Surgical Single Stage Treatment for Obstructive Hypertrophic Cardiomyopathy and Aortic Arch Aneurysm

Davide Margonato, MD1,2  Valerio Stefano Tolva, MD, PhD3  Giuseppe Vaccari, MD4  Paolo Bianchi, MD3  Renato Casana, MD5  Gianfranco Parati, MD, FESC6  Paolo Ferrazzi, MD4

1 Department of Cardiology, Policlinico di Monza, Monza, Italy  
2 Department of Cardiology, Fondazione Policlinico San Matteo, Pavia, Italy  
3 Department of Vascular Surgery, Policlinico di Monza, Monza, Italy  
4 Department of Cardiac Surgery, Policlinico di Monza, Monza, Italy  
5 Vascular Surgery Research Laboratory, Istituto Auxologico Italiano, Milano, Italy  
6 Cardiology Unit and Department of Cardiovascular, Neural and Metabolic Science, Istituto Auxologico Italiano, S. Luca Hospital, Milano, Italy

AORTA 2020;8:144–147.

Abstract

Coexistence of obstructive hypertrophic cardiomyopathy and severe aortic pathology is extremely rare; nonetheless, the association between these two diseases is fascinating. Here we present a unique case report of a patient with obstructive hypertrophic cardiomyopathy and aortic arch aneurysm treated by a single surgical procedure.

Keywords

- hypertrophic cardiomyopathy
- aortic arch aneurysm
- aortic replacement
- aortic stent graft
- myomectomy
- myectomy

Introduction

Hypertrophic cardiomyopathy (HCM) is the most common heritable cardiovascular disease, affecting 1 out of 500 people. It is characterized by asymmetric left ventricular (LV) hypertrophy in the absence of a secondary cause, and by dynamic obstruction of the LV outflow tract (LVOT). Aortic dilation is a major cause of morbidity and mortality, and its association with obstructive HCM (OHCM) has recently been investigated because of common signaling pathways that may play a significant role in both diseases. We present a unique case report of a patient affected by OHCM and saccular aortic arch aneurysm treated by surgical septal myectomy, mitral valve plasty, and ascending aortic replacement, as well as aortic arch endoprosthesis with concomitant upper trunk rerouting during the same surgical procedure.

Case Presentation

A 65-year-old male with a history of OHCM was admitted to our cardiology department for elective surgical septal myectomy. OHCM was diagnosed in 2012 and treated with β-blockers; however, in the last year, he developed worsening dyspnea (NYHA class III [New York Heart Association functional classification]), exercise intolerance and atrial fibrillation. Shortly before, admission chest computed tomography (CT) scan disclosed a thrombosed sacciform aortic arch aneurysm, small saccular lesion of the proximal descending aorta, and an infrarenal abdominal aortic aneurysm (Fig. 1A).

Preoperative echocardiography showed interventricular septal hypertrophy (both anterior and posterior, 19 and 17 mm, respectively, Fig. 2A and B), systolic anterior motion of the mitral valve with LVOT obstruction (basal LVOT obstruction).
gradient 33 and 77 mm Hg after Valsalva’s maneuver, ►Fig. 2C), and normal biventricular contractility. Coronary angiography revealed no significant obstruction.

We decided to approach both the lesions (OHCM and aneurysm) concurrently during the same surgical procedure. We performed standard transaortic anterior septal myectomy for OHCM enriched by various interventions on the subvalvular mitral apparatus according to our experience in this technique,4 through resection of fibrous structures connecting the papillary muscles to the ventricular septum, and resection of fibrotic secondary chordae. This intervention included ascending aorta graft repair (►Fig. 3A; ascending aorta contained intimal ulcers), the great vessels were rerouted via an aortic bypass, and complete arch endovascular repair was performed. Ascending aorta and aortic arch were treated using a hybrid approach. A tubular 28-mm Dacron graft was used to replace the ascending aorta. A bifurcated Dacron prosthesis (24 mm × 12 mm) was sutured “end to side” to the ascending graft and “end to end” with the innominate and left common carotid artery. After aortoendarteriotomy bypass has been performed, we treated the aortic arch using a Gore C-TAG (TGMR404020E) through a right femoral artery approach.

Fig. 1 (A) Presurgery computed tomography (CT) scan showing a thrombosed sacciform aortic arch aneurysm with maximum diameter of 43 mm. (B) Arch vessel vascular graft patency and complete sealing of the aortic lesion at 1-month follow-up CT scan.

