Author’s response to reviews

Title: A case of hypertrophic cardiomyopathy combined with muscular ventricular septal defect and abnormal origin of right coronary artery

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Author’s response to reviews:

Dear Editor:

We thank you, the Associate Editor, and the two reviewers for carefully evaluating our manuscript and providing valuable comments and suggestions. We have addressed the reviewer’s concerns by providing further explanation for some of the observation procedures and results that were not stated clearly in the initially submitted manuscript. The revisions are shown in red-colored words. We hope that the revised manuscript is acceptable for publication in BMC Cardiovascular Disorders.

Sincerely yours,

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Reviewer #1:

Concern #1: I have a question. Some authors have reported that left ventricular crypts (clefts) were often observed in HCM patients. [Reference 1, 2] Crypts were defined as narrow and deep blood-filled invaginations contiguous with the left ventricular cavity extending ≥ 50% of wall thickness. What is the difference of the crypt and incomplete spontaneous closure of muscular VSD that you have reported? Or, is your case classified into myocardial crypt which has an extremely deep cavity?

References:


Response: Thanks for your great question. I have carefully read the references recommended by reviewers. The literatures indicated that myocardial crypts (clefts) can occur in the myocardium of patients with either hypertrophic cardiomyopathy, or carriers of cardiac hypertrophy related genes without clinical phenotype, most of which were observed by magnetic resonance imaging, and rarely found by ultrasound. Morphologically, the crypts (clefts) in the ventricular septal myocardium are not much different from the VSDs due to incomplete spontaneous closure. However, in our case, the features of 2D and color Doppler Echocardiography of this "crypt" are similar to the muscular VSD, allowing us to diagnose it as a muscular VSD with incomplete spontaneous closure. Of interest, the presence of myocardial crypts (clefts) can be highly correlated with the genotype of HCM, but the mechanism underlying the formation of myocardial crypts remains unclear. Combined with our case, we speculated that the clefts in the interventricular septum (or part of them) could be the remnants of the VSDs during spontaneous closure, which could be associated with following reasons. At first, the possible mechanisms of spontaneous closure of muscular VSD are: cardiac hypertrophy, fibrous tissue hyperplasia, etc. [1]. In agreement with the pathological changes of hypertrophic cardiomyopathy, the presence of myocardial crypts (clefts) was highly correlated with HCM. And the process of spontaneous closure of muscular VSD may promote the progression of myocardium hypertrophy. Secondly, Dasgupta S [2] proposed another possible mechanism for closure of muscular VSD: right ventricular endocardial tissue and proliferation and coverage. It was reported that a 4 mm
midmuscular VSD with a left-to-right shunt in a 2-month-old female infant was found by echocardiography. 3 years later, a repeat ECHO showed the defective right ventricular side was closed, but there was still a 4 mm defect in the left ventricular side. Left-to-right shunt communicating with the right ventricular cavity was not observed. At last, Tikanoja T [3] has reported that the incidence of muscular VSD was higher in children with HCM, rare in adults based on clinical evidences. However, these clinical observations could not be excluded from the possibility of septal myocardial hypertrophy in adults, apart from the spontaneous closure of most VSDs. With the development of modern diagnostic instruments, the remnants of these VSDs with incomplete spontaneous closure (right ventricular side closure only) could be more easily detected.

References


Concern #2: On Movie 1, systolic anterior motion of mitral valve or chordae without septal contact was observed. Was the left ventricular outflow tract obstruction provoked under Valsalva maneuver, sitting, or standing position, because the present patient complained of chest pain?

Response: Thank you for your meticulous observation. The left ventricular outflow tract obstruction was not observed at rest. Unfortunately, we did not perform a stress echocardiography or an excitation test on him. In recent months, we have tried to contact the patient to have an examination of Echocardiography again, but he was reluctant to come to the hospital due to preparing his wedding.
Concern #3: In the explanation of Figure 1, you've used the expression 'the left ventricle anterior lateral wall leads' or 'on the leads of left ventricle anterior lateral wall', but it may be better, 'on the leads of left ventricular anterolateral wall ' or other expression.

Response: Thank you very much for the great suggestion. We have made the change.

Concern #4: In several places, you've used the expression 'sinus of valsalva', but it may be better to spell 'sinus of Valsalva'.

Response: They are all changed as you suggested.

Reviewer #2:

Concern #1: General comment: This is a nicely described clinical case of a patient with HCM and associated congenital lesions (septal defect+anomalous RCA) with great descriptive images. That is the good thing; the bad thing is that it requires deep and comprehensive revision of the language

Response: Thank you for your encouragement. We have comprehensively revised the language.


Response: We have added this reference in the revised paper.

Concern #3: ADDITIONAL REQUESTS/SUGGESTIONS: English needs revision

Response: The revised manuscript has been polished by a professional English expert.