Author's response to reviews

Title: New Insight into Treatment of Mid-apical Obstructive Hypertrophic Cardiomyopathy: Transapical Myectomy

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Author's response to reviews:

July 29th, 2014.
Michael Kidd
Editor-in-Chief,

Journal of Medical Case Reports

Dear Editor,

Attached please find the manuscript entitled: “New Insight into Treatment of Mid-apical Obstructive Hypertrophic Cardiomyopathy: Transapical Myectomy”, and responses to reviewer’s comments.

The manuscript has been edited for spelling, grammar, style, syntax, sentence structure, punctuation, and conciseness and formatted according to journal guidelines.

Sincerely,

Thiago Luis Scudeler, MD.
Whady Hueb, MD, PhD.
Heart Institute, University of Sao Paulo

Author's responses to reviewer

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Authors:

Thiago Luis Scudeler (tlscud@ig.com.br)
Paulo Cury Rezende (rezendepaulo@hotmail.com)
Reviewer's report:

General

The case presented is nicely depicted. However, in my opinion, some important information are missing.

Dear Reviewer,

Thank you very much for your careful analysis of the manuscript. I am sure that all questions were valuable and the changes made contributed to the improvement of this case report.

Question 1

Does the patient have a restrictive filling pattern?

Good point. The echocardiograms showed end systolic and diastolic volumes within the range of normality. This may have happened because the hypertrophic myocardium was confined only to the mid-apical portion of the left ventricle.

Question 2

Did the authors require a genetic test to ascertain the diagnosis? They stated that the hypertrophy was only mild and concentric in 2009 and then was present and moderate after only one year, and was also associated with apical aneurysm. This feature is unusual for HCM, a condition in which the phenotype usually develops over many years.

Excellent question. The diagnosis in this case was based on clinical symptoms, echocardiographic images, and by cardiac magnetic resonance imaging. In addition, the diagnosis was also ascertained by macroscopic characteristics confirmed by the surgeon at the time of the procedure and microscopically by histological analysis. Based on all of these findings, the diagnosis was clear, and
we therefore did not perform a genetic test. The image of the apical aneurysm may be caused by the presence of chronic occlusion of the LAD.

Question 3
The surgery procedure described is rarely performed because of its technical difficulty. Adverse remodeling of the left ventricle tends to develop after time, and may cause left ventricular dysfunction with worsening of symptoms and functional status. It would be interesting to know how many years/months of follow-up they have on this patient.

Good question. The patient had complete remission of symptoms after surgery and therefore reassumed her daily activities after recovery from surgery. One and half years after surgery, the patient was free of symptoms caused by cardiomyopathy.

Question 4
Some comorbidity such as obesity and hypertension may influence the phenotype expression of a genetic-determined cardiomyopathy. Do the authors think that the patient’s comorbidity described may have affected the cardiac morphology and function?

You are right. These comorbidities, especially hypertension may influence the phenotype expression of hypertrophic cardiomyopathy, but this is very difficult to prove. Unlike hypertrophic cardiomyopathy, hypertrophy caused by hypertension regresses when the patient achieves adequate control of blood pressure levels. Our patient was well medicated, with well-controlled pressure.

Question 5
Did the patient undergo transesophageal echocardiography and cardiopulmonary test before surgery?

Unfortunately, these procedures were not performed. Only transthoracic echocardiography was accomplished. Intraoperative transesophageal echocardiographic control was performed and decreased apical hypertrophy was observed, with increased volume of the left ventricular cavity and absence of papillary muscle dysfunction.

Question 6
Which medical treatment did the authors try before surgery?

Excellent point. Before surgery, this patient received optimal medical therapy, with the use of beta-blockers and calcium-channel blockers, at maximum tolerated doses and nitrates. Her blood pressure was controlled, reaching levels lower than 140 x 90 mmHg, and all the other comorbidities were also controlled. Despite this, the patient still had limiting symptoms, so that she opted for cardiac surgery.

Question 7
Did they consider an ICD for this patient with a high-risk subtype of HCM? If not,
why?

Excellent question. We did not consider an ICD for this patient, despite her having a high-risk subtype of HCM, because demand for this device at our institution is great compared with capacity to provide it. Currently, we have a waiting list of more than 1 year for an ICD implant. However, as the patient is a candidate for an ICD implant, she was indicated to be evaluated for this device.