

Stress Cardiomyopathy in a Patient with Hypertrophic Cardiomyopathy: Case Presentation and Review of the Literature

Juan M. Vinardell¹, Christos G. Mihos², Adam Nader³, Richard Ro³, Esteban Escolar³ and Orlando Santana^{3,*}

¹Department of Internal Medicine, Mount Sinai Medical Center, Miami Beach, Florida 33140

²Cardiac Ultrasound Laboratory, Massachusetts General Hospital, Harvard Medical School, Boston MA 02114

³Columbia University Division of Cardiology at the Mount Sinai Heart Institute, Miami Beach, Florida 33140

*Correspondence to Orlando Santana: osantana@msmc.com

DOI: 10.31083/j.rcm.2018.02.893

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Stress cardiomyopathy and hypertrophic cardiomyopathy are two distinct entities with different pathophysiologic causes. In the recent medical literature their concurrency has been described. During the acute phase of a stress cardiomyopathy making the diagnosis of a concomitant hypertrophic cardiomyopathy may be challenging, and has important implications in both the acute and long-term clinical management. Herein, we present a case of a stress cardiomyopathy occurring in a patient with hypertrophic cardiomyopathy, along with a review of the literature.

Keywords

Hypertrophic cardiomyopathy; stress cardiomyopathy; hemodynamic instability; myocardial edema; mitral regurgitation

1. Introduction

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant disorder, occurring in approximately 1 in 200 individuals, and is associated with nearly 1,500 identified genetic mutations (Sem-sarian et al., 2015). It is characterized by hypertrophy of the left ventricle in the absence of chamber dilatation, and any other cardiac or systemic condition capable of producing such a degree of hypertrophy (Maron et al., 2014). Stress cardiomyopathy (SCM), on the other hand, is an acquired condition mostly affecting women (90%) in their 7th decade, usually triggered by mental or physical stress, and characterized by a transient left ventricular systolic dysfunction in the absence of obstructive coronary artery disease (Nascimento et al., 2011; Pelliccia et al., 2015). The hypothesized pathophysiology of SCM includes epicardial coronary artery vasospasm or microvascular impairment, catecholamine-induced cardiotoxicity, and neurogenic myocardial stunning (Akashi et al., 2008). Recently, SCM has been reported in cases of HCM. In some, the SCM occurred in patients with a known history of HCM, and in others the diagnosis of HCM was made after the resolution of the left ventricular systolic dysfunction caused by the SCM (Brabham et al., 2011; Daralammoni et al., 2012; Gziut et al., 2012; Egea-serrano et al., 2015; Modi and Ramsdale, 2011; Ochiumi et al., 2015; Patrianakos et al., 2013; Roy et al., 2014; Singh et al.,

2008; Pastor et al., 2015; Miyoshi et al., 2005; Jaber et al., 2006). Herein, we describe a case of a patient with a known history of HCM who developed SCM, along with a review of the literature.

2. Case presentation

A 67-year-old African American female, with an established history of obstructive hypertrophic cardiomyopathy with a left ventricular outflow gradient of 35 mmHg noted on previous echocardiograms. She presented after two hours of atypical chest pain of moderate intensity. The day prior, she was under significant emotional stress due to her financial situation. Her only medication was metoprolol succinate 50 mg daily. There was no history of tobacco, alcohol or recreational/illicit drug use. The physical exam was remarkable for normal vital signs, a 3/6 systolic ejection murmur heard at the left 2nd intercostal space, and the presence of an S4. The Troponin I level was elevated at 1.7 ng/mL. An electrocardiogram revealed normal sinus rhythm, right bundle branch block with left axis deviation, and ST-segment elevations in the precordial leads (Fig. 1). A transthoracic echocardiogram showed a decreased left ventricular ejection fraction of 35% with akinesis of the anteroseptal and apical segments, as well as systolic anterior motion of the mitral valve (SAM) resulting in a left ventricular outflow tract gradient of 35 mmHg, and moderate mitral regurgitation Fig. 2. The coronary angiogram demonstrated normal coronary arteries, a left ventricular ejection fraction of 30%, and left ventricular apical ballooning consistent with a SCM. The hospital course was uneventful, and the patient was discharged on day 3 of hospitalization. A transthoracic echocardiogram performed two weeks after discharge showed a hypertrophic cardiomyopathy with a left ventricular ejection fraction > 65%, and SAM causing a left ventricular outflow tract gradient which was unchanged when compared with the study performed in the inpatient setting and to her previous echocardiograms (Fig. 3).