Fig. 2 (A) Preoperative transthoracic echocardiography showing anterior and (B) posterior interventricular septal hypertrophy; (C) showing significant left ventricular outflow tract obstruction after Valsalva’s maneuver.
To reduce carotid clamping time and avoid cerebral ische-
mia, we used antegrade selective cerebral perfusion during
innominate trunk anastomosis (Fig. 3B) using the right
axillary artery as the arterial line. Ultimate angiograms
showed a residual saccular aneurysm in the descending
thoracic aorta that we have preoperatively decided not to
treat to reduce the risk of paraplegia.

One month follow-up (Fig. 1B) showed vascular graft
patency and complete sealing of the aortic lesion. Descend-
ing aorta aneurysm was stable in terms of morphology and
diameters and we elected a regular 6-month CT scan
monitoring.

Discussion

Surgical septal myectomy is the gold-standard treatment for
severe OHCM with refractory symptoms, achieving perma-
nent resolution of the LVOT obstruction, reduction in intra-
ventricular pressure, reduction of mitral regurgitation, and
significant improvement of quality of life and long-term
prognosis.5

Recently, OHCM has been associated with aortic abnor-
malities. While aortic stiffness has been proven to be in-
creased in patients with OHCM,3 recent data have suggested
an increased prevalence of aortic dilatation, although there is
no clear consensus.2 To our knowledge, this is the first case
report describing a combined surgical intervention on OHCM
and aortic aneurysm. Ours is a reference center for OHCM,
with 570 cases surgically treated between 2013 and 2019.
However, we have never before faced the coexistence of both
OHCM and aortic pathology requiring contemporary surgical
treatment.

Different factors could play a role in the association
between aortic pathology and HCM: TGF-β (transforming
growth factor-beta) overexpression, neurohormonal distur-
bance, endothelial dysfunction, and an abnormal barorecep-
tor response of the LV.2 For all these reasons, an exclusive
subset of patients with both OHCM and aortic aneurysm
could be at higher risk for adverse events such as aneurysm
rupture and dissection.

Concerning clinical decision making, whereas our patient
fulfilled all the criteria of surgery for OHCM, we were not
guided solely by aneurysm dimension. In this patient, a
hybrid procedure instead of complete surgical arch repair
was deemed to be safer for both neurological and bleeding
complications.

There is no consensus regarding surgical indications for
small saccular aneurysm of the aortic arch. Empirically,
eccentric saccular aneurysms are thought to pose a higher
risk for rupture. Although there is weak evidence, the
current ACC/AHA (American College Cardiology/American
Heart Association) guidelines recommend concomitant re-
placement of a significantly enlarged or weakened ascend-
ing aorta at the time of cardiac surgery.7 Moreover,
saccular aortic lesions are thought more prone to sponta-
neous rupture and standard surgical sizing criteria are
considered insufficient when aortic bulging or penetrating
aortic ulcer are evaluated. Therefore, the dynamic, unpre-
dictable and global clinical picture of our patient led us to
the decision of a complete approach to both the cardiac
and aortic disease.

Funding
None.

Conflict of Interest
The authors declare no conflict of interest related to this
article.

Acknowledgments
None.

References

1 Geske JB, Ommen SR, Gersh BJ. Hypertrophic cardiomyopathy:
clinical update. JACC Heart Fail 2018;6(05):364–375
mopathy with aortic dilation: a novel observation. Eur Heart J
Cardiovasc Imaging 2017;18(12):1398–1403
3 Boonyasirinant T, Rajiah P, Setser RM, et al. Aortic stiffness is
increased in hypertrophic cardiomyopathy with myocardial fi-
brosis: novel insights in vascular function from magnetic reso-
mitral valve repair for obstructive hypertrophic cardiomyopathy
with mild septal hypertrophy. J Am Coll Cardiol 2015;66(15):
1687–1696
5 Maron BJ, Yacoub M, Dearani JA. Controversies in cardiovascular
medicine. Benefits of surgery in obstructive hypertrophic cardio-
mypathy: bring septal myectomy back for European patients.
Eur Heart J 2011;32(09):1055–1058

Fig. 3 (A) Intraoperative specimen reveals an ulcer-like lesion of the
ascending aorta (see arrowhead). (B) Antegrade perfusion during
graft and innominate trunk end-to-end anastomosis.