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Case Report

Our Anesthesia Experience in a Patient with Neurofibromatosis

Ozkan Onal[1], Cansu Ciftci[1], Emine Aslanlar[1], Fatmanur Erdoğdu[1], Jale Bengi Celik[1]
Selcuk University Medical Faculty Department of Anesthesiology and Intensive Care, Konya, Turkey

***Corresponding author**
Dr. Ozkan Onal
Email: drozkanonal@selcuk.edu.tr

Abstract: Neurofibromatosis (NF) is an otozomal dominant and multisistemic disorder. Two types were defined: Neurofibromatosis type 1 (NF1) and Neurofibromatosis type 2. Neurofibromas can be found multisystemic and can be cause difficulty for either general anesthesia or regional anesthesia. The patient is 4 years old, 15 kilograms weight, who has common neurofibroms mostly at back and cafe au lait in whole body. She was planned to undergo operation for left inguinal hernia. After 8mg/kg propofol and 2mg/kg fentanyl administration, i-gel was placed and general anesthesia was administered. End of the operation the patient was awakened without any problem. The vital findings of the patient were stable intraoperatively. In postoperative period, no neurological or anesthetical complications occurred and the patients was transferred to the clinic. With this case report, we want to tell anesthesic method in pediatric patient with NF1.
Keywords: Neurofibromatosis (NF), multisistemic disorder, anesthesia

INTRODUCTION

Neurofibromatosis is an autosomal dominant genetic, multisystemic diseases and its two forms have been defined. Neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). NF2, occurs more rarely. NF1 (Von Recklinghausen disease, peripheral neurofibromatosis) is a neurocutaneous disease which influences cell growth primarily in neural tissues and involves many systems. "Cafè au lait" spots are the earliest findings. Neural tumors may appear and optic gliomas may lead to progressive loss of vision. Neurofibromas, which is the most important characteristic finding, of the disease, may make anesthesia administration more difficult, by affecting more than one system. Neurofibromas may obstruct upper airways and may close spinal needle hole during regional anesthesia. We wanted to share our experience of anesthesia in patient with NF1.

CASE REPORT

A 4 year old female patients at the weight of 15 kg. had many cafe au lait spots all around her body at different sizes and neurofibromas mostly at back (Figure 1). She was followed with the diagnosis of NF1 and was planned to undergo operation for left inguinal hernia. In her history, it was learned that she was followed for a long time in neonatal intensive care unit intubated with thorax tube due to pneumothorax associated with barotrauma after a premature birth at 31 weeks. In preoperative evaluation, no respiratory distress was observed and under ambient conditions, oxygen saturation value measures as 97%. Pediatry department was consulted about physical examination and when it was seen that neurofibromatosis was limited to skin involvement, no drawbacks for operations were seen. Preoperative laboratory findings were normal.

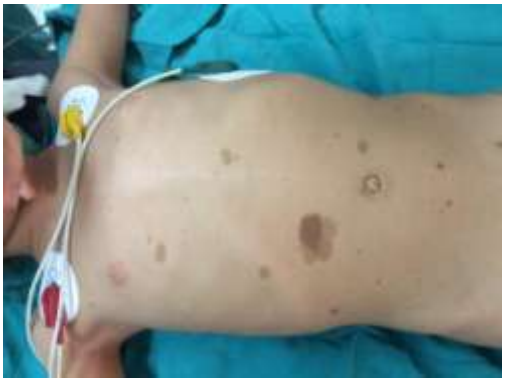

Fig-1: Our patient with Neurofibromatosis.