3. Discussion

HCM, on echocardiographic evaluation, is characterized by a maximal left ventricular wall thickness > 15 mm, with almost one third having wall thickening localized to a single segment, which

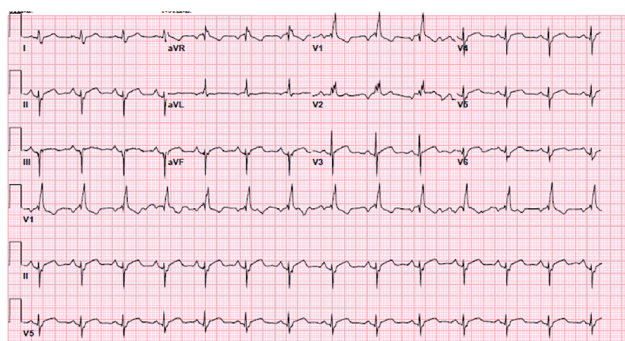


Figure 1. Electrocardiogram Demonstrating ST-segment Elevation in V3-V6 Precordial Leads.



Figure 2. The Apical Ballooning, Hypertrophic Septum with Systolic Anterior Motion of the Mitral Valve And a Left Ventricular Outflow Tract Gradient of 35 mmHg Assessed by Apical Four-chamber View of Transthoracic Echocardiogram.

may be isolated to the apex, in the absence of any identifiable cause for the hypertrophy (Maron et al., 2014; Silbiger, 2016). As many as 70% of patients with HCM may have SAM causing a dynamic obstruction of the left ventricle outflow tract, which may be accompanied by anatomic abnormalities of the mitral apparatus (Silbiger, 2016). Conversely, the most common finding in SCM is a transient apical hypokinesis with hyperdynamic motion of the basal myocardial segments, which may lead to the development of left ventricular outflow tract obstruction due to SAM in up to 25% of patients (El Mahmoud, 2008). The presence of SAM in either cardiomyopathy may be associated with significant mitral regurgitation, which predisposes to volume overload of the left ventricle, and in the acute setting can trigger hemodynamic instability. Therefore, in the acute phase of a SCM it may be difficult to differentiate the two entities, and a high index of clinical suspicion is warranted.

A search of the medical literature rendered 14 cases in which SCM occurred in the context of HCM (Brabham et al., 2011; Daralammori et al., 2012; Gziut et al., 2012; Egea-serrano et al., 2015; Modi and Ramsdale, 2011; Ochiumi et al., 2015; Patrianakos et al., 2013; Roy et al., 2014; Singh et al., 2008; Pastor et al., 2015;

Miyoshi et al., 2005; Jaber et al., 2006). Excluding the present case, 11 were female, the age ranged from 43 to 85 years, all had an abnormal electrocardiogram suggestive of myocardial ischemia, and the most common presenting symptoms were chest pain (N = 9) and dyspnea (N = 3), with 2 patients having both chest pain and dyspnea. The data were limited in that there was no uniform reporting of the triggering events of the SCM, or severity of SAM and/or mitral regurgitation. In the patients where the cardiac biomarkers were reported, all were elevated, and the mean left ventricular ejection fraction on presentation was 40%. A history of HCM was reported in 8 of the cases, and in the rest the diagnosis of HCM was made after the resolution of the SCM. Left ventricular outflow tract obstruction was evaluated in 9 cases, 8 of which demonstrated increased gradients that improved or resolved at follow-up. Complete resolution of the left ventricular systolic dysfunction was seen in all of the cases where follow-up was obtained. In total, 11 patients had HCM consisting of asymmetrical septal hypertrophy, and 3 had apical HCM as shown in Table 1.

As previously mentioned, during the acute phase of SCM it is challenging to differentiate the two entities, especially the apical variant of HCM, since in those cases the disease process may be entirely masked by apical thinning or aneurysm caused by the SCM (Roy et al., 2014). The clinical scenario is further confounded in that there have been reports of a recently detected phenomenon of apparent left ventricular apical hypertrophy seen during the subacute and chronic phases of a resolving SCM, in patients without apical HCM (Izgi et al., 2015). In these circumstances the apical hypertrophy is due to transient myocardial edema, which can be detected utilizing cardiovascular magnetic resonance imaging (Izgi et al., 2015). Differentiating between left

