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APICAL ANEURYSM OF THE LEFT VENTRICLE IN ATYPICAL HYPERTROPHIC CARDIOMYOPATHY AFTER ACUTE MYOCARDIAL INFARCTION IN POSTMENOPAUSAL WOMAN: A CASE REPORT

ANEURISMA APICAL DO VENTRÍCULO ESQUERDO NA CARDIOMIOPATIA HIPERTRÓFICA ATÍPICA APÓS INFARTO AGUDO DO MIOCÁRDIO EM MULHER PÓS MENOPAUSADA: RELATO DE CASO

ATYPICAL HYPERTROPHIC CARDIOMYOPATHY IN WOMAN

Descriptors: Case Report (Relato de Caso), Cardiomyopathy Hypertrophic (Cardiomiopatia Hipertrófica), Aneurysm (Aneurisma), Myocardial Infarction (Infarto do Miocárdio).

ABSTRACT

We report an atypical case of a 65-year-old patient with hypertrophic cardiomyopathy (HCM), ventricular media and apical aneurysm, associated with Coronary Artery Disease involving the anterior descending coronary artery, with acute myocardial infarction, primary percutaneous revascularization and intracoronary stent implantation, which represented a diagnostic and therapeutic challenge. Patient with hypertension, diabetes, dyslipidemia and asthma, three months before the interview, present symptoms of Heart Failure, compatible with functional class III of the New York Heart Association, even with optimized therapy.

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The echocardiogram also demonstrated the presence of elevated intraventricular pressure gradient between the aneurysm region and the ventricular chamber (around 76 mmHg). Magnetic resonance imaging confirmed the presence of mean HCM of the left ventricle, "trapping" the contrast in the aneurysm region. The wall thickness of the middle segment of the lower septum measured 20 mm in the diastole and lateral infero- 15 mm with myocardial fibrosis area affecting the apex. Treatment options for individuals not responsive to optimized therapy are: surgical aneurysmectomy and cardioverter pacemaker and defibrillator implantation. Both would reduce the intraventricular gradient. However, the surgical procedure presents a high risk, even though the pacemaker implantation would not apply, because the patient did not develop complex arrhythmias and it is not certain whether it would reduce the intraventricular gradient, one of the factors responsible for the probable symptoms.

RESUMO

Relata-se um caso atípico de uma paciente de 65 anos, portadora de cardiomiopatia hipertrófica, forma médio ventricular e aneurisma apical, associado a Doença Arterial Coronariana envolvendo a artéria coronária descendente anterior, com infarto agudo do miocárdio, revascularização percutânea primária e implante de “stent” intracoronário, que representou um desafio diagnóstico e terapêutico. Portadora de hipertensão arterial, diabetes,

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dislipidemia e asma, há três meses da entrevista, apresenta sintomas de Insuficiência Cardíaca, compatíveis com classe funcional III da *New York Heart Association,* mesmo com terapia otimizada. O ecocardiograma demonstrou ainda a presença de gradiente de pressão intraventricular elevado entre a região do aneurisma e a câmara ventricular (em torno de 76 mmHg). A Ressonância Nuclear Magnética confirmou a presença de CMH média do ventrículo esquerdo, "aprisionando" o contraste na região do aneurisma. A espessura da parede do segmento médio do septo inferior mediu 20 mm na diástole e ínfero- lateral 15 mm com área de fibrose miocárdica acometendo o ápice. As opções de tratamento para indivíduos pouco responsivos à terapia otimizada são: aneurismectomia cirúrgica e o implante de marcapasso cardioversor e desfibrilador. Ambos reduziriam o gradiente intraventricular. Entretanto, o procedimento cirúrgico apresenta um risco elevado conquanto que o implante do marca-passo não se aplicaria, pois, a paciente não desenvolveu arritmias complexas e não está certo se reduziria o gradiente intraventricular, um dos fatores responsáveis pelos prováveis sintomas.

INTRODUCTION

An apical aneurysm is recognized, echocardiographically, as a region of the left ventricle with a very tapered wall, with very distinct margins and that presents dyskinetic movement during ventricular contraction[1]. In patients with Hypertrophic Cardiomyopathy

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(HCM) and subaortic obstruction, the most frequent form of presentation of this heart disease, the presence of an apical aneurysm is very rare (approximately 2-5%)[2,3]. This association has been reported in 15% of cases with HCM, apical form, and about 28% with midventricular HCM[1,2,4]. The existence of apical aneurysm in HCM is related to a worse prognosis, including heart failure not responsive to optimized treatment, malignant ventricular arrhythmias, thromboembolic events, and sudden death[5]. The association of HCM and obstructive coronary atheromatous disease adds a higher risk of complications and makes it difficult to choose the best treatment option in these cases[6,7]. We report a case of a 67-year-old woman who developed an apical aneurysm of the left ventricle after acute myocardial infarction, with HCM. All relevant data are available in the article and Supplemental Material. This case report follows CARE Guidelines[8] and was approved by the Ethics and Research Committee of UNIMES - CAAE: 19690619.9.0000.5509.

