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Case Report

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Alcohol septal ablation in hypertrophic obstructive cardiomyopathy: a case with a very high left ventricular outflow tract gradient

Abstract

For more than 20 years, alcohol septal ablation (ASA) has shown to be a safe and effective procedure in the treatment of hypertrophic obstructive cardiomyopathy (HOCM), with results similar to those of surgical myectomy. To our knowledge, we present the first documented case of a HOCM with a very high left ventricular outflow tract (LVOT) gradient (>250 mm Hg) managed with ASA, with a decrease of > 50% of gradient after the procedure.

Keywords: ablation, cardiomyopathy, hypertrophic, left ventricular outflow gradient, catheter, genetic disorder, hypertrophic obstructive cardiomyopathy, beta-myosin heavy chain, syncope

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Abbreviations: HCM, hypertrophic cardiomyopathy; MYH7, beta-myosin heavy chain, MYBPC3, myosin-binding protein C; HOCM, hypertrophic obstructive cardiomyopathy; ASA, alcohol septal ablation; LVOT, left ventricular outflow tract; SAM, systolic anterior motion; OTW, over-the-wire; ESC, European society of cardiology; NYHA I, NYHA functional class

Introduction

Hypertrophic cardiomyopathy (HCM) is a genetic disorder of the cardiac muscle characterized by left ventricular hypertrophy, myofibrillar disarray and myocardial stiffness. This entity affects approximately 1:500 individuals and is the most common cause of sudden death in young athletes.[1] Approximately 40% of the cases of HCM are associated with mutations in genes that encode for beta-myosin heavy chain (MYH7) and myosin-binding protein C (MYBPC3).[2] The majority of hypertrophic obstructive cardiomyopathy (HOCM) patients are detected incidentally; dyspnea is the frequent symptom in 90% of the cases followed by chest pain in a 70-80% and less frequently syncope in 20%, or to debut with sudden death.[2] Currently, septal reduction therapy for patients with HCM can be done either by surgical myectomy or by alcohol septal ablation (ASA).[3] Surgical myectomy is the most commonly performed surgical procedure, in patients with a resting or maximum provoked left ventricular outflow tract (LVOT) gradient of ≥50 mm Hg, who are in NYHA functional Class III-IV, despite maximum tolerated medical therapy.[2] ASA was introduced as a percutaneous alternative to surgical myectomy[3] with the first case reported by Dr. Ulrich Sigwart published in 1995, and subsequently in North America in 1996.[4] Since then, it has shown results similar to that of surgical myectomy. To our concern, we present the first clinical case of a very high LVOT gradient (>250 mm Hg) HCM treated with ASA with significant reduction >50% of gradient after procedure.

Case Series

65-year-old male patient with a history of systemic arterial hypertension; diagnosis of asymmetric HOCM of 3 years of evolution described by echocardiography as LV hypertrophy with a basal distribution, septum width of 18 mm, LVOT maximum gradient of 55 mm Hg at rest, LVEF 75%; managed with ACE inhibitor and beta-blockers. He was seen in consultation due to symptoms of dyspnea, palpitations and progressive angina, NYHA functional class II-III. A new transthoracic echocardiogram was performed with LVOT obstructive gradient of 65 mmHg (Figure 1), 21 mm septum with, LVEF 76%, moderate dilation of left atrium, PASP 32 mm Hg; EKG Sokolow-Lyon index 35 mm. Dobutamine stress echocardiogram was performed with dynamic obstructive gradient of 257 mm Hg, moderate-severe systolic anterior motion (SAM) of mitral valve. Next, cardiac catheterization and ventriculography were performed observing coronary arteries without significant lesions and adequate diameter of septal branches, severely hypertrophied LV with LVEF 80% and end-diastolic pressure of 16 mm Hg. We decided to perform ASA.

Technique: Bilateral femoral arterial accesses were made and a pigtail catheter was placed in LV for pressure assessment during the procedure. Through right femoral artery, the left main coronary artery was cannulated with a JL4 6 Fr guide catheter; a 0.014 guidewire was introduced into the second septal branch and an over-the-wire (OTW) ballon catheter of 2.0 x 9 mm was crossed through the guidewire placing it in the ostium of the septal branch and it was inflated to 14 atm (Figure 1). It was observed by transthoracic echocardiogram,

a decrease of the interventricular gradient and of the contractility in the basal septal region after 10 minutes of balloon occlusion. Once confirmed by echocardiography that the area irrigated by the septal branch corresponded directly to the territory of hypertrophy, a 96% alcohol infusion was slowly initiated at a total of 3 ml. The OTW balloon catheter 2.0 x 9 mm was inflated during the alcohol infusion to protect the anterior descending artery for 40 minutes. After ablation, immediate echography with akinesia in septal basal zone, as well as a decrease of the LVOT gradient of 30 mm Hg mm Hg; EKG with a 3 mm ST segment depression in the anterior face and ventricular ectopy of left branch morphology (Figure 2). Contrast was injected to anterior descending artery with an adequate TIMI III flow, indicator of hemodynamic stability. Follow up at 6 months with improvement in functional class, NYHA I. Dobutamine stress echocardiogram with a dynamic LVOT gradient of 131 mm Hg (Figure 3) right and left cavities of normal diameters and thickness, severe hypokinesia of the basal third of the anterior septum, LVEF 65%, mild mitral valve SAM.

