CASE REPORT

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Anesthesia in Carvajal syndrome; the first case report

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ABSTRACT

Carvajal syndrome is a rare genetic disorder. Patients reporting for surgery pose some difficulties in anesthesia management. In this case report we present the case of a 12-year-old boy, who was a known case of Carvajal syndrome, referred for surgical resection of perianal condyloma. Close monitoring of hemodynamic status is the mainstay of anesthetic considerations in such patients. As in any other challenging scenario, it should be kept in mind that 'there is no safest anesthetic agent, nor the safest anesthetic technique; there is only the safest anesthesiologist'.

Key words: Carvajal syndrome; Anesthesia; Keratoderma; Cardiomyopathy

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INTRODUCTION

Carvajal syndrome (CS) is an autosomal recessive pattern duo to mutation in desmoplakin gene. It is a variant of Naxos disease with predominantly left ventricular involvement.[1] Also, known as striate palmoplantar keratoderma with woolly hair and cardiomyopathy. Clinical manifestations are usually apparent during childhood. Most of the patients develop heart failure and die due to syncope and/or sustained ventricular tachycardia during adolescence with a peak frequency in early adulthood.[2] These patients may be brought in to the operating room for different kind of surgeries. In this case report, we opted for light general anesthesia as the patient had moderate heart failure. We report this rare case after obtaining an informed written consent from the parents of the patient.

CASE DESCRIPTION

A 12 years old boy with a history of CS was admitted for resection of perianal condyloma. His disease was diagnosed during infancy and it was confirmed by genetic testing. Family history was notable for his two cousins, who were also diagnosed for CS. One of his cousins had died because of arrhythmia duo to dilated cardiomyopathy. On physical examination he was nervous and fearful. He had woolly hair, keratotic skin over palm, sole and lower half of both lower limbs, stomatitis, distal extremity weakness and easy fatigability (Figure 1). Electrocardiogram (ECG) demonstrated normal sinus rhythm (QTc= 430 msec), echocardiography revealed minimal mitral regurgitation (MR), mild dilated left atrium and left ventricle and decreased left ventricular ejection fraction (LVEF = 44%). He was not taking any medication and his weight was 30 kg.

The patient was brought to the operating room with a 20G intravenous (IV) line in dorsum of his right hand. 2 mg of midazolam and 20 mg of ketamine were injected to decrease anxiety and facilitate parental separation. Monitoring was started (pulse rate: 126/min; blood pressure 105/53 mmHg, SpO_2 95% in room air) and it was continued for 30 min after arrival to Post Anesthesia Care Unit (PACU) after which he was transferred to the ward. Because the expected surgery time was short, IV sedation anesthesia was planned for him. Inj fentanyl 30 μg and inj propofol 30 mg were injected and lithotomy position was made. Depth of anesthesia was monitored by BIS and propofol infusion was continued for anesthesia maintenance @75 μg /kg/min. Bag mask ventilation was used to assist the ventilation and efficacy of spontaneous ventilation was monitored by controlling of expiratory parameters, especially expiratory tidal volume on anesthesia machine. Vital signs were stable during the procedure and bispectral index (BIS)

monitoring was continued in a standard range (50-60). At the end of the operation the patient was waken up without any complication and transferred to the PACU after complete emergence. No subsequent sequel was observed.

