Anesthesia Management in a Patient with Unclassified Cardiomyopathy for Transureteral Lithotripsy Surgery

Abstract
Anesthesia management has always been challenging in cardiac patients, especially patients with cardiomyopathy. There are a variety of cardiomyopathies such as unclassified cardiomyopathy as a complex type that can occur in many forms like left ventricular noncompaction (LVNC) that is an uncommon primary genetic cardiomyopathy typified by noticeable trabeculation of the left ventricular (LV) wall and intertrabecular recesses. We report anesthesia management in a 53-year-old female patient who admitted to the hospital for the transureteral lithotripsy surgery due to dysuria and urolithiasis with a medical history, and echocardiographic examination indicated the diagnosis of hypertension and unclassified cardiomyopathy (LVNC).

Keywords: Anesthesia, noncompaction cardiomyopathy, spinal anesthesia, unclassified cardiomyopathy

Introduction
Cardiomyopathies are a heterogeneous group of myocardial diseases related to electrical and mechanical dysfunction and typically (but not always) caused by inappropriate ventricular hypertrophy or dilatation because of various reasons that are often genetic. Cardiomyopathies often result in death or progressive disabilities related to the heart failure. Despite the fact that the cardiomyopathy is not treatable, its symptoms can be well managed and patients can have a good life expectancy. Cardiomyopathies are divided into primary and secondary groups; the primary type is often limited to the heart muscles and has genetic, acquired, or both causes. The secondary group includes the pathophysiology of heart involvement in the field of disorders of all organs.

Based on the European Society of Cardiology (ESC) Classification of Cardiomyopathies, the main types of cardiomyopathy contain hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, restrictive cardiomyopathy, and unclassified cardiomyopathy. The treatment, which may include medications, surgical implants, or heart transplant in severe cases, depends on what type of cardiomyopathy you have and how serious it is. Clinical symptoms include dyspnea, orthopnea, palpitations, angina, gallop rhythm, syncope, functional decline, left ventricle ejection fraction <45%, cardiomegaly, and plural effusion. The clinical prognosis varies from minimum symptoms to acute myocardial infarction or sudden death. Echocardiography and cardiac magnetic resonance imaging (cMRI) can be used to diagnose cardiomyopathy.

The anesthesia management is usually accompanied by many challenges in this group of patients. The primary goals of anesthesia management of patients with a history of cardiomyopathy are adequate tissue oxygenation and perfusion, as well as maintaining adequate cardiac output.

Based on the ESC Classification of Cardiomyopathies, left ventricular noncompaction (LVNC) cardiomyopathy is ordered in unclassified cardiomyopathy. LVNC type is commonly seen as abnormal trabeculations in left ventricles, which often affects the apex region as well as hypertrophy, dilatation, and systolic or diastolic dysfunction or both. The molecular mechanism in noncompaction type is not fully known now, but it is hereditary in 30%-50% of patients.

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occur as systolic disorders with heart failure, thrombolytic complications, arrhythmia, and sudden cardiac arrest.\cite{10,11}

In the present report, the anesthesia management was investigated in a transureteral lithotripsy (TUL) candidate due to dysuria and urolithiasis. It was also diagnosed the existence of unclassified cardiomyopathy with the LVNC type.

**Case Report**

This case was a 53-year-old female patient who admitted in Seyed-Al-Shohadaee Hospital, Sanandaj, Iran, for the TUL surgery in May 2019 with a history of hypertension, ventricular arrhythmias, and unclassified cardiomyopathy. Based on the echocardiographic and CMRI findings in the studied patient, California criteria\cite{12} were observed. In the echocardiography, mild hypertrabeculation in LV apex was observed, and mild increased LV size without LV hypertrophy and reduced systolic function was reported. LV ejection fraction (LVEF) = 30\%, right ventricular ejection fraction = 52\%, pulmonary artery pressure = 30 mmHg, and partial regurgitation of bicuspid and tricuspid valves. Figures 1 and 2 show right ventricular in systolic and diastolic phase. There was also an increase in the left ventricular (LV) size as well as a decrease in LV systolic function [Figures 3 and 4]. She had a history of consuming Metohexal (23.75 BD), ASA (80 mg daily), atorvastatin (40 mg daily), and lisinopril (2.5 mg daily).

The patient visited for chest pain and shortness of breath 6 months ago; the echocardiography indicated the LV. Table 1 presents the patient laboratory results.

The patient was visited the night before the surgery. The bilateral lung sound was clear in physical examination. Heart sounds were reduced s1 with no soufflé. In the airway examination of the patient, there was no finding of the airway problem and has Mallampati Class II. In the patient’s electrocardiogram, premature atrial contraction and ST-segment depression and T-wave inversion were visible in precordial leads, especially V1 and V4 to V6. According to the cardiologist, the risk of surgery was moderate.

