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Clinical Case

Anesthetic management during endocardial radiofrequency ablation of septal hypertrophy – a case report

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Background: Hypertrophic cardiomyopathy (HCM) is a genetic disorder present in up to 1/500 individuals, about 20–30% of them presenting with hypertrophic obstructive cardiomyopathy (HOCM) due to left ventricle outflow tract obstruction. This is an important cause of sudden cardiac death. Endocardial radiofrequency ablation of septal hypertrophy (ERASH) might be an attractive treatment for HOCM, particularly in patients who do not respond to transcoronary alcohol septal ablation (TASA). **Aim:** To describe technical aspects related to the procedure and anesthetic management of an ERASH case. **Case report:** A 64-year-old woman with HOCM was scheduled for ERASH. She had worsening of dyspnea on exertion and generalized fatigue for the previous weeks after previous surgical myomectomy about 6 months ago. The anatomy was unfavorable for TASA and the patient was not willing to undergo another surgery. Preoperative transthoracic echocardiography (TTE) showed asymmetric mid-septal hypertrophy, systolic anterior motion with septal contact and left ventricular outflow tract maximum gradient of 68 mmHg at rest and 105 mmHg after the Valsalva maneuver. General anesthesia was performed. Pulse pressure variation, echocardiography parameters and passive leg raising test where used to guide fluid therapy. At the end of the procedure, analgesia was provided together with prophylaxis of nausea and vomiting. Extubation was uneventful and the patient was transported to the intensive care unit eupneic and hemodynamically stable. On the fourth postoperative day, TTE showed septal hypocontractility and maximum gradient reduction of 33% at rest (68 mmHg to 45 mmHg) and 31% after the Valsalva maneuver (105 mmHg to 73 mmHg). The patient was discharged from hospital at the sixth postoperative day. One month later, she reported progressive improvement of symptoms and expressed satisfaction with the results. **Conclusion:** Better understanding of the pathophysiology and natural history of HCM has enabled earlier diagnosis, as well as a more adequate therapeutic approach. Anesthesiologists should be aware of the pathophysiology of HOCM and must be prepared to anticipate the hemodynamic changes and cardiovascular instability that such patients may show perioperatively. ERASH is a promising therapeutic modality increasingly used for HOCM and anesthesiologists should become more familiar with it.

Key words: anesthesia, hypertrophic obstructive cardiomyopathy, radiofrequency catheter ablation

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Hypertrophic cardiomyopathy (HCM) is a genetic disorder characterized by unexplained hypertrophy of the left ventricle (LV). Its prevalence is up to 1/500 individuals, about 20–30% of them presenting with hypertrophic obstructive cardiomyopathy (HOCM) due to left ventricle outflow tract (LVOT) obstruction [1, 2]. Although it is an important cause of sudden cardiac death among young people, a reduction in mortality has been observed in the last decades [3]. This advance has been due to better understanding of the disease's pathophysiology, its earlier diagnosis and the advances in therapeutic strategies.

It is well known that surgical or percutaneous septal reduction therapies (SRT) are the only effective methods for the reduction of the LVOT gradient. Among SRTs, endocardial radiofrequency ablation of septal hypertrophy (ERASH) might be an attractive treatment for sympathetic HOCM, particularly in patients who do not respond to transcoronary alcohol septal ablation (TASA) [4].

Here, we describe technical aspects related to the procedure and anesthetic management of an ERASH case.

Case report

A 64-year-old woman with HOCM was scheduled for ERASH. She had worsening of dyspnea on exertion and generalized fatigue for the previous weeks, after previous surgical myectomy about 6 months ago. The anatomy was not favorable for TASA and the patient was not willing to undergo another surgery. Other comorbidities included hypertension, paroxysmal atrial fibrillation and anxiety. Her medications included furosemide, spironolactone, atenolol, warfarin, zolpidem, and omeprazole. Preoperative transthoracic echocardiography (TTE) showed asymmetric mid septal hypertrophy, systolic anterior motion (SAM) with septal contact and LVOT maximum gradient of 68 mmHg at rest and 105 mmHg after the Valsalva maneuver. Her previous ergometric test showed a 5 METs functional capacity.

After detailed discussion with the interventional cardiologist, it was decided to proceed with general anesthesia. Standard monitoring of the patient included five-lead electrocardiography, pulse oximetry, intermittent noninvasive blood pressure measurements, capnography and gas analysis. Body temperature was measured by an esophageal thermometer. A judicious dose of midazolam was administered to reduce anxiety. Radial artery catheterization was performed before induction anesthesia and orotracheal intubation. Etomidate (0.2 mg/kg), lidocaine (1 mg/kg), fentanyl (3 mcg/kg), and cisatracurium (0.15 mg/kg) were chosen for the induction. Anesthesia was maintained with remifentanil (0.15 mcg/kg/min) and sevoflurane (1% expired fraction). Pulse pressure variation, echocardiography parameters and passive leg raising test where used to guide fluid therapy. Boluses of phenylephrine (50 mcg) were administered when necessary to increase afterload and avoid hypotension.

