A Case Series of Hypertrophic Cardiomyopathy (Hypertrophic Obstructive Cardiomyopathy and Dilated Phase of Hypertrophic Cardiomyopathy)

Henry A Mayala¹, Khamis Hassan Bakari², Mark Mayala², Semvua Kilonzo² Waheeda Shokat Kara², Prof. Mohamed Janabi³, Prof. Wang Zhao Hui³

¹Department Of Cardiology, Wuhan Union Hospital, Tongji Medical College Of Huazhong University Of Science And Technology

Abstract

Introduction: Hypertrophic Cardiomyopathy is abnormal asymmetrical thickening of the interventricular septum. It affects 1 in 500 of general population. Hypertrophic Cardiomyopathy may be accompanied by life threatening arrhythmias VT, VF. The clinical presentation of the patients varies according to the stage or type of Hypertrophic Cardiomyopathy.

Case presentation: We present a 63 years old patient a Chinese descent, who presented to Wuhan union hospital with chief complains of awareness of heart beat, blurred vision, and dizziness. He has no history of hypertension, no history of Diabetes mellitus and no history of hyperthyroidism. Clinical and imaging findings are suggestive of hypertrophic obstructive Cardiomyopathy

We also present a 73 years old patient a Chinese descent, who presented to Wuhan union hospital chief complains of productive cough, difficulty in breathing on lying flat and awareness of heart beat. He has no history of hypertension or diabetes mellitus, nor hyperthyroidism. The clinical and imaging findings are suggestive of dilated phase of hypertrophic Cardiomyopathy or end stage hypertrophic Cardiomyopathy.

Conclusion: Hypertrophic Cardiomyopathy is the second most common form of Cardiomyopathy, its presentation is different in different individuals and to diagnose it might be very challenging and may require expertise thus we thought, it will be very important to review this case and share it as it is a common cause of sudden death.

Keywords: HCM-hypertrophic Cardiomyopathy, VT-ventricular tachycardia, VF- Ventricular fibrillation, LVOT-left ventricle outflow tract, ECG and Echocardiography

I. Introduction

Hypertrophic Cardiomyopathy is abnormal asymmetrical thickening of the interventricular septum. There are two types of hypertrophic Cardiomyopathy namely: the non obstructive hypertrophic Cardiomyopathy and obstructive hypertrophic Cardiomyopathy. It affects 1 in 500 of general population and with male predominance and blacks are more affected than white. ¹

Hypertrophic obstructive Cardiomyopathy is a disease characterized by asymmetrical thickening of the interventricular septum extending to the left ventricle outflow tract. During systole, the thickened muscle in the left ventricle outflow tract narrows thus producing obstruction to the left ventricular ejection. The most common symptoms of hypertrophic obstructive Cardiomyopathy are difficulty in breathing, chest pain (angina), dizziness, awareness of heart beat and syncope.²

Hypertrophic Cardiomyopathy runs in families, with an autosomal dominant pattern. It involves mutation in cardiac sarcomeric proteins thus having an effect on cardiac sarcomeric function, currently there more than 27 hypertrophic Cardiomyopathy susceptible genes that are identified.^{1, 3} Hypertrophic Cardiomyopathy is associated with sudden death, and/or a history of sudden death from one of a family member, despite the above fact hypertrophic Cardiomyopathy may be accompanied by life threatening arrhythmias VT, VF and SVT.

In some patient population, hypertrophic Cardiomyopathy may progress to dilated phase of hypertrophic Cardiomyopathy a so called end stage hypertrophic Cardiomyopathy, presenting with dilated left ventricle, impaired left ventricular systolic function mimicking idiopathic dilated Cardiomyopathy, such patients tend to have poor prognosis and might require heart transplant. ^{7,8}

² Department Of Radiology And Nuclear Medicine, Wuhan Union Hospital, Tongji Medical College Of Huazhong University Of Science And Technology

³ Director Department Of Cardiology, Wuhan Union Hospital, Tongji Medical College Of Huazhong University
Of Science And Technology

II. Case Presentation

We present two cases of hypertrophic Cardiomyopathy

Case 1

We present a 63 years old Chinese Male patient, who presented to Wuhan union hospital with chief complains of awareness of heart beat, blurred vision, and dizziness. He has no history of hypertension, no history of Diabetes mellitus and no history of hyperthyroidism. On physical examination the only finding was cold extremities. The vital signs Blood pressure was 110/65mmhg, heart rate = 75b/min, regular, Temperature=37 °C, respiration rate 16 breaths/min. Different investigation were done, the full blood count, renal function test, liver function test, thyroid function test, lipid profile and there were normal Further investigations were done chest radiography, Electrocardiogram and Echocardiogram Chest x-ray was normal ECG revealed Q- waves in leads I and aVL Echo revealed asymmetrical basal septal hypertrophy, dynamic LVOT obstruction with the pressure gradient of 61mmhg across the LVOT with the velocity of 3.9m/s, and diastolic dysfunction with impaired relaxation (E/A <1), normal ejection fraction 65%