DISCUSSION

CS is characterized by palmoplantar keratoderma, woolly hair, dilated cardiomyopathy, especially on the left ventricle side, and early morbidity.[3] The keratoderma involves mainly pressure areas in the palms and soles, and appears during the first years of life. CS with predominantly left ventricular involvement, early morbidity and clinical overlapping with dilated cardiomyopathy was described by Kaplan et al. from Ecuador in 1998.[4] It is a progressive heart disease and may cause sudden death in a child with early age. Progressive replacement of cardiac myocytes by adipose tissue and fibrosis can result in ventricular dilatation and dysrhythmia.[5] More than fifty percent of affected patients develop heart failure, and most of them die during adolescence.[1] A particular mutation in Ecuadorian families, that truncates the intermediate filament-binding site of desmoplakin, results in CS.[4] In our case, sedation/anesthesia for resection of a perianal condyloma in a 12 year old boy with moderate left ventricular failure is presented. Easy fatigability and reduced physical activity were the prodromal heart failure symptoms in this case. In patients at risk of arrhythmias and sudden death, close monitoring of hemodynamic parameters is the mainstay of safe anesthesia practice.[6] Cardiac study by means of echocardiography and electrocardiography can help in detecting most of the structural and electrophysiological abnormalities with major impact on anesthesia plan. Cardiomyopathies per se require meticulous perioperative care. Maintenance of adequate preload, sustained normal heart rate and avoiding increase in systemic vascular resistance are the principles of anesthetic management. According to these considerations, cardiovascular support, invasive hemodynamic monitoring and postoperative care in an intensive setting should strongly be considered in patients with severe disease undergoing lengthy procedures[7] Because CS is a variant of arrhythmogenic right ventricular cardiomyopathy (ARVC), close monitoring of heart rate and preparing for appropriate intervention following any perioperative dysrhythmia is inevitable.[8] For this reason 5 lead ECG monitoring was applied for our patient, defibrillator was kept ready in whole perioperative period and heart rate maintained 80-100 beats/min as Yildiz et al. recommended in the first case report of anesthesia for Naxos disease.[5] Near normal ventricular function permitted us to administer commonly used premedication drugs with standard doses. In the same way, minimal anesthetic

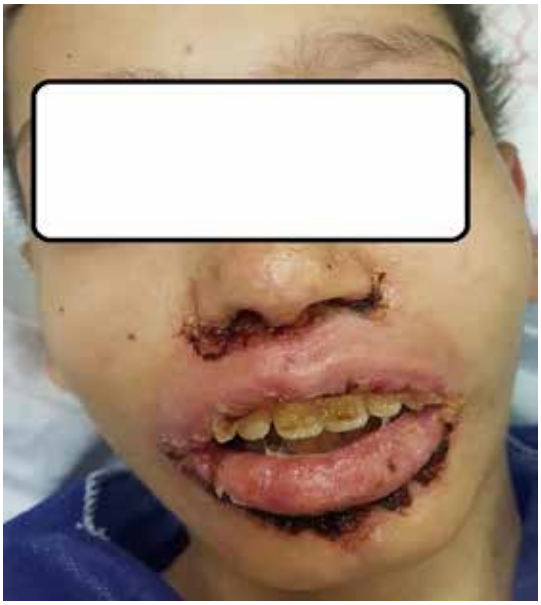

Figure 1: Facial features of CS

dose of ketamine, a small bolus dose of propofol as an induction agent, and followed by a cool maintenance infusion of propofol resulted in stable hemodynamic parameters, avoiding hypotension that is critical in the case of left ventricular failure. However, etomidate could have been a good alternative for anesthesia induction in such cases at risk of hemodynamic perturbations. Because of short duration of surgery, we decided to apply bag mask ventilation for airway management and maintain adequate oxygenation but presence of peri-oral lesions forced us to apply vitamin A ointment beneath the face mask cushion. However, use of more advanced airway devices such as laryngeal mask airway can be considered for longer procedures without any further risk. Short duration of the procedure beside near normal ventricular function allowed us monitor the hemodynamic state by the use of noninvasive monitoring instead of invasive methods, thus avoiding skin trauma and arterial puncture in a patient with a vulnerable skin. However, arterial catheterization or transesophageal echocardiography with the aim of beat to beat control of ventricular function is sometimes prudent, especially in hemodynamically unstable patients for lengthy or more complicated procedures.[7]

In conclusion, any anesthetic procedure in patients with CS requires standard monitoring and close observation, because anesthesia may lead to myocardial depression, heart failure, dysrhythmia and death. In other words, in CS, anesthesia is nothing but monitoring and preparation to deal any adverse event. As in any other challenging scenarios, it should be kept in mind that " there is no safest anesthetic agent, nor the safest anesthetic technique; but the safest anesthesiologist".[9]

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Authors' contribution:

SS: Conceived idea, wrote the primary manuscript

RA: Wrote the discussion section and edited the entire manuscript

AS: Revised the manuscript, contributed to the final version

MM: Performed the surgery, contributed to the final version.

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