Abstract: 1348

Alcohol septal ablation in a patient with hypertrophic obstructive cardiomyopathy due to Anderson-Fabry disease

Authors:
N Verheyen¹, H Seggewiss², K Ablasser¹, E Kolesnik¹, D Zach¹, J Binder¹, A Zirlik¹, GG Toth¹, ¹Medical University of Graz, Clinical Department of Cardiology - Graz - Austria, ²Klinikum Wuerzburg Mitte, Standort Juliussspital, Department of Cardiology - Wuerzburg - Germany,

Topic(s):
Hypertrophic Cardiomyopathy

Citation:

Introduction

A 63 year-old male patient with known Anderson-Fabry disease (AFD) presented with progressive deterioration of functional capacity (NYHA III). In the medical history, he had coronary artery disease (CAD) treated with PCI, ICD-implantation due to malignant ventricular arrhythmia, chronic kidney disease stage IV and depression. Besides enzyme replacement therapy he was treated with AT-II antagonist, beta-blocker, calcium antagonist, low-dose aspirin, statin and ezetimibe.

Methods

Echocardiography showed severe global cardiac hypertrophy with pronounced interventricular septum thickening (maximal end-diastolic thickness of 31 mm). Left ventricular ejection fraction was preserved with grade II diastolic dysfunction and evidence of increased filling pressures, supporting the diagnosis of HFrEF. Maximal LVOT gradient was 52 mmHg at rest and 86 mmHg at post-extrasystolic beat. Mitral valve showed incomplete systolic anterior motion (SAM). At the time of presentation NT-proBNP levels were 1.108 pg/ml. After treatment with verapamil at maximally tolerated doses NT-proBNP was lowered to 426 pg/ml and peak LVOT gradient declined to 26 mmHg at rest. However, during bicycle echocardiography the gradient increased to 76 mmHg after maximal workload. Also, the patient did not report relevant improvement in symptoms indicating septal reduction therapy. In face of comorbidities and high perioperative risk, percutaneous transluminal septal myocardial ablation (PTSMA) was preferred.

By PTSMA of the first septal branch provoked peak LVOT gradient dropped from 80 mmHg to 30 mmHg as measured invasively. Peak post-procedural levels of CK and high-sensitive Troponin T were 1.766 U/l and 6.600 pg/ml, respectively. Pre-discharge echocardiography showed a peak LVOT gradient of 16 mmHg without relevant increase during provocation and no evidence of SAM.

Conclusion

Cardiac manifestation of AFB can mimic the phenotype of hypertrophic obstructive cardiomyopathy (HOCM). Therefore, AFB patients with signs or symptoms of heart failure should undergo a comprehensive diagnostic HOCM assessment. In case LVOT obstruction is confirmed, PTSMA in experienced hands can be a safe and effective treatment option for these patients.
Abstract: 1348
Alcohol septal ablation in a patient with hypertrophic obstructive cardiomyopathy due to Anderson-Fabry disease

Introduction
A 63 year-old male patient with known Anderson-Fabry disease (AFD) presented with progressive deterioration of functional capacity (NYHA III). In the medical history, he had coronary artery disease (CAD) treated with PCI, ICD-implantation due to malignant ventricular arrhythmia, chronic kidney disease stage IV and depression. Besides enzyme replacement therapy he was treated with AT-II antagonist, beta-blocker, calcium antagonist, low-dose aspirin, statin and ezetimibe.

Methods
Echocardiography showed severe global cardiac hypertrophy with pronounced interventricular septum thickening (maximal end-diastolic thickness of 31 mm). Left ventricular ejection fraction was preserved with grade II diastolic dysfunction and evidence of increased filling pressures, supporting the diagnosis of HFpEF. Maximal LVOT gradient was 52 mmHg at rest and 86 mmHg at post-extrasystolic beat. Mitral valve showed incomplete systolic anterior motion (SAM). At the time of presentation NT-proBNP levels were 1.108 pg/ml. After treatment with verapamil at maximally tolerated doses NT-proBNP was lowered to 426 pg/ml and peak LVOT gradient declined to 26 mmHg at rest. However, during bicycle echocardiography the gradient increased to 76 mmHg after maximal workload. Also, the patient did not report relevant improvement in symptoms indicating septal reduction therapy. In face of comorbidities and high perioperative risk, percutaneous transluminal septal myocardial ablation (PTSMA) was preferred.

By PTSMA of the first septal branch provoked peak LVOT gradient dropped from 80 mmHg to 30 mmHg as measured invasively. Peak post-procedural levels of CK and high-sensitive Troponin T were 1.766 U/l and 6.600 pg/ml, respectively. Pre-discharge echocardiography showed a peak LVOT gradient of 16 mmHg without relevant increase during provocation and no evidence of SAM.

Conclusion
Cardiac manifestation of AFB can mimic the phenotype of hypertrophic obstructive cardiomyopathy (HOCM). Therefore, AFB patients with signs or symptoms of heart failure should undergo a comprehensive diagnostic HOCM assessment. In case LVOT obstruction is confirmed, PTSMA in experienced hands can be a safe and effective treatment option for these patients.

Figure 1: Peak-to-peak systolic LVOT gradient before (top, 80 mmHg) and immediately after (bottom, 30 mmHg) alcohol injection at post-extrasystolic beat with Valsalva provocation, respectively