Anesthesia for laser lead extraction in a patient with catecholaminergic polymorphic ventricular tachycardia: a case report

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Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an uncommon hereditary arrhythmogenic syndrome, with an estimated prevalence of ~0.1/1000. In this disease, myocardial calcium receptors are, more than usual, sensitive to catecholamine release, leading to severe tachyarrhythmias after emotional or physical stress. To the best of our knowledge, only a few cases of general anesthesia (GA) in patients with CPVT have been reported in literature, and none for potentially arrhythmogenic procedures like laser lead extraction. We describe the case of a 17-year-old male patient with CPVT, undergoing GA, for laser lead extraction of an old implantable cardioverter-defibrillator. The procedure, conducted in totally intravenous anesthesia, was uneventful, and just a few sporadic ventricular extrabeats owing to mechanical manipulation of the old wire were reported. The patient was extubated in the operation theater and discharged the following day. In the discussion, the case is analyzed, the main issues highlighted, and different anesthetic strategies considered. In conclusion, laser lead extraction in patients affected by CPVT can be performed safely under GA, provided that patients are cautiously treated and assessed during the whole perioperative period.

Keywords:
catecholaminergic polymorphic ventricular tachycardia, general anaesthesia, laser lead extraction

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Introduction

Catecholaminergic polymorphic ventricular tachycardia (CPVT) is an uncommon hereditary arrhythmogenic syndrome. Although data from systematic assessment are not available, the prevalence of this disease has been estimated at ~0.1/1000 [1]. Two types of CPVT have been identified. The more common (also known as CPVT1) is autosomal dominant and is due to an abnormal pool of ryanodine-2 calcium receptors (RyR2) [2]. Calsequestrin receptors (CASQ2) can also been involved in the less common autosomal recessive variant (CPVT2) [3]. Both receptors are more sensitive than usual to intrinsic or extrinsic catecholamines [4,5]. Moreover, other even more uncommon genetic variations have been reported [6].

Clinically, patients affected by CPVT present syncopal episodes, starting usually between the first and the second decade of life, owing to severe tachyarrhythmias on occasion of physical efforts or acute emotional stress, although sudden death or aborted sudden death can be the first manifestations of the disease [4]. During an effort or after isoproterenol infusion, the ECG can show isolated ventricular premature beats, runs of ventricular tachycardia, and polymorphic bidirectional bursts. When syncope is accompanied by seizure-like activity, the diagnosis can be missed and the patient treated with anti-epileptic medications.

CPVT is part of the very heterogeneous group of inherited primary arrhythmogenic disorders. Despite similar to arrhythmogenic right ventricular dysplasia, CPVT is characterized by an anatomically normal heart [7].

Therapeutic resources include high dose of nonselective beta-blockers, implantable cardioverter-defibrillator (ICD), flecainide, and left cervical sympathetic denervation [1,8].

Extraction of fibrotic tissue-encapsulated implantable electronic device leads can be a cause for lead failure (i.e. owing to rupture or high output threshold) and/or infections. The procedure can be done on open surgery or, more frequently, percutaneously. Open heart

surgery is usually considered when the patient has failed prior percutaneous extraction procedure or when cardiac imaging identifies large lead masses (vegetation or thrombus >2.5 cm) [9]. Percutaneous extractions can be successfully completed using different approaches: simple manual traction, locking stylets, telescoping sheaths, femoral snares, mechanical cutters, and laser sheaths.

Laser-assisted lead extraction is now the standard procedure adopted in many institutions owing to its efficacy and low incidence of major adverse events. The first laser lead extraction was performed by Byrd in 1994 [10]. The initial model of laser sheath (SLSI) was modified, and a second-generation more flexible laser sheath (SLSII) was released in 2002 [11].

The laser sheath fiber-optical device used in our institution (CVX 300; Spectranetics, Colorado Springs, Colorado, USA) delivers the laser energy to the distal end of the sheath. The sheath is constructed by 82 optical fibers, each with a core diameter of 100 μm, around an inner lumen. The device emits an excimer laser beam using xenon chloride, in the ultraviolet region, not visible for humans. This cool cutting laser has an absorption depth of 0.05 mm, the energy being absorbed by proteins and lipids. These parameters allow cutting of the tissue with minimal risk of damaging the veins or insulation of lead. In case of fibrotic calcified encapsulation, the laser is less effective in releasing the leads, and different techniques must be considered.

In literature, only a few cases have been reported regarding anesthetic management in patients affected by CPVT [12–14].

Case report

A 17-year-old male patient with CPVT due to CASQ2 receptor hypersensitivity, on regular nadolol 80 mg postoperatively OD, flecainide 50 mg postoperatively BD, with a transvenous ICD positioned at the age of 6 years, and left cervical sympathetic denervation at the age of 15 years, was scheduled for laser extraction of an old lead, reinsertion of new intracavitary leads, and implantation of a new generator for battery depletion.

The patient's electrophysiological history reported six shocks from his ICD after a football game, three years ago, when he held his nadolol and flecainide for 4 days.

No any other relevant past medical history was reported. His weight was 76 kg and height 180 cm, with a BMI of 23.5 kg/m^2.