Two femoral artery punctures were performed by the interventional cardiologist, one for retrograde aortic approach to the LV and the other for arterial LV pressure measurement. Three femoral vein punctures were performed for catheter placement, right ventricle catheter placement and transseptal puncture, using a SL01 Abbott Long Sheath. Left septal approach was tried using transseptal puncture with an Agilis™ XT deflectable sheath. After that, the retrograde

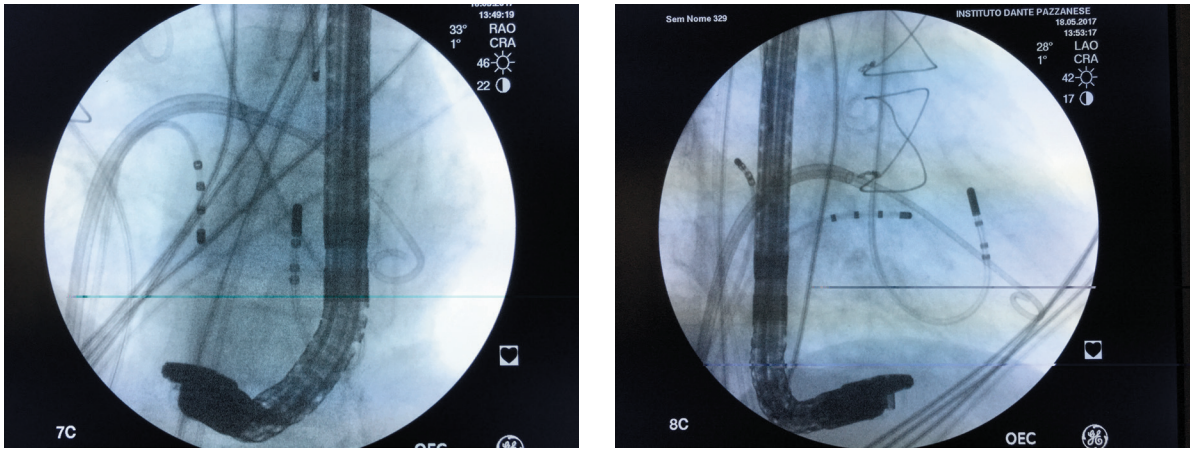

Fig. 1. Fluoroscopy obtained during the left septal ablation procedure. Gradient measurements were obtained using a pig tail catheter inserted through transseptal puncture and a SL01 Abbott Long Sheath. A quadripolar diagnostic catheter was used for recording. Left septal ablation was performed via retrograde aortic approach. Trans-esophageal echocardiography and when necessary transtoracic echocardiography assessments were used to guide the electrophysiologyst during the entire procedure

approach was guided by trans-esophageal echocardiography (TEE) and occasionally TTE to achieve the best catheter position (Fig. 1). Radiofrequency was delivered as per the institution protocol until the LVOT gradient was reduced by 40% from the initial measured values (as measured by intraoperative TEE and a pig tail catheter placed in LV apex and LVOT), as previously described [5].

At the end of the procedure, analgesia was provided together with prophylaxis of nausea and vomiting. Extubation was uneventful done and the patient was transferred to the intensive care unit, being eupneic and hemodynamically stable.

During the first postoperative day low doses of noradrenaline were needed to maintain adequate afterload, despite patient's good recovery. Serum troponin levels remained elevated during the first 48 hours and then decreased progressively (Fig. 2). The patient stayed in the intensive care unit for 3 days before being transferred to the ward. On the fourth postoperative day, TTE showed septal hypocontractility and a maximum gradient reduction of 33% at rest (68 mmHg to 45 mmHg) and 31% after the Valsalva maneuver (105 mmHg to 73 mmHg). The patient was discharged from the hospital at the sixth postoperative day. One month later she reported progressive improvement of symptoms and expressed satisfaction with the results.

