

Anesthetic management of neurofibromatosis type 1 patient

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Abstract

Neurofibromatosis type 1 is an autosomal dominant neurocutaneous disorder. It is multisystemic disorder with multifaceted implications throughout nearly every organ system. Anesthesiologists must perform a through preoperative evaluation of patient. Moreover the anesthesiologist must consider the possibility of each of the multisystemic complications when evaluating and managing the patient for surgical procedures. (J Med Life Sci 2013;10(1):44-46)

Key Words : Neurofibromatosis, Anesthesia, Management

Introduction

Neurofibromatosis type 1 is an autosomal dominant inherited disorder characterized by multiple neurofibromas and cafe-au-lait spots. Patients with neurofibromatosis type 1 require caution during anesthesia since neurofibromas present in tongue and laryngopharynx could interfere with endotracheal intubation or neurofibromas present in the spinal cord could deteriorate pulmonary function by inducing scoliosis or kyphosis.

Case Report

A 56-year-old female patient with a height of 158 cm and weight of 52.6kg was diagnosed with neurofibromatosis type 1. Since her childhood, the patient had some skin lesions similar to a small lump and the lesion at the back did not protrude to cause any trouble in her daily activity. However, a skin lesion on the back became bigger enough to be saggy from a week before the hospital visit. She was admitted through the emergency room since bleeding had started from the lower part of the lesion (Figure 1).

Fever was associated with the finding of a decreased hemoglobin level of 7.3 g/dL according to blood test. In addition, she was suspected of the finding of a rapid expansion due to intra-tumor bleeding according to a

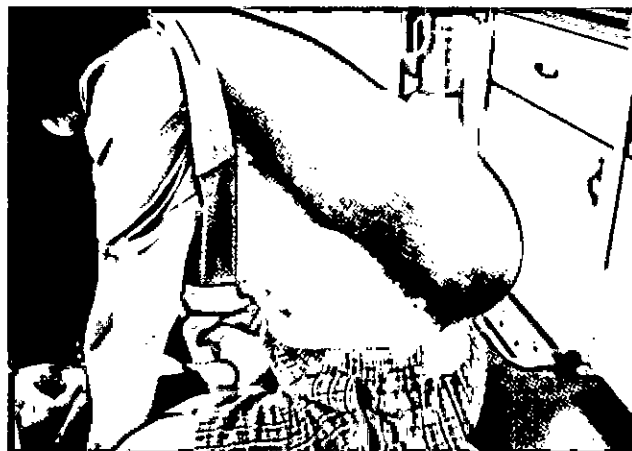


Figure 1. Appearance at the time of admission the patient's skin lesion.

vascular computed tomography. Cerebral and thoracoabdominal computed tomography and endoscopic examination were conducted preoperatively. No specific findings were detected other than the lesion on the back and subcutaneous lesions in the body. However, the patient complained inconvenience of having difficulty in lying on a supine position, daily activity, and particularly sleeping. Embolization was performed in blood vessels supplying nutrients to the tumors by requesting to the radiology department one day preoperatively.

Since the patient had difficulty in taking a supine position on the day of surgery due to the lesion at the back, arterial blood monitor was placed for the patient after measuring ECG, oxygen saturation, and blood pressure in a lateral decubitus position. The patient underwent anesthesia with a

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glide scope and a bronchoscope prepared to be used in case of undetected lesions that might result in difficulty in endotracheal intubation. After sufficient preoxygenation with 100% oxygen, anesthesia was induced using a 200 mg of thiopental and the ventilation of oxygen mask was checked. Subsequently, endotracheal intubation was conducted through a reinforced tube with an inner diameter of 7.0 mm under laryngoscopy after administering a 40 mg of rocuronium. Any specific resistance or endotracheal lesions were not observed during intubation. Although embolization was performed in advance, the MAC™(Two-lumen central venous access set, ARROW) catheter was inserted into the central venous line under the induction of ultrasonic waves in veins of right internal jugular vein by taking the risk of massive bleeding into consideration due to the enlarged lesion. Rocuronium was steadily administered at a 20 mg/hr by maintaining anesthesia at a 20 mg/hr of sevoflurane, a 1.5 L/min of oxygen, and a 1.5 L/min of nitrous oxide intraoperatively. Sodium nitroprusside was constantly injected at a 0.5mcg/kg/min to minimize bleeding and systolic blood pressure was maintained in between 100~120 mmHg. After the patient was confirmed with normalized voluntary respiration postoperatively, extubation was conducted in the operating room. Due to the potential risk of unexpected difficulty in breathing and sudden bleeding, she was observed in the intensive care unit for three days postoperatively before being transferred to the general ward.

Discussion

Neurofibromatosis type 1 was first documented by Freidrich von Recklinghausen in 1882 and named after the researcher also known as von Recklinghausen's disease, recognized as tumors arising from nerve tissues. Since neurofibromas might influence both the ectoderm and endoderm, lesions and associated symptoms could be involved during anesthetic procedure throughout the body including the central nervous, respiratory, cardiovascular, musculoskeletal, digestive, and genitourinary system. Thus, caution is essential with anesthesia. In cases where difficulty in swallowing, dysarthria, voice change, and other symptoms are particularly associated in the aspects of airway management, performing intubation may be challenging due to lesions in tongue and laryngopharynx. Since pulmonary lesions could induce difficulty in breathing

due to the advancement of pulmonary fibrosis or spinal deformity, the presence of such lesions needs to be carefully examined. Patients with a suspected bleeding like the patient in this case report need to be taken into consideration the fact that bleeding may occur due to undetected lesions in gastrointestinal tract[1]. Therefore, anesthetists need to be equipped with anesthetic plans and adequate countermeasures regarding any potential complications through sufficient preoperative assessment and history taking in all patients[2]. Patients with neuromuscular diseases are known to be sensitive to nondepolarizing muscle relaxants like the patient in this case report. For this reason, the use of TOF (train of four) or DBS (double burst stimulation) is recommended to steadily monitor the neuromuscular system[3]. We conducted extubation after confirming that the patient restored voluntary breathing and sufficient muscle relaxation of clenching her fist for more than five seconds. However, the study had a limitation of insufficient use of the neuromuscular system monitoring that could be taken as an objective indicator. Additional risk factors associated during anesthesia were not detected in preoperative examination including tracheal lesions, spinal deformity, or lesions in abdominal cavity. Moreover, she had recovered well in muscle relaxation and minimizing bleeding.

It was difficult in maintaining a posture due to neurofibromas. Factors causing trouble in airway management during anesthesia were masses in the neck area and at the back restraining a supine position and requiring caution. Methods enabling anesthesia in a supine position have been introduced by making holes in table for patients who are anticipated to have difficulty in trachea management due to associated tracheal deviation during preoperative assessment due to masses at the back[4]. Alternative methods were prearranged for the situation of complicated intubation with no anatomical deformity in airway management in spite of unsecured supine position preoperatively. Intubation under bronchoscopy was smoothly performed in lateral position. Ultrasonography was used to minimize complications such as arterial puncture or damage in adjacent tissues that could arise during central venous cannulation due to changed lateral position by safely placing central venous line. Therefore, anesthesia was able to be safely induced without any particular problems.

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