On the surgical procedure day, the patient received 20 mg of Temazepam 1 h before the procedure. In the anesthetic room, before the induction of general anesthesia (GA), a second dose of benzodiazepine (midazolam 4 mg intravenous) has been administered, once the intravenous cannula was inserted. The GA was conducted with remifentanil infusion (0.5 μg/kg/min) up to a total dose of 1 μg/kg then reduced to 0.3 μg/kg/min, propofol TCI (target plasmatic concentration between 2.1 and 3.6 μg/ml), and atracurium besilatus 50 mg followed by 10 mg every 30 min. Phenylephrine was available as a vasoconstrictor.

External defibrillator patches have been positioned before the induction of the GA.

The patient was intubated and ventilated without any complication. After oral endotracheal intubation, a transesophageal echocardiography probe was inserted with no complications.

Before the induction of the GA, but after midazolam intravenous, a cannula was inserted into the patient's right radial artery to monitor the blood pressure. A 8.5 Fr 16-cm central venous catheter into the right internal jugular vein was positioned when the patient was under GA. SaO_2, $ETCO_2$, an esophageal temperature probe, two-lead ECG, and a BIS 2-Channel Monitor (Aspect Medical Systems Inc., Norwood, Massachusetts, USA) were positioned as a routine monitoring procedure.

The procedure lasted 65 min and was uneventful. Just a few sporadic ventricular extrabeats owing to mechanical manipulation of the old wire were observed. At the end of the procedure, the patient received 5 mg of morphine intravenous when the remifentanil infusion was interrupted, paracetamol 1 g, ondansetron 4 mg and cyclizine 50 mg intravenous, before being extubated in the theater. After 2 h spent in recovery room, the patient was transferred to the ward and discharged the following day.

Discussion

In case of surgical procedures, patients affected by CVPT are at risk of life-threatening rhythm disturbance during the whole perioperative period. Before GA induction, the simple stimulus of venipuncture can lead to arrhythmias. Furthermore,

in patients with an ICD, a single shock can increase the sympathetic tone leading to further shocks and, eventually, to a malignant cycle of them [15]. In the case reported, after the usual oral premedication, another dose through the peripheral venous line has been given to keep the patient deeply sedated and to tolerate arterial cannulation.

Beside a proper premedication, it is fundamental for patients to have their regular antiarrhythmic treatment (usually beta-blockers and, sometime like in this case, flecainide). On the patient's past medical history, there was an episode of ICD shock following a 4-day medication cessation, but ICD firings after just 1 day of drugs suspension have been reported, if associated with emotional or physical triggers [16].

Laser lead extraction is a common procedure to remove pacing, ICD, or cardiac resynchronization therapy intracavitary wires. Despite usually safe, vascular laceration, perforation, hemothorax, tamponade, and arrhythmias have been reported as complications [17]. The mechanical stimulation of the right ventricle, and the sympathetic response owing to the laser action itself can potentially, on CPVT patients, lead to malignant arrhythmias. For those reasons, monitoring the anesthetic depth with BIS to avoid sympathetic response to surgical stimulus is, in our opinion, mandatory. Furthermore, it allows minimizing hypotension episodes and, consequently, vasoactive drugs requirement.

In literature, total intravenous anesthesia and combine spinal-epidural anesthesia have been reported in patients with CPVT, although volatile anesthetic agents (but not halothane) are considered safe. Particularly, because of CPVT pathogenetic mechanism, sevoflurane, and other drugs commonly used in anesthesia and known to cause QT prolongation (i.e. ondansetron) can be safely administered in this case [7]. Furthermore, because ryanodine receptors involved in malignant hyperthermia are of a different subtype (RyR1) compared with those related to CPVT (RyR2), there is no risk of triggering of malignant hyperthermia by halogenated anesthetics.

Although keeping the patient in spontaneous ventilation with a laryngeal mask, to avoid direct laryngoscopy sympathetic response, has been suggested [12]. In case of transesophageal echocardiography probe insertion, an endotracheal tube has to be positioned. In this case, common muscle relaxants (but not pancuronium, because of its sympathetic enhancement) can be safely administered.

Both pain and postoperative nausea and vomiting can result in discomfort and sympathetic activation. Opioids (remifentanil infusion and morphine intravenous) and paracetamol have been given to achieve a complete pain relief, but also NSAIDs can be considered. As antiemetic drugs, ondansetron and cyclizine, an antihistamine medication whose sedative effect can be particularly useful in this case, have been administered.

In patients with CPVT, vasoactive drugs have to be given carefully; however, catecholamine infusions (especially isoproterenol) have to be obviously avoided. Trying to keep the patient hemodynamically stable is an important issue. Proper anesthetic depth assessment, correct fluid infusion, and spontaneous ventilation can help to avoid hypotension and vasoactive drugs requirement. Anyway, in case of anesthesia-related vasodilation, direct alpha 1 agonists like phenylephrine are considered safer than direct/indirect agonistic medications, like metaraminol or ephedrine. On the contrary, in occasion of hypertension and sinus tachycardia, esmolol infusion and/or central alpha 2 agonists, like clonidine or dexmedetomidine, which have also sedative and analgesic properties, can be considered.

In conclusion, despite being challenging, laser lead extraction in patients affected by CPVT can be performed safely under GA, provided that patients are cautiously treated and assessed during the whole perioperative period.

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Conflicts of interest
There are no conflicts of interest.

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