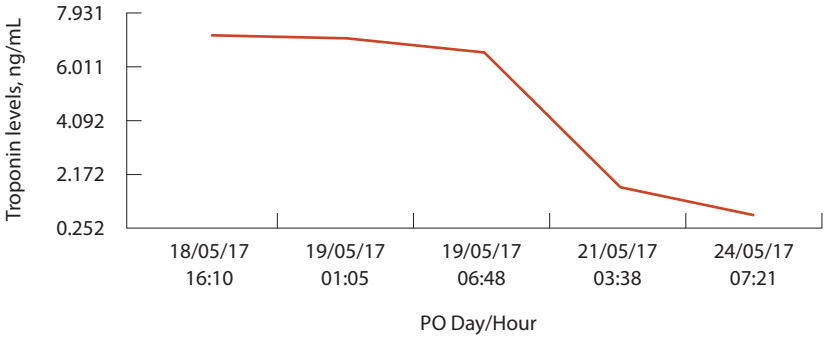

Fig. 2. Troponin levels during perioperative period; *PO* postoperative. Upper level: below 0.034 ng/mL (normal); Cut-off point: 0.12 ng/ml

Discussion

As described, better understanding of the pathophysiology and natural history of HCM has enabled earlier diagnosis, as well as a more adequate therapeutic approach. Studies have shown that the main causes of death in patients with HCM are sudden cardiac death secondary to ventricular arrhythmias, heart failure and stroke [6].

To improve functional capacity, reduce symptoms and prevent disease progression drug therapy, based on the use of beta-blockers and/or verapamil, disopyramide, antiarrhythmics and diuretics should be started [5]. However, patients with HOCM who remain severely symptomatic despite maximal drug therapy are candidates for invasive procedures, including permanent pacemaker insertion, percutaneous SRT, or surgical myectomy [1].

Among invasive procedures, the clinical eligibility criteria for surgical myectomy and TASA are the same, according to the guidelines by American College of Cardiology/American Heart Association (ACC/AHA) and European Society of Cardiology (ESC). The eligibility criteria can be summarized as follows:

a. Clinical: severe dyspnea or chest pain (usually New York Heart Association (NYHA) functional classes III or IV) or occasionally other exertional symptoms (such as syncope or near syncope) that interfere with everyday activity or quality of life despite optimal medical therapy.

b. Hemodynamic: dynamic LVOT gradient at rest or with physiologic provocation ≥ 50 mmHg associated with septal hypertrophy and SAM of the mitral valve.

c. Anatomic: targeted anterior septal thickness sufficient to perform the procedure safely and effectively in the judgment of the individual operator [7, 8].

None of these guidelines, however, cites ERASH as an invasive treatment alternative. First described in 2004 by T. Lawrenz, ERASH has become an increasingly used treatment alternative [9]. ERASH has its best indications in patients with the criteria described above who are not eligible or refuse to perform surgical myectomy, as well as those who do not respond to TASA, since a significant limitation of the latter technique is its dependence on the coronary anatomy to provide access for ablation, and up to 15% of patients have no septal vessel suitable for TASA procedure [10, 11].

To better understand how ERASH works, we must comprehend the pathophysiology of HOCM. For LVOT obstruction in HOCM, basal septum hypertrophy and SAM of the mitral valve are key elements. In the majority of cases, asymmetric septal hypertrophy leads to LVOT obstruction causing rapid acceleration of blood flow apical from the mitral valve (the Venturi effect). It is believed that this narrowing of the LVOT favors the movement of the valve apparatus towards the septum. However, this is not the sole pathophysiological factor of LVOT obstruction. In some patients, obstruction may occur despite low velocities in LVOT. Abnormal posteriorly directed flow due to the septal hypertrophy may circulate around the mitral valve and back towards the LVOT. If there is a structural anomaly on the anterior mitral leaflet like a redundancy, the leaflet may be caught by this flow. Then, the anterior leaflet may move towards the hypertrophic septum. Once the anterior mitral leaflet contacts the

interventricular septum, the LVOT orifice is narrowed further and greater obstruction to flow develops, resulting in a higher pressure difference. This may cause positive amplifying feedback loop throughout ventricular systole. Main deleterious effects of LVOT obstruction are reduction of cardiac output, mitral regurgitation due to SAM, diastolic dysfunction and coronary flow abnormalities, causing the symptoms described above [12, 13].

ERASH reduces LVOT gradient by inducing, via endocardial radiofrequency, a local alteration of septal contractility, thereby interrupting contact of the mitral valve anterior leaflet and the basal septum. As it is true for other septal ablation modalities, the most important part of ERASH procedure is the exact localization of this contact point. For this purpose, transesophageal, transthoracic or intracardiac echocardiography, as well as fluoroscopy guidance, are used. In addition, the use of CARTO® (Biosense Webster, Diamond Bar, CA, USA) and LocaLisa (Medtronic EP Systems, Minneapolis, MN, USA) as electroanatomical mapping systems has been described [4, 14]. The approaches used for catheter insertion can vary from the right or left ventricle to aortic retrograde approach.

