

Anesthesia Management of Escobar (Multiple Pterygium Coli) Syndrome Case Report

Escobar (Multiple Pterygium Coli) Sendromunda Anestezi Yaklaşımı Olgu Sunumu

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ABSTRACT

Escobar syndrome is a genetic disease advancing with various malfunctions. Anesthesia application on these patients is of consideration due to risk of respiratory difficulties and malignant hyperthermia development.

In this presentation, our anesthesia approach on the 1 year old female diagnosed with Escobar syndrome, and to have a pyeloplastic operation due to uretero-pelvic stenosis.

Key words: Escobar syndrome, Anesthesia

ÖZET

Escobar sendromu genetik olup, çeşitli malformasyonlarla seyreden bir hastalıktır. Havayolu zorluğu ve malign hipertermi gelişme riski açısından, bu hastalarda anestezi uygulaması özellik arz etmektedir.

Bu sunumda, Escobar sendromu tanısı almış olan, üretero-pelvik darlık nedeniyle piyeloplasti operasyonu geçirecek olan, 1 yaşındaki kız çocuğunda anestezi yaklaşımımız sunulmuştur.

Anahtar Kelimeler: Escobar Sendromu, Anestezi

INTRODUCTION

Escobar (multiple pterygium coli) syndrome is a rarely encountered autosomal recessive syndrome characterized with pterygia, camptodactile, syndactile and cleft palate, and developing with restrictive lung disease, dyspnea attacks, recurrent pneumonia and cardiac defects (1,2). Due to decreased mouth clearance, serious micrognathia, anchiloglossy, cervical fusion, presence of cifoscoliosis, current lung and cardiac abnormalities and risk of malignant hypothermia, anesthesia application on these patients is of consideration.

In this presentation, our anesthesia approach on the patient with Escobar syndrome, and to have a pyeloplastic operation due to uretero-pelvic stenosis.

CASE REPORT

A pyeloplastic operation has been planned for the 1 year old patient weighing 5.500 grams, monitored with diagnosis of Escobar syndrome, due to uretero-pelvic stenosis. In the pre-operative examination, the patient has been classified as difficult respiration due to decreased mouth opening, micrognathia and limited neck extension (Figure 1). While the pre-operative cardiologic examination was within the normal limits, no pathologies have been detected in the blood analysis and lung graphs. In accordance with the criteria of the American Anesthesia Association, all the pre-operative equipments were present. The patient has been pre-medicated with 0.08 mg/kg rectal midazolam and EMLA cream has been applied onto the area where the intravenous catheter was planned to be inserted. Pulse blood pressure and oxygen saturation of the patient has been monitored in the operation room. The vessel route has been opened on the hand with intravenous catheter anesthesia induction has been commenced with 0.015 mg atropine sulfate and propofol, after the pre-

oxygenization. The patient's spontaneous respiration has been protected and ventilation has been tried with face mask and as there was no difficulty with the mask respiration, after the sufficient anesthesia has been provided, laryngoscope condition has been determined to be Cormack-Lehane Class III using a direct laryngoscope. However, the external laryngeal pressure and style 4 mm diameter endotracheal tube have been intubate at the first try. Anesthesia maintenance has been provided in 70 % N₂O, 30 % O₂ and with infusion of propofol at a rate of 60-160 mcg/kg/minute and muscle slackness with sis-atracurium. During surgery, 0.05 mg/kg sis-atracurium has been given as additional dose as needed. During surgery, the end tidal carbon-dioxide value, body temperature and urine output have been monitored with urine catheter. The myoglobine level of the urine has been determined and serum electrolytes, protein, glucose, lactate and blood gas values have been monitored hourly following the anesthesia induction. During the operation of three hours, all the blood gas and biochemical values of the patient were within the normal limits and vital findings were stable. The patient with full respiratory reflexes and breathing spontaneously has been extube at the end of the operation. 1 mg/kg meperidin has been titrated intravenously for post-operative pain treatment and 20 mg/kg paracetamol has been applied rectally.

Though the myoglobuline level of the urine increased at the 1st post-operative hour, it was within the normal limits. The patient has been sent to normal service beds without complication after monitoring.



Figure 1: A pyeloplastic operation has been planned for the 1 year old patient weighing 5.500 grams, monitored with diagnosis of Escobar syndrome, due to uretero-pelvic stenosis.

DISCUSSION

The articles published in the literature regarding the anesthesia approach to the Escobar syndrome cases defined in the year 1978 commonly argue respiration difficulties (1,5). Decreased mouth opening, micrognathia, cleft palate, pterigiums on the neck and vertebra abnormalities accompanying the syndrome most frequently cause respiratory difficulties.

In this case, during the pre-operative anesthesia examination, the possibility of airway difficulty has been foreseen during the physical examination of the patient and pre-surgical preparations have been provided. However, the current cardiac and respiration system problems of these patients are of the same significance as the airway difficulties (4). Repeated pneumonia attacks are accompanied by problems related with frequent cifoscoliosis pre-operative lung ventilation (5).

Increased malignant hyperthermia frequency that has not been discussed in the literature before is of a life-threatening level. In this presentation, we have considered the risk of malignant hyperthermia development and preferred the intravenous anesthetic agents and monitored the blood gas and biochemical values and myoglobin levels in the urine of the patient, contrary to the literature. While no change was observed in the blood gas and biochemistry levels, in the post-operative period, even though the urine myoglobin level of the urine has increased, it remained within the normal limits. However, as the metabolic acidosis to arise due to maintenance of anesthesia with propofol infusion may be confused with malignant hypothermia, the blood gas analysis of these patients need to be evaluated together with other malignant hyperthermia indications such as hypercarbia and sudden increases of the body temperature (6).

For the anesthesia planning of the Escobar syndrome cases, pre-operative measures should be taken against potential respiratory difficulties and during and after

application of anesthesia, patients should be closely monitored with consideration of risks of development of current lung and cardiac problems and malignant hyperthermia.

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