Introduction

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant disease characterized by ventricular hypertrophy whose prevalence in the general population is 1:500\(^1\). Obstruction of the left ventricular outflow tract occurs in 25% of cases and is a marker of poor prognosis. It occurs due to a combination of mechanical and hemodynamic factors, such as interventricular septal hypertrophy, hyperdynamic left ventricular contraction and systolic anterior motion of the anterior cusp of the mitral valve\(^2\). In patients refractory to medical therapy, percutaneous alcohol septal ablation (ASA) may be indicated as a strategy to reduce obstruction. The association of transesophageal echocardiography (TEE) during alcoholization has increased the procedural success to approximately 90%\(^3\).

We report a case of asymmetric septal HCM treated with ASA guided by three-dimensional TEE, followed by literature review and critical discussion of the interaction and involvement of invasive cardiology with several other medical specialists as the ideal approach for this type of procedure.

Case Report

A 31-year-old male patient with hypertension and a history of asymmetric HCM diagnosed 6 years earlier. He had a family history of HCM and sudden death and his father had undergone surgery for correction of septal hypertrophy. On admission, he complained of dyspnea at rest (NYHA class IV), associated with orthopnea and paroxysmal nocturnal dyspnea. He was on atenolol 100 mg/day, verapamil 160 mg/day and furosemide 80 mg/day.

During the diagnostic investigation four years earlier, cardiac magnetic resonance imaging had shown the presence of left ventricular hypertrophy with septal predominance, which determined outflow tract obstruction, in addition to mitral regurgitation and focal myocardial delayed enhancement in the anteroseptal wall, consistent with fibrosis at the ventricular junction. The anterior septal wall thickness was 11 mm and the posterior lateral wall thickness was 14 mm.

On admission, an echocardiogram was performed, which showed a left ventricle with hyperdynamic systolic performance, with no alterations in segmental motion or restrictive pattern, 19-mm ventricular septum, 14-mm LV posterior wall, diastolic and systolic left ventricular diameter of 47 and 26 mm, respectively, and mitral valve with anterior systolic motion of the anterior cusp. The peak intraventricular systolic gradient was estimated at 92 mmHg.

Due to the persistence of limiting symptoms throughout optimized drug treatment, we chose to perform ASA aiming at symptom relief. The procedure was performed using three-dimensional TEE, which showed an intraventricular gradient of 104 mmHg at the beginning of the procedure (Figure 1).

Some authors advocate septal ablation by selective catheterization of the largest septal branch (in this case, the 2\(^{nd}\) septal branch - Figure 2), which is responsible for the irrigation of the mid-basal portion of the interventricular septum, as observed after echographic contrast injection, followed by slow infusion of absolute alcohol\(^2,3\). However, in this case, after both occlusion of the branch and alcohol infusion, there was no decrease in the gradient. In view of the lack of effective results, we searched for another branch with therapeutic potential. Despite the reduced caliber and length, catheterization of the 1\(^{st}\) septal branch with balloon-catheter occlusion resulted in gradient decrease, observed at the three-dimensional transesophageal echocardiography, associated with septal reduction, which was restored to the baseline aspect after balloon deflation. Moreover, the injection of echographic contrast showed a larger irrigation area of the mid-basal portion of the interventricular septum of the 1\(^{st}\) septal branch, when compared to the 2\(^{nd}\). Based on these findings, we performed alcoholization of the 1\(^{st}\) septal branch with 2 mL, with reduction of the intraventricular gradient, as observed by invasive manometry, to 21 mmHg and by three-dimensional TEE to 26 mmHg, with no complications.

After three days of intervention, a new echocardiogram was performed and disclosed a ventricular septum of 16 mm, LV outflow tract gradient of 46 mmHg and improvement of the diastolic alteration (pseudonormal

Keywords

Cardiomyopathy, Hypertrophic; Hypertrophy, Left Ventricular; Echocardiography, Transesophageal; Ablation Techniques.
Case Report

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Septal alcoholization with multidisciplinary team

Figure 1 - Two-dimensional transesophageal echocardiography before percutaneous alcoholic ablation, showing increased thickness of the interventricular septum (A). Peak gradient in the left ventricular outflow tract (LVOT), measuring 104 mmHg before the procedure (B). Three-dimensional transesophageal echocardiography after alcoholization, showing a hyperechoic spot, appropriately treated with ethyl alcohol injection (white arrow) (C). LVOT gradient measuring 26 mmHg after the procedure (D)

Discussion

The main finding of this case report is that the best ASA technique involves the interaction of multiple professionals from different fields of Cardiology and Medicine dedicated to this type of intervention.

The initial alcoholization of the large septal branch did not result in the expected hemodynamic improvement, as readily identified by TEE. Soon after, the morphological identification of another therapeutic target by angiography (1st septal branch) was confirmed as functionally important by the TEE, allowing a successful procedure.

Typically, the first accessible major septal branch is identified by angiography and selected for ablation. However, echocardiographic images during the procedure may suggest the alcoholization of another branch, according to the location of the septal fibrosis caused during the procedure. Three-dimensional TEE stands out in this context to help in correct identification of the artery responsible for septal hypertrophy, thus avoiding alcoholization of branches that irrigate the papillary muscle or the RV free wall.

General anesthesia is often used, allowing greater safety during the procedure. This is especially advisable when there is risk of cardiac instability during the procedure. The anesthesiologist must be aware of the peculiarities of invasive cardiological procedures, and particularly of ASA, since the manipulation of the catheter, balloon catheters, guide wires and alcohol can cause acute events.

Complete atrioventricular block is a complication that resolves spontaneously in most cases within 13 days, with less than 10% requiring a permanent pacemaker, mainly if the intervention is guided by echocardiography. Transmural myocardial infarction, with the formation of potentially arrhythmogenic areas, and ventricular septal defect are other possible complications. Basically, they depend on the size of necrosis caused by the injected amount of alcohol. The use of echographic images during the procedure was effective in locating the best site to inject alcohol, as well as in reducing such complications.

pattern). The patient was hemodynamically stable and asymptomatic, and was discharged four days later, using atenolol 50 mg a day.

At the 30-day follow-up visit, the patient complained of an episode of chest pain at rest followed by syncope, and the atenolol dose was increased to 100 mg daily. At the four-month follow-up visit he was asymptomatic.

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The syncope episode after the procedure may have occurred secondary to arrhythmia, as the patient had a family history of sudden death and focal myocardial delayed enhancement on magnetic resonance imaging and he might be eligible to an implantable cardioverter-defibrillator.

It is yet to be defined in the literature whether percutaneous septal alcoholization of asymmetric septal HCM can alter patient prognosis. However, their quality of life, represented by symptom relief and increased exercise capacity, certainly improves\textsuperscript{1,2}. This improvement was observed in the present case, since the patient is asymptomatic and used only one type of medication four months after the procedure.

**Potential Conflict of Interest**

No potential conflict of interest relevant to this article was reported.
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Study Association

This study is not associated with any post-graduation program.

References