In a study in 19 patients, T. Lawrenz et al. observed that ERASH was effective in reducing LVOT gradient and resulted in significant improvement in exercise capacity and NYHA functional class after 6 months [14]. In another study by N. Sreeram et al., including 32 children who underwent ERASH procedure, an immediate decrease was detected in the catheter pullback gradient and further reduction in the echocardiographic gradient was seen at follow-up [15]. More recently, R.M. Cooper et al. described 5 patients undergoing ERASH and observed a reduction in resting and Valsalva exercise-induced LVOT gradients, as well as an improvement of their NYHA status [16].

During the preoperative anesthetic evaluation, in addition to assessing the clinical status of the patient, it is important to ensure the correct use of the prescribed medications. Ideally, they should be continued perioperatively, including beta-blockers, in order to keep a target heart rate of 60–65 beats/min. Moreover, prolonged fasting should be avoided, since hypovolemia may exacerbate LVOT obstruction. Maintenance of preload and afterload, heart rate, sinus rhythm, mild suppression of myocardial contractility, and myocardial perfusion pressure are the goals of anesthetic management [13, 17, 18].

As it was the first case of ERASH performed at our institution, in addition to the standard monitoring recommended for procedures done under general anesthesia, we have chosen to ensure continuous invasive blood pressure monitoring from the induction of anesthesia on. This tool was useful to avoid hypotension, which is poorly tolerated in these individuals, and to evaluate fluid responsiveness through pulse pressure variation. A bolus of 100 mL of crystalloids was given when delta pulse pressure exceeded 10% [18–20].

In order to allow adequate handling of TEE, ensure better hemodynamic control, as well as immobility throughout the procedure, general anesthesia with orotracheal intubation was the technique of choice. During the induction, in addition to fluid management and titration hypnotics and opioids, we used phenylephrine to avoid hypotension, since the use of α1-adrenergic agonists increases systemic vascular resistance and decreases LVOT obstruction, whereas β-adrenergic agents worsen it due to their positive inotropic and chronotropic effects [21, 22]. As a neuromuscular blocker, cisatracurium was chosen because of its hemodynamic stability and lack of histamine release. We believe that as long as TTE is performed and as interventional cardiology gains expertise, a less invasive anesthetic technique may be a plausible alternative. One should bear in mind the importance of maintaining adequate pain control during septal ablation.

It is important not to inactivate an implanted DDD-pacemaker during anesthesia when it has been placed for timed atrial contraction and gradient reduction. Tachycardia associated with direct laryngoscopy and intubation can be prevented by pretreatment with intravenous metoprolol or esmolol. This practice is recommended, especially in patients with history of heart failure symptoms, angina, or HCM-related syncope or high resting gradients.

Mechanical ventilation parameters were adjusted to ensure an $EtCO_2$ of 35–40 mmHg. In addition, to reduce intrathoracic pressures and thus favor venous return, we maintained a low tidal volume and reduced the positive end expiratory pressure and inspiratory: expiratory ratio.

Special attention should be given to potential risks of this technique. Complete heart block was reported in 21% of cases, with permanent pacemaker dependency [14]. These patients can also develop other types of arrhythmia such as bradyarrhythmia, supraventricular tachycardia, ventricular tachycardia or fibrillation for which appropriate pharmacologic agents and/or defibrillation should be kept ready [17]. Other complications include cardiac tamponade, superficial burning from electrocautery plate, risks related to any percutaneous procedure (retroperitoneal hemorrhage, limb ischemia and groin hematoma, for example) and even death. Death report was described in a patient that developed acute left ventricular

dysfunction secondary to paradoxical increase in the degree of LVOT obstruction due to tissue edema at the ablation sites [12, 14, 15]. Anesthesiologists should be aware of the pathophysiology of HCM when handling PCR in these individuals, since inotropic agents such as adrenaline may worsen LVOT obstruction and should be used judiciously [18].

To our knowledge, this is the first paper describing the anesthetic management of the endocardial radiofrequency ablation of septal hypertrophy for HCM. Although ERASH seems a technically feasible strategy, short- and longterm results and complications were not clearly defined, since most trials currently available have a small number of cases and are single centered. Moreover, at this time we cannot make a statistical comparison between TASA and ERASH due to the small sample size with ERASH. Further largescale studies are needed to clarify this issue [4, 12, 15]. However, as this is a promising therapeutic modality increasingly used for HCM with LVOT obstruction, we believe that anesthesiologists should become more familiar with it.

Conflict of interests

The authors declare that they have no conflict of interest.

Author contributions

All authors have contributed equally to the paper. All authors have read and approved the final manuscript.

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