The patient was fasting for 8 h. The defibrillator and invasive and noninvasive blood pressure monitoring were prepared before the induction of anesthesia. Antiarrhythmic and vasoactive drugs were prepared. Eighteen-gauge intravenous line was established from the left cubital for the patient and she received 500 ml of Ringer’s solution before the anesthesia. One h before start of surgery, 2 g of ampicillin in 100 ml of normal saline was administered to the patient for prophylaxis. According to the patient’s clinical condition and the advice and recommendation of the cardiologist, as well as the decision of the anesthesiologist based on the existing conditions, the patient was prepared for spinal anesthesia. After obtaining the patient’s consent to perform spinal anesthesia, the patient sat in sitting position and the anesthesia was performed by sterile 26G Quincke needle in the intervertebral space L4–L5 by an anesthesiologist with injection of 5 mg bupivacaine 0.5\% and 5 µg sufentanil. After performing spinal anesthesia, the patient was placed in the supine position. Anesthesia confirmation was performed with loss of cold sensation test, using an alcohol pad. The patient was then placed in a lithotomy position. Six L/min of oxygen therapy via a Hudson face mask was performed during the operation. The patient underwent continuous electrocardiogram, invasive blood pressure, and oxygen saturation monitoring during surgery. Hemodynamics of the patient was stable during the operation and changing in at least 20\% of baseline.

| Table 1. Patient serology test results |
|---------------------------------------|
| Hb=11.9 mg/dl                        |
| HCT=34%                               |
| BUN=8 mg/dl                           |
| PLT=256×10^3                         |
| INR=1                                 |
| Cr=1.4                                |
| WBC=9×10^3                            |
| M.C.V=80 fL                           |
| RBC=4.25×10^6                        |
| PT=12                                 |
| M.C.H=28 pg                           |
| BS=110                                |
| PTT=40                                |
| M.C.H.C=35 gr/dl                      |

**Figure 1: Right ventricular systolic phase**

**Figure 2: Right ventricular diastolic phase**

**Figure 3: Left ventricular systolic phase**

**Figure 4: Left ventricular diastolic phase**
The operation was completed uneventfully without pain or discomfort. After surgery, the patient was transferred to the recovery room and underwent monitoring. The patient was prescribed 1 g of Apotel in 100 ml of normal saline for infusion during 20 min to prevent postoperative pain. In addition, ranitidine 50 mg and ondansetron 4 mg were administered intravenously due to preventing of itching in the patient. She was then transferred to the coronary care unit (CCU) and underwent continuous cardiac monitoring. She was discharged after 24 h in good general condition without complications. Conscious consent was obtained from the patient to write the case report.

Discussion
Cardiomyopathy causes myocardial dysfunction by involving the myocardium. According to the ESC Classification of Cardiomyopathies, the main types of cardiomyopathy contain hypertrophic cardiomyopathy, dilated cardiomyopathy, arrhythmogenic right ventricular cardiomyopathy, restrictive cardiomyopathy, and unclassified cardiomyopathy. LVNC has been introduced as an primary genetic disorder by the American Heart Association, while the ESC described it as the unclassified cardiomyopathy.

In the unclassified or noncompacted types of left ventricle, it can be seen by prominent trabeculae of the left ventricle and deep intertrabecular recesses. The myocardial wall is often thickened with a thin, compact pericardial layer and a thick endocardial layer. In some patients, the LVNC is associated with LV dilatation and systolic dysfunction that can be transient in infants.

Echocardiography is recognized as the first diagnostic pathway with four diagnostic criteria: California criteria, Zurich criteria, Vienna criteria, and Milwaukee criteria. This type of cardiomyopathy can be also diagnosed by a cMRI. The present case report aimed to present a rare case of cardiomyopathy that was diagnosed by the echocardiography and CMRI in the unclassified cardiomyopathy group. Diagnosis of this type of cardiomyopathy in this patient was based on the California echocardiography as well as the CMRI criteria. Hypertrophy of walls causes permanent or intermittent left ventricle outflow tract obstruction. The rate of left ventricular outflow tract obstruction (LVOTO) increases in higher myocardial contractility, lower left ventricle end-diastolic diameter (increased heart rate and decreased venous return), and lower peripheral vascular resistance. Beta-blockers are widely used to control the patients’ symptoms because they reduce LVOTO (reducing contractility, decreasing heart rate) and are antiangiina and antiarrhythmic. The overadministration of prespinal or intraoperative fluid can cause severe postoperative symptoms. The lower intraoperative blood pressure can be treated with alpha-agonists such as phenylephrine. Beta-agonists are not indicated because they increase the heart rate and increase the risk of LVOTO. Monitoring is recommended for the diagnosis of subendocardic ischemia, cardiac arrhythmia, and hypotension. The accurate monitoring, availability of drugs, and defibrillators are recommended because of the possibility of urgent need. The continuation of monitoring is recommended in recovery because of the risk of pulmonary edema. Our patient was under the accurate monitoring in the CCU for 24; nothing happened by the treatment.

Conclusion
In this case report, we performed the anesthesia and surgery with the least complications by doing the spinal anesthesia with the lowest effective dose and the proper level of anesthesia with a procedure without any noticeable hemodynamic changes.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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