myofiber disarray)^{6,7}. Every result was in line with hypertrophic cardiomyopathy (HOCM). A section of the myocardium becomes hypertrophied in HOCM, a condition for which there is no known etiology. It is the main reason for unexpected deaths in young adults.⁸

According to recent research, this condition may be more prevalent now than it was in the past. Oftentimes, HOCM is asymptomatic until Unexpected cardiac mortality, and as a result, the prevalence of HOCM is 0.2 and 0.5% of the overall population has HOCM.¹⁰

CONCLUSION

The autopsy findings presented in this case report provide crucial insights into the structural and histopathological characteristics of Hypertrophic Obstructive Cardiomyopathy (HOCM), shedding light on the underlying mechanisms contributing to sudden cardiac death in affected individuals. The observed myocardial hypertrophy, dynamic left ventricular outflow tract obstruction, and extensive myocardial fibrosis underscore the complexity of HOCM and its potential for adverse cardiovascular events.

Correlating these findings with existing literature on HOCM reinforces the importance of early detection, accurate

diagnosis, and targeted management strategies in mitigating the risk of sudden cardiac death. The integration of clinical, imaging, and pathological data is essential for risk stratification and personalized treatment approaches tailored to the individual's disease phenotype and prognosis.

Ultimately, this case underscores the imperative for heightened awareness, comprehensive evaluation, and multidisciplinary collaboration in the care of individuals with HOCM. By advancing our understanding of the pathophysiological basis of the disease and its clinical implications, we can strive to improve outcomes and prevent tragic outcomes such as the sudden death observed in this case.

This conclusion encapsulates the significance of the autopsy findings in the case report and highlights the implications for clinical practice and future research in the management of Hypertrophic Obstructive Cardiomyopathy.

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