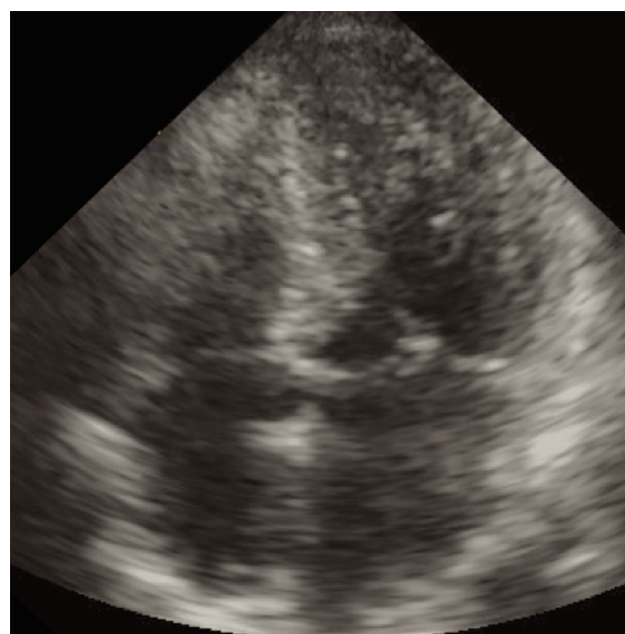


Figure 3. Transthoracic Echocardiogram Apical Four-chamber View Performed Two Weeks After Discharge Revealing a Hypertrophic Septum with Systolic Anterior Motion of the Mitral Valve and an Ejection Fraction of 65%.

Table 1. Cases of Stress Cardiomyopathy in Patients with Hypertrophic Cardiomyopathy.

Authors	Age (years)	Gender	Triggers	Symptoms	History of HCM	Trop I	Ejection Fraction(%)	LVWT (mm) [s or a]	LVOT Rest /Valsalva gradients (mmHg)	follow up			
										Time to follow up	Ejection Fraction (%)	LVOT gradient (mmHg)	LVWT (mm)[s or a]
Brabham et al (Brabham et al., 2011)	48	Male	No	Chest pain, dyspnea	No	H	35	20[s]	96/105	1 year	66	27/95	19[s]
Daralammori et al (Daralammori et al., 2012)	70	Female	NR	Dyspnea	No	H	35	24[s]	NR	3 months	NR	20/70	24[s]
Gziut et al (Gziut et al., 2012)	74	Female	NR	Chest pain	Yes	H	40	22[s]	NR/90	3 months	55	NR/75	NR
Egea-Serrano et al (Egea-serrano et al., 2015)	51	Male	No	Dyspnea	Yes	H	43	19[s]	85/150	15 days	Normal	7/NR	NR
Modi et al (Modi and Ramsdale, 2011)	54	Female	NR	Chest pain	No	H	NR	19[s]	NR	3 days (deceased)	-	-	-
Ochiumi et al (Ochi-umi et al., 2015)	84	Female	Mental stress	Chest pain	Yes	H	NR	NR	158/NR	10 days	NR	Improved	NR
Patriakanos et al (Patrianakos et al., 2013)	49	Male	Alcohol intoxication	Chest pain	Yes	H	30	18[s]	Present but not quantified	2 days	NR	Present but not quantified	NR
Roy et al (Roy et al., 2014)	43	Female	Alcohol abstinence	Confusion, dyspnea	No	H	47	NR	NR/NR	2 weeks	69	NR	13 [a]
Singh et al (Singh et al., 2008)	70	Female	NR	Chest pain, dyspnea	No	H	39	NR	NR/NR	1 month	65	NR	14 [a]
Pastor et al (Pastor et al., 2015)	79	Female	NR	Chest pain	Yes	H	25	22[s]	20/NR	2 months	NR	Less severe	NR
Miyoshi et al (Miyoshi et al., 2005)	85	Female	NR	Chest pain	Yes	NR	Low	14	NR/120	3 months	Normal	NR/100	NR
Jaber et al (Jaber et al., 2006)	65	Female	Death of spouse	Chest pain	Yes	NR	67	12[s]	No gradients	NR	NR	NR	NR
	65	Female			Yes	H	NR	20[s]	40/NR	NR	NR	Present Unchanged	

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ventricular hypertrophy due to apical HCM or SCM-induced myocardial edema can be achieved by observing subsequent electrocardiograms for markedly increased precordial and standard lead voltages and chronically persisting giant negative T waves, and by comparing old and follow up echocardiograms (Madias, 2014; Maron, 2001). There is no conflict of interest to disclose from the author and co-authors.

In conclusion, it is important to consider coexistent HCM within the differential diagnoses in patients presenting with SCM, which requires a high index of suspicion. HCM with a baseline SAM and mitral regurgitation may significantly worsen in the acute phases of SCM, given the hyperdynamic function of the basal myocardial segments, which may precipitate hemodynamic instability. Finally, while SCM typically resolves fairly quickly, patients with HCM require much closer clinical surveillance and risk stratification for sudden cardiac death and possible prophylactic implantable cardioverter defibrillator.

Acknowledgement

Thanks to all the peer reviewers and editors for their opinions and suggestions.

Conflict of Interest

The authors declare no competing interests.

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