Routine monitorization electrocardiogram (ECG), pulse oximeter, noninvasive artery pressure was carried out. Vital finding were stable. In one of the dorsal hand veins, catheter was placed with 22 G canula and isotonic sodium chloride infusion was initiated at the rate of 100 ml/ hour. After 8mg/kg propofol (Fresenius Kabi, Istanbul ,Turkey), 2mg/kg fentanyl (Talinat,Vem Drug, Ankara, Turkey) administration, 2 no i-gel was placed and general anesthesia was administered. Ventilation was provided with flow 3,5 lt/min. %40 oxygen and anesthesia maintenance and analgesia was carried out with 0,15mg/kg min. Propofol and 0,15 mcg/kg/min. remifentanyl (Ultiva, Glaxo Smith Kline, Istanbul, Turkey) infusion. Following the

operation lasting 30 minutes, the patient was awakened without any problem and then monitorized in recovery room. The vital findings of the patient were stable intraoperatively. In postoperative period, no neurological or anesthetical complications occurred and the patient was transferred to the clinic.

DISCUSSION

Neurofibromatosis is a multisytemic disease and in this groups patients, anesthesia management requires care. In anesthesia management, difficulties may be encounterd due to present systemic abnormalities and hemodaynamic responses of the patients may be diffeerent. NF may involve airways, lung parenchyme and chest cage [1]. Thoracic symptoms include neurofibromas, interstial lung disease, costal deformations and kyphoscolisosis. In addition, aorta coarctation, cardiomyopathy and renovascular diseases occur commonly as well [2]. Neurofibromas may be present in central nervous system, cardiovascular system and respiratory system. In patients with NF1, neurofibromas may be seen in base of the tongue and may cause difficulties during aeration and intubation of the patients. In patients with symptoms of cough, wheezing and dyspnea, the presence of neurofibromas obtructing airway should be considered. Mandibular anomalies and macroglossia are other factors making anesthesia management more difficult [2-4]. In NF1 the development of bilateral diaphragmatic paralysis due to the involvement of phrenical nerve roots has been reported [1,5]. In cardiovascular system, it mostly presents with hypertension and pheochromacytoma occurs more frequently in neurofibromatosis compared to general community [6]. The involvement of respiratory system and cardiovascular system may influence the hemodynamic response of the patients to anesthesia. In patients with neurofibromatosis, deciding on suitable anesthesia management requires a thorough systemic evaluation. For this purpose, at first airway should be evaluated and subsequently respiratory, cardiovascular and central nervous system involvements and the presence of vertebral anomalies should be evaluated. Considering that cranial or spinal involvement may occur and regional anesthesia may impair present neurological status in asymptomatic patients, general anesthesia may be considered the preferred method. However, in general anesthesia, the probability of difficult intubation owing to neurofibromas located in oropharynx and pharynx and the fact that a hemodynamically stable anesthesia may be difficult due to multisytemic involvement should be borne in mind. Furthermore, in patients with neurofibromatossis, there may be variable sentitivity to succynilcholine and neuromuscular blocker agents. Although the risk of abnormal response to neuromuscular agents has been reported to be minimal, currently routine neuromuscular monitorization via peripheric nerve stimulator is recommended during anesthesia in patients with neurofibromatosis [2,7]. In the present case, as the patients could not adapt to spinal regional anesthesia due to her age, we decided on general anesthesia. As the operation was predicted to last short and the probability of difficult intubation was present, we thought that i-gel application would be suitable. Anesthesia induction and management and ventilation was uncomplicated during operation. In perioperative period, patient was quite stable hemodynamically. This argued against probability of cardiovascular anomalies and pheochromacytoma. After the operation, patient was awakened without any complications.

In conclusion, when deciding between general anesthesia or regional anesthesia modalities in a patient with neufibromatosis, the benefit of the patient's should be considered and in preoperative period, a complete systemic evaluation must be made. Preoperative evaluation of spinal cord will influence the decision on neuraxial anesthesia and hemodynamic status of the patient and characteristics of multisytemic involvement will affect the decision in favor of general anethesia. Irrespective of the anesthesia method that will be employed, it should be considered that in these patients the probability of difficult intubation is high and anesthesia administration should be initiated after all preparations are made for the safety of airway.

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