Yeditepe Medical Journal 2009;(10):198-201

Anesthetic Management of Maffuccı's Syndrome Out Side the Operating Room: Case Report

Maffucci Sendromlu Hastada Ameliyathane Dışı Anestezi Uygulaması: Olgu Sunumu

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ABSTRACT

Maffucci's syndrome is a congenital disease characterized with enchondromatosis and multiple soft tissue haemangiomata due to neuro-ectodermal dysplasia. Maffucci's syndrome is a rare clinical condition that presents difficulties concerning its anesthetic management.

In this presentation, our anesthesia approach on the patient with Maffucci's syndrome with multiple haemangiomata along the upper airways requiring intralesional sclerotherapy is described.

Keywords: *Maffucci's Syndrome, Anesthesia*

ÖZET

Maffucci sendromu, nöro-ektodermal displaziye bağlı olarak gelişen, enkondromlar ve multiple yumuşak doku hemangiomları ile karakterize konjenital bir hastalıktır.

Bu sunumda, üst hava yolunda çok sayıda hemangiomları olan ve lezyon içine sklerozan madde enjeksiyonu yapılan hastada anestezi yaklaşımımız sunulmuştur.

Anahtar Kelimeler: *Maffucci Sendromu, Anestezi*

INTRODUCTION

Maffucci's syndrome, is a congenital osteochondromatosis, which develops associated to neuroectodermal dysplasia, and progresses with hemangiomas, asymmetric enchondromas and extremity development (1). Enchondromas may occur in long bones, hands, feet or pelvis, and fractures are encountered frequently

due to enchondromas (2). Enchondromas, depending on their location, may make it difficult or even impossible to put the patient in position during anesthesia (3). Hemangiomas associated with the disease on the other hand are located at subcutaneous level or dermis. Difficulties may be encountered in both airway management and peripheral venous catheterization associated with the location, size and number of hemangioma (3).

This report presents our anesthetic approach outside the operating room in a patient with Maffucci`s syndrome who underwent intralesional injections to multiple hemangiomas arising from the root of the tongue and leading to upper respiratory obstruction by protruding to oropharynx.

CASE REPORT

A 10 years old, female patient was scheduled for sclerosant substance injection into the hemangiomas which is located in oral cavity at the radiology unit. She has a history of respiratory arrest requiring emergency tracheostomy when she was 5 years old. During the evaluation of the aforementioned hospital admission widespread hemangiomas was found in her mouth, she had been later diagnosed with Maffucci`s syndrome. In her follow-up, tracheostomy was again performed at the age of 10 due to the respiratory distress. Until that hospital admission she had been living without a tracheostomy cannula for the past five years.

Angiographically guided sclerosant substance injection was planned into the patient's lesions. Hemangiomas were extending from the root of the tongue to vallecula. During preoperative examination tracheostomy cannula and hemangiomas in the mouth arising from the root of the tongue and fully obstructing upper respiratory tract was observed (Fig. 1).

Figure 1: *Hemangiomas in oral cavity.*

Following the local anesthetic cream (EMLA % 5 krem, AstraZeneca, Türkiye) application, an intravenous catheter was placed on the hand of the patient who was agitated following the 6 hours starvation period. Premedication was given intravenously by administering 1 mg of midazolam, and the patient was taken into invasive radiology unit. Heart rate, non-invasive blood pressure, and oxygen saturation with pulse oxymetry were monitored. Following anesthesia induction with intravenous propofol (Propofol, Abbott, Turkey) (2 mg/kg) and fentanyl (Fentanyl, Abbott, Turkey) (1 mcg/kg), muscle relaxation was achieved with mivacurium (Mivacron, Glaxo Smith Kline, Turkey) (0.2 mg/kg). Mechanical ventilation was initiated by means of tracheostomy cannula. Anesthesia maintenance was sustained with sevorane titration in 60% N2O, 40% O2. End-tidal carbon dioxide values were monitored throughout the procedure. Extremities of the patient were supported by silicon pads. Hemangiomas in oral cavity were sclerosed in a period of approximately 2 hours (Fig. 2). There were no active bleeding in the surgical area at the end of surgery and vital symptoms were stable perioperatively. Patient was transferred to her room when she was fully awake. The lesions were regressed in later follow-ups and the patient was decannulated approximately six months after the operation.

Figure 2: *Peroperative surgical approach.*

Figure 3: *Postoperative evaluation of the airway.*

DISCUSSION

Maffucci syndrome was first defined by Maffucci in 1881. This syndrome is characterized by enchondromas, bone deformities, and irregular placement of hemangiomas (4). This is a rare, nonhereditary, congenital disorder of unknown origin, and anesthetic management is also specialized (3). While information with regards to anesthetic approaches is rarely encountered in the literature, surgical diagnosis and approach is generally emphasized (1-5). Whereas, as has been highlighted in this presentation, extreme caution should be demonstrated during positioning of the patient with Maffucci syndrome due to the risk of fractures that may be caused in connection to enchondromas (5). Extremities of our case were supported by position pads throughout the anesthesia period to prevent any possible problems as skin lesions, peripheral neuropathies. Care should be taken to prevent extremity fractures while positioning these patients. No problems were encountered during positioning and operation in our case.

Another issue in patients with Maffucci syndrome is the difficulty in vascular access due to lesions at the back of hands and feet (5). However no problems were encountered in venous access in this case.

As a general anesthetic approach in anesthesia performed outside of the operating room, thorough evaluation of the patient prior to invasive procedure, anesthesia device, oxygen system, aspirator, emergency equipment, monitor, complete availability of anesthesia and resuscitation medicine as well as close monitoring of the patient during invasive procedure are important (6). In this patient, where invasive procedure was performed on the lesion blocking upper respiratory tract, problems could have been encountered arising from removal of tracheostomy cannula. In our case, hemangiomas located in the mouth may lead to respiratory problems. Therefore all equipment required for an emergency airway problem was kept ready in pre- and early postoperative period.

During intralesional injections, damages that might occur due to the radiation are important. In such invasive procedures, where there may be radiation pollution, anesthetist needs to take precautions oriented towards self-protection (7).

Preoperative evaluation of a patient with Maffucci`s syndrome should include a careful examination of the airway, and veins to be cannulated to prevent the

potential difficulties of airway management, and intravenous access. A careful preoperative evaluation and perioperative management provided a successful outcome in this case. All equipment should be kept ready for emergency situations, especially when the procedure was planned out side the operating theatre.

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