Case 2

We also present a 73 years old Chinese male patient, who presented to Wuhan union hospital chief complains of productive cough, difficulty in breathing on lying flat and loss of consciousness for some few seconds. He has no history of hypertension or diabetes mellitus, no history of hyperthyroidism. He was diagnosed of Hypertrophic non obstructive Cardiomyopathy 20 years ago, but was not attending clinics for follow up. On physical examination the only finding was bilateral fine crackles on the inter-scapular regions of the chest. The vital signs: Blood pressure was 110/65mmhg, heart rate =108b/min, regular, respiration rate was 26breaths/min, Temperature was 37 °C Different investigation were done, the full blood count, renal function test, liver function test, thyroid function test, lipid profile and there were normal Further investigations were done chest radiography, Electrocardiogram and Echocardiogram Chest x-ray revealed features of pulmonary edema ECG revealed narrow complex tachycardia which is regular with absent T-waves Supra ventricular tachycardia (SVT) Echo revealed asymmetrical basal septal hypertrophy, dilated left ventricle 6.1cm,impaired LV systolic function with Ejection fraction of 44% and diastolic dysfunction with impaired relaxation (E/A <1), the LVOT velocity was 1m/s with the pressure gradient of 7mmhg indicating no LVOT obstruction

III. Discussion

By comparing the two cases we can see how different the patients might present with hypertrophic Cardiomyopathy depending on which stage (early or late) and type (obstructive on non obstructive), for patient case 1 he presented with hypertrophic obstructive Cardiomyopathy and looked more stable compared to the other one with Hypertrophic non obstructive Cardiomyopathy who presented at the end stage(dilated phase) with all features of heart failure, and further more patients with dilated phase of hypertrophic Cardiomyopathy tend to have poor prognosis and end up to have heart transplant compared to the later. we also learn that both patients are men, concurring with the literature that men are more affected than women and also the importance of echocardiogram which is now the accepted tool for its diagnosis lets no forget that hypertrophic Cardiomyopathy is a second most common type of Cardiomyopathy after dilated Cardiomyopathy, far more worse with the history of sudden death, and running in families with challenges in diagnosing it because it might not present with any symptom in some patients and the history may not be that helpful furthermore it has been diagnosed after an episode of syncope especially in athletes, so we need to have a high suspicion index on its diagnosis and improve our skills especially on echo and as the way the our patients had classical symptoms for hypertrophic Cardiomyopathy and the Echo confirmed it. And the other important thing is the risk of developing arrhythmias such as SVT, VT or VF which are more life threatening making it important for this case series. Our patient 1 is currently on Diltiazem and scheduled for Septal myomectomy and patient 2 is on lasix, aldactone, enalapril on the list of heart transplant.

IV. Conclusion

It's really important to understand the clinical presentation and diagnostic modalities for hypertrophic Cardiomyopathy, as with the current technology and advancement genetic testing is now done to confirm the diagnosis and testing the family members to screen for the disease.

Conflict Of Interest: We declare there is no conflict of interest

Ethical Clearance: Ethical clearance was obtained from Tongji medical college ethical committee

All patient consented for this case report

Author's contributions:

Drafting of manuscript: Henry Anselmo Mayala, Khamis Hassan Bakari, Mark Mayala, Waheeda Shokat Kara, Semvua Kilonzo

Critical Revision: Prof Mohamed Janabi, Prof. Wang Zhao Hui

There was no funding on this research

References

- [1]. Parag Patel, Harry Lever. Hypertrophic Cardiomyopathy,
- [2]. http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/cardiology/hypertrophic-cardiomyopathy/, published April 2014
- [3]. Eugene Braunwald, Costas T, et al. Idiopathic hypertrophic subaortic stenosis: a description of the disease based upon analysis of 64 patients. Circulation 1964; 29:IV-3-IV-119
- [4]. Dao Wen Wang, et al. Hypertrophic Cardiomyopathy Complicated by Severe Bradycardias: A Pedigree Report. Clin. Cardiol. 25, 76–80 (2002)
- [5]. Conolly H M, et al, Echo in Cardiomyopathy, chapter 14 Braunwald's 8th ed. 2008
- [6]. Feigenbaum's 6th ed. chapter 17. 2005
- [7]. Maria-Angela Losi, et al. echocardiography in patients with hypertrophic Cardiomyopathy: usefulness of old and new techniques in the diagnosis and pathophysiological assessment, cardiovascular ultrasound 2010 8:7, BioMed Central
- [8]. Hamada et al clinicl features of dilated pahse of hypertrophic Cardiomyopathy in comparison with those of dilated Cardiomyopathy. Clin Cardiol. 2010 Jul;33(7):E24-8.
- [9]. Goto D et al. clinical characteristics and outcomes of dilated phase of hypertrophic Cardiomyopathy: report from registry data in Japan. J cardiol. 2013 Jan; 61(1):65-70.
- [10]. Ikeda H et al. Prognosis of hypertrophic and dilated Cardiomyopathy. Nihon Rinsho. 2000 Jan; 58 (1): 86-92
- [11]. Yan Xiao et al. Clinical characteristics and prognosis of End stage hypertrophic Cardiomyopathy. Chinese Medical Journal, 2015; Vol.128, issue 11:14831489