NARRATIVE

Patient Information

The patient is a woman, born in 1954 (67 years), white, housewife. She is a smoker, hypertensive, diabetic, dyslipidemic, asthmatic, and sedentary. She has a weight of 78kg, height of 1.58m, body mass index of 31.2, body surface 1,797m^2. She has HCM, a ventricular medium form with apical aneurysm, associated with coronary atheromatous disease involving the anterior descending coronary artery, with acute myocardial infarction, primary percutaneous revascularization, and intracoronary stent implantation.

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Clinical Findings

1st Hospitalization: At the age of 60 years old, she had an episode of segmental obstruction of 70% of the proximal third of the anterior descending coronary artery and performed a percutaneous coronary intervention with pharmacological coronary stent implantation.

2nd Hospitalization: Nine months later, at the age of 61 years old, she was hospitalized with precordial pain in tightening at rest, with irradiation to the left upper limb and mandible.

On physical examination (PE), she presented a good general condition, acyanotic, anicteric and afebrile, hearted, hydrated and eupneic. The blood pressure (BP) values were 130x65mmHg, heart rate (HR) 60 beats per min (bpm), oxygen saturation ($SatO_2$): 97%.

An echocardiogram (ECHO) was performed and presented the following results shown in Table 1. It was concluded that there was hypokinesia in the middle and basal segment of the lateral inferous wall and dyskinesia of the apical portion of the LV, characterizing a grade I diastolic dysfunction.

3rd Hospitalization: Three years later, at the age of 64 years old, she returned to the emergency room complaining of precordialgia with irradiation to the back and left upper limb and dyspnea to moderate efforts.

In the PE, she had regular general condition, acyanotic, anicteric, and afebrile, anchored +/4+, hydrated and eupneic. BP 80x40 mmHg, HR 120 bpm, $SatO_2$ 93%. They

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performed an ECHO (Table 1) that concluded the presence of akinesia of the apical segment of the septum and anterior wall with slight apical dyskinetic movement. There was a change in the diastolic filling of the LV. The aortic valve exhibited a slight reflux without hemodynamic repercussion. Then, the patient presented alterations in segmental contractility with normal LV function, characterizing grade I diastolic dysfunction.

Catheterization was also performed showing tortuosity in the anterior descent artery that was free of obstructive lesions. The stents in the middle third were pervios and had no sign of restenosis.

Evaluation and Diagnosis

At 65 years of age, the patient returns for routine ECHO and revealed akinesia of the apical segment of the septum and anterior LV wall with the slight dyskinetic movement of the apical region, characterized by an apical aneurysm (Fig 1). In the mean region of the LV, there was a greater thickening of the myocardium with systolic narrowing, producing a dynamic intraventricular gradient of up to 62 mmHg, which may be compatible with HCM, an atypical form. The diastolic filling of the VE was altered type ventricular compliance and the mitral and aortic valves exhibited mild reflux.

It was concluded that the patient presents atypical HCM, mitral and aortic insufficiency of discrete degree, alterations in segmental contractility with normal LV function (apical aneurysm), and grade II diastolic dysfunction.

To confirm the diagnostic hypothesis, a magnetic resonance imaging study of the

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heart was requested which was compatible with asymmetric HCM of the VE, especially distributed in the midventricular portion, with an important decrease in the ventricular cavity in the systole, without the involvement of the basal portion and LV outflow tract. The infero-septal wall measured 20 mm in diastole (Fig 2) and lateral infero- 15mm. The presence of regional alteration of the myocardial contractility of the LV was observed, with chromium dyskinesia, configuring an apical aneurysm. In addition to mitral insufficiency and myocardial fibrosis area affecting the entire apex.

Diagnostic challenges

Her initial clinical diagnosis was only acute myocardial infarction and coronary atheromatous syndrome. Due to attention was not given, in previous hospitalizations and interventions, the possibility of being HCM. As a result of the primary diagnosis, however, our patient did not seek an additional evaluation for a long time because she believed that there was no treatment for her condition, so the delay in diagnosis resulted in a severe gradual deterioration of our patient.

