Anaesthesia Management of a Patient with Undiagnosed Hypertrophic Cardiomyopathy

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Dear Editor,

Hypertrophic cardiomyopathy (HC) is the most common autosomal dominant inherited cardiac disease and is the most frequent cause of cardiac disease-induced death among young people (1). Epidemiological studies report its prevalence to be 0.2% in the general adult population, and it has a high risk of sudden cardiac death (2). Moreover, it has a risk in anaesthesia practice because it cannot be diagnosed with routine preoperative evaluations and history findings in many cases. In this paper, it was aimed to share the case of a patient with HC who could not be diagnosed previously in view of literature.

Our patient was a 27-years-old, 168-cm tall male, and he weighed 69 kg. He underwent elective surgery due to American Society of Anaesthesiologists (ASA) I varicocele. In his preoperative evaluation, he and his family had no history of any disease. His physical examination did not reveal any symptom that could suggest a cardiac or respiratory disease. Moreover, the results of laboratory tests (haemogram, routine biochemistry, coagulation tests and serology) were within normal intervals. The patient was informed, and his written informed consent was obtained before operation. The patient was taken into the operating room for surgery, and he was monitored. Heart rate (HR), blood pressure (BP), electrocardiography (ECG) and peripheral oxygen saturation (SpO₂) were found to be normal. The patient, who was planned to be administered general anaesthesia in the intervention, was sedated with 2 mg of midazolam. Induction was then performed with 2.5 mg kg⁻¹ propofol; a classical laryngeal mask airway (LMA) was placed, and 50/50% O₂/N₂O was initiated with 1 MAC of sevoflurane. In the ECG monitor of the patient whose BP, SpO₂ and end-tidal CO₂ (ETCO₂) values were within normal limits, ST-depression and T-wave negativity suddenly occurred. When his ECG was taken, it was

observed that T negativity continued, and a blood sample was taken for cardiac enzyme evaluation. It was then decided that a cardiology consultation would be requested for the comprehensive examination of the patient whose ECG changes did not return to normal after being followed for a while under these conditions and that the patient would be awakened and the intervention would be postponed. After the end of general anaesthesia the patient was transferred to the intensive care unit for a monitored follow-up and treatment. In the echocardiography evaluation conducted by a cardiologist, it was observed that he had apical hypokinesia, apical hypertrophy and thin-septum structure, and his left ventricular ejection fraction was found to be 45–50%. The patient was referred to another health centre for an advanced cardiac examination.

The diagnosis of HC is established based on asymmetrical left ventricular hypertrophy, which is not associated with another cardiac or systemic disease in two-dimensional echocardiography. The cause of left ventricular hypertrophy is considered to be glycogen storage in the myocardium as a result of the mutation of two non-sarcomeric genes. Moreover, in two-thirds of these patients, primary malformation of the mitral valve, which leads to the obstruction of the left ventricle outflow can be seen, and intramural coronary arteries with thick walls and narrowed lumen can cause myocardial ischemia (2-4).

The data on the risks that patients with HC can have under anaesthesia are insufficient. It has been reported that complications, including congestive heart failure, myocardial ischemia, systemic hypotension and supraventricular and ventricular arrhythmia, can occur in these patients (5). Although myocardial ischemia and death are rarely seen, they can pose a danger during anaesthesia administration in patients with unspecified HC who are to undergo non-cardiac surgery.

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An invasive arterial catheter inserted before anaesthesia induction is important for the detection of hypotensive periods. Monitoring the end pressure of pulmonary capillaries is significant because it demonstrates the evaluation of real volume that occurs in association with a decreased diastolic compliance of patients. In patients with HC, increased or decreased intravascular volume can be dangerous for patients, and excessive fluid therapies applied for providing preload can lead to heart failure. Therefore, the main target must be to use drugs that suppress oxygen consumption and myocardial contraction at the lowest level while maintaining intravascular fluid volume and systemic vascular resistance and to provide an appropriate fluid volume. Considering pathophysiological mechanisms related to the obstruction of the left ventricle outflow, acute treatment strategies must be developed for the development of this condition and other complications associated with anaesthesia (6).

Considering the frequency of HC to be 0.2% in the population and the increasing rates of ambulatory surgeries, it is seen that the rate for anaesthetists to encounter a young patient with undiagnosed HC is not so low. In our case, HC was incidentally found, and it was defined with the analysis of the patient's parameters. The applications that would be life-threatening were probably prevented by this means.

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