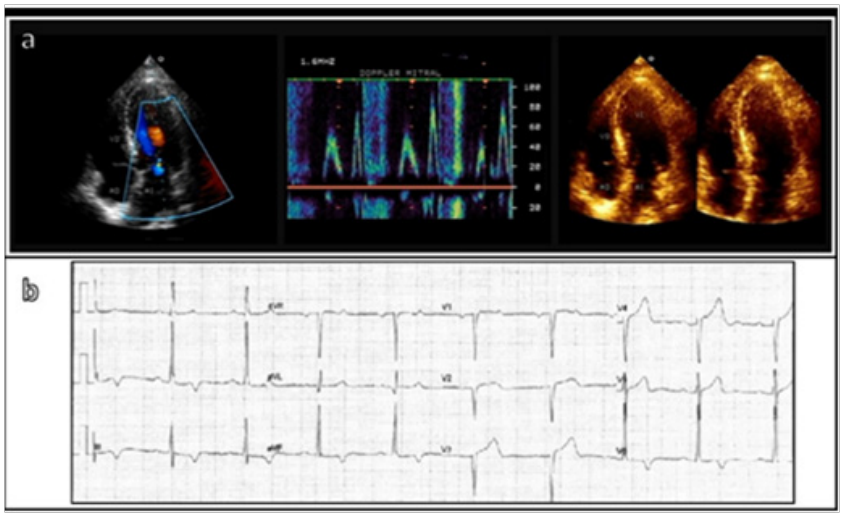

Figure 1 A) TT echocardiogram, B) Electrocardiogram with systolic overload.

Figure 2 A) Pressure gradients and EKG before alcohol ablation, B) Pressure gradients and EKG after alcohol ablation.

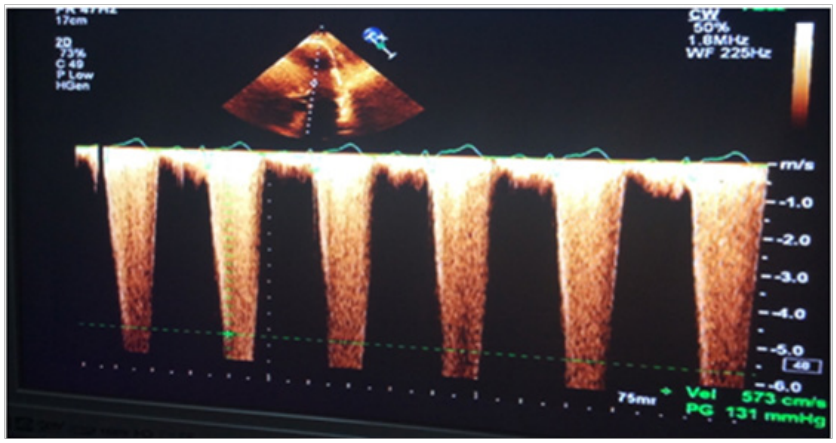

Figure 3 Dynamic LVOT gradient of 131 mm Hg.

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Discussion

ASA has merged as a percutaneous alternative to surgical myectomy. However, literature tends to support better long-term symptom relief in those patients who undergo septal myectomy, lower rate of complications, and immediate results over ASA with an immediate reduction in LVOT gradient. Also, patients with veru high LVOT gradients are considered poor candidates for ASA[5] those patients with a very high LVOT gradient. According to the 2014 European Society of Cardiology (ESC) Guidelines, surgical myectomy is recommended in patients with a resting or maximum provoked LVOT gradient of ≥50 mm Hg, who are in NYHA functional class III-IV, despite maximum tolerated medical therapy (class of recommendation and level of evidence IB).[2] Extremely high LVOT gradients (>200 mm Hg) are uncommon,[6] with several cases reported by Joshi et al.,[7] of >200 mm Hg and a case over 300 mm Hg described echocardiographic records of patients undergoing dynamic changes of LVOT gradients[7]. However, to our knowledge this is the first documented case of a HCM with a very high dynamic LVOT gradient >250 mm Hg and an immediate reduction of resting gradient >50% after ASA, and at 6 month follow-up reduction of dynamic LVOT gradient >100 mm Hg; without complications during and after procedure, no development arrhythmogenic events during follow-up, improvement of NYHA functional class (NYHA I), and the expected hypokinesia in the site of septo-basal hypertrophy.

Conclusion

We present a unique case of HCM with a very high LVOT gradient and a successful management of obstructive gradient with ASA. We consider ASA as a safe and effective procedure in the reduction of very high LVOT in HCM.

Acknowledgements

None.

Conflicts of interest

The author declares there is no conflict of interest.

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