Therapeutic Interventions

Treatment options for individuals not responsive to optimized therapy are surgical aneurysmectomy and cardioverter pacemaker and defibrillator implantation. Both would reduce the intraventricular gradient. However, the surgical procedure presents a high risk for our patient (Euro score: 56%) and the pacemaker implantation would not apply because it did not develop complex arrhythmias and it is not certain whether it would reduce the

intraventricular gradient, one of the factors responsible for the probable symptoms. That is, both surgery and pacemaker would not provide a significant functional benefit.

Monitoring

Since the last routine consultation, the patient continues with dyspnea at small and medium efforts. She is being treated for heart failure, compatible with functional class III of the New York Heart Association, with optimized therapy. Because it is an extremely rare case with a high risk of mortality for her, so far there is no consensus on what the best procedure for our patient is.

DISCUSSION

HCM is a genetic disease characterized by the disorganization and hypertrophy of cardiac myocytes, interspersed with areas of interstitial fibrosis, with a prevalence between 0.16 and 0.29% (≈1:625–1:344) of the general population[9]. It is usually asymmetric, with greater involvement of the basal interventricular septum. Other regions of the LV may be compromised such as the apex, middle section, and inferolateral wall. The LV, in general, is with normal volume and normal or increased ejection fraction[9].

Most cases of HCM are asymptomatic. However, a subgroup may develop symptoms such as fatigue, dyspnea, chest pain, palpitations, and syncope[9].

Chest pain presents aspects very suggestive of angina pectoris, and electrocardiogram may recall acute myocardial infarction even in the absence of coronary atheromatous disease,

confusing diagnoses. In some patients, when these conditions may be coexisting (coronary atheromatous disease and HCM), morbidity and mortality become very high (prevalence of 23.3%)[10].

Aneurysms involving the apical region of the LV may arise in patients with HCM, associated or not with the coronary atheromatous disease. Particularly in the media ventricular and apical forms (15 to 28%)[1,2,4], this finding is associated with adverse results including sudden cardiac death, complex ventricular arrhythmias, and heart failure[11].

Patients with medio ventricular HCM and apical aneurysm, in general, present high intraventricular gradient and reduction of the effective volume of LV, a situation that promotes a "vicious circle", dilating the apical region and further reducing the systolic volume, exactly as it occurs in our patient.

CONCLUSION

Our patient meets the diagnostic criteria by ECHO and Nuclear Magnetic Resonance tests for HCM, but it is an atypical form. In addition, after myocardial infarction, the patient developed an apical aneurysm, making her case extremely rare in the medical literature. There must be still many studies based on this pathology so that we can always offer the best for our patients.

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Supplemental Material

Table 1 - Echocardiogram values during 2nd, 3rd hospitalization and routine examination

Echocardiogram	2*nd* hospitalization	3 *rd* hospitalization	Routine examination	Reference Values
Right Cavities	Normal	Normal	Normal	Normal
Ventricular Septum Movement	Normal	Normal	Normal	Normal
Aorta	29mm	30mm	37mm	20-38mm
Left Atrium	29mm	38mm	37mm	20-40mm

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Diastolic diameter of the Left Ventricle	47mm	55mm	48mm	35-56mm
Systolic Diameter of the Left Ventricle	27mm	37mm	29mm	20-40mm
Diastolic thickness of the Ventricular Septum	9mm	7mm	9mm	7-11mm
Diastolic thickness of the posterior wall	9mm	7mm	9mm	7-11mm
Ejection Fraction	0,72	0,60	0,66	>0,55
Left ventricular mass	149 gr/m^2	161 gr/m^2	141 gr/m^2	<95gr/m^2
Mitral Valve	No changes	No changes	No changes	No changes
Aortic Valve	No changes	No changes	No changes	No changes
Tricuspid Valve	No changes	No changes	No changes	No changes
Pulmonary Valve	No changes	No changes	No changes	No changes

Source: Larissa Gomes Peres Bomfim

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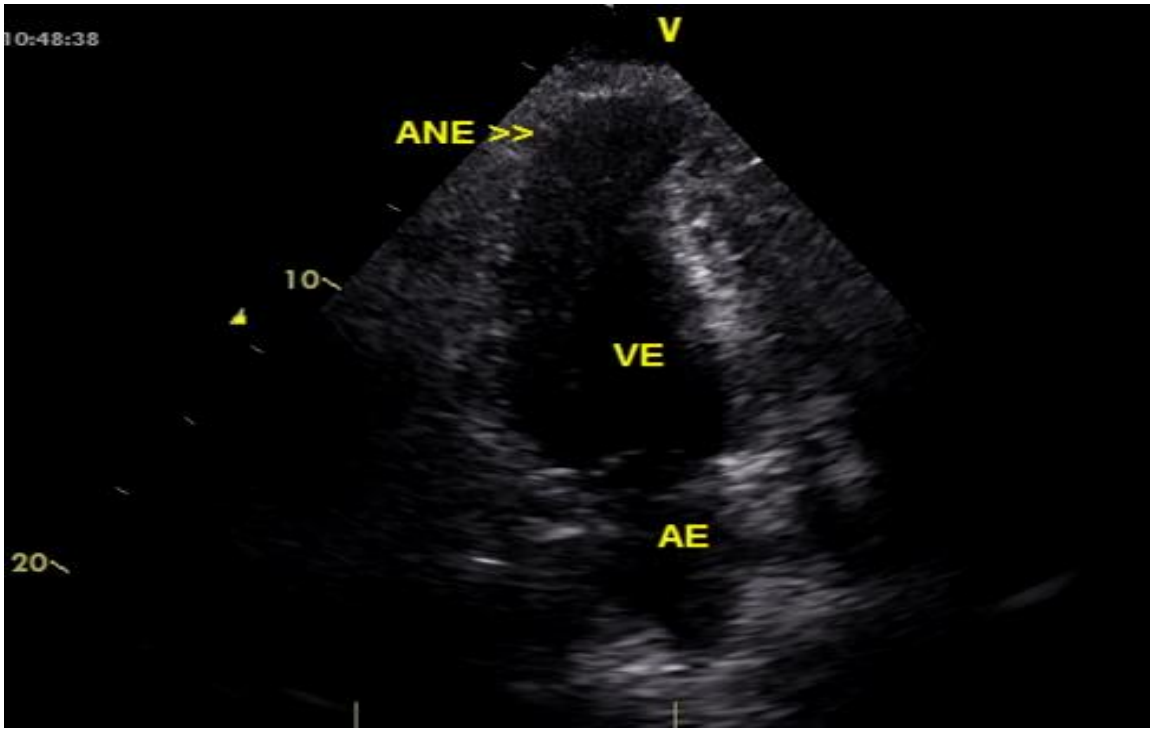
Fig 1- Echocardiogram evidencing atypical Hypertrophic Cardiomyopathy with apical aneurysm of the left ventricle



Source: Larissa Gomes Peres Bomfim

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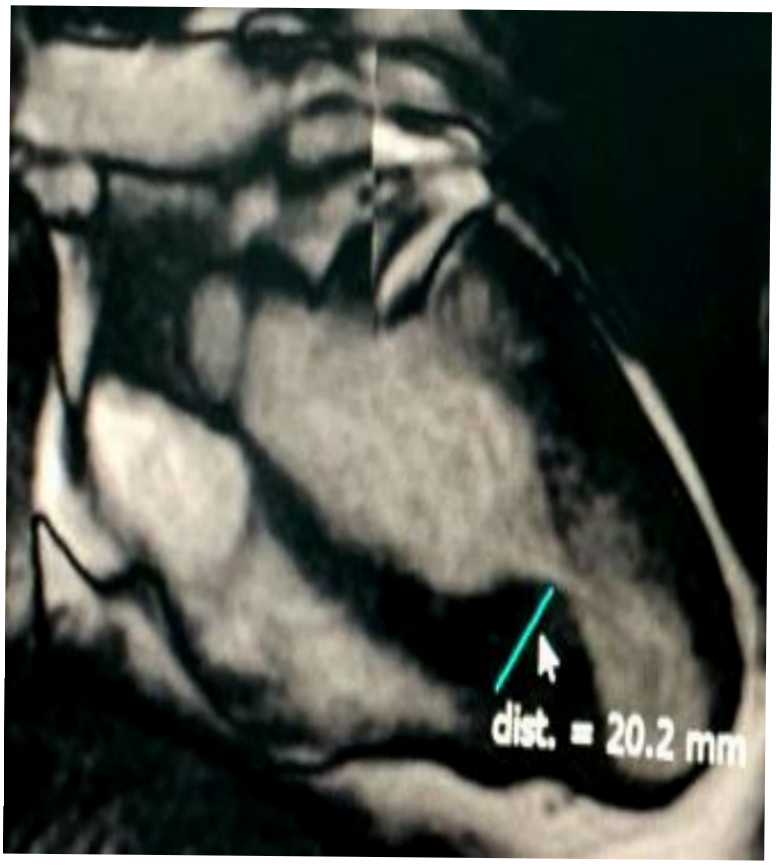
Fig 2- Magnetic Resonance: thickness of the lower septum wall



Source: Larissa Gomes Peres Bomfim