

# Anesthesia management in a pregnant patient with neurofibromatosis

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## Abstract

Neurofibromatosis (NF) is a genetic and multisystemic disease with autosomal dominant transition. It can affect anesthesia applications by affecting more than one system. In the presence of neurofibromas in the airway, it can cause airway obstruction and difficulties in respiratory delivery during general anesthesia. The presence of tumors affecting the central nervous system makes spinal anesthesia risky. Anesthesiologists should act with awareness of each multisystemic complication when evaluating and managing patients. This case report aims to present our general anesthesia practice in an elective cesarean operation in a pregnant patient with neurofibromatosis type 1.

**Keywords:** Neurofibromatosis, Pregnant, General anesthesia

## Introduction

Neurofibromatosis is an autosomal dominant disease and is divided into two groups: neurofibromatosis type-1 (NF-1) and neurofibromatosis type-2 (NF-2) [1]. Type 1 neurofibromatosis, also known as von Recklinghausen disease, is the most common type and is characterized by café-au-lait spots and benign skin neurofibromas. Type 2 neurofibromatosis affects the central nervous system due to spinal cord tumors and bilateral vestibular schwannomas [2, 3]. In NF-1, neurofibromas in the tongue, pharynx, and larynx can prevent intubation by making the airway more constricted. For this reason, in pregnant women with NF-1, difficult airway has been the main cause of anesthesia-related deaths. In these cases, it is important for anesthesiologists to perform airway examinations carefully [4]. Increasing the risk of bleeding while applying regional anesthesia, there is an increased risk of hematoma and intracranial pressure. However, successful computer spinal anesthesia has been reported in patients by excluding the presence of spinal neurofibroma by brain computer tomography (CT) and magnetic resonance imaging (MRI) [5]. We present our anesthetic approach to cesarean surgery performed under elective conditions in a pregnant patient diagnosed with NF-1.

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The authors stated that the written consent was obtained from the patient presented with images in the study.

### Conflict of Interest

No conflict of interest was declared by the authors.

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## Case presentation

A 38-year-old pregnant woman weighing 72 kg was brought into the operation room after receiving a confirmation form due to cesarean surgery. In the preoperative evaluation, it was determined that she had hypothyroidism for which she used drugs; she smoked two cigarettes per day. It was determined that she had been operated on ten years ago for a cesarean birth. In the physical examination performed in the preoperative period, it was determined that there were diffuse neurofibromas and café-au-lait spots on the body (Figures 1, 2). Patient consent was obtained for the images presented in the study. No mass was observed in the oral examination and this was evaluated as Mallampati II. Respiratory, cardiovascular, and neurological system examinations of the patient were normal. There were no restrictions in the mouth opening or neck movement. Apart from the high TSH, no abnormal results were found in the preoperative laboratory tests. In terms of hypothyroidism, an internal medicine doctor was consulted preoperatively and his recommendations were followed. Operational risk was determined as ASA 2 according to the classification of the Association of Anesthesiologists (ASA).

Figure 1: Diffuse neurofibromas



Figure 2: Café-au-lait spot



When she entered the operating room, first, her heart rate (HR), noninvasive blood pressure (TA), and peripheral oxygen saturation (SpO<sub>2</sub>) were monitored. Her TA value was 130/85 mmHg, HR 90 / min, SpO<sub>2</sub> 97. Vascular access was opened with 18 G and 20 G cannula from both hand ridge veins, preop 500 mL isotonic sodium chloride was given, and fluid infusion continued during the operation. Due to the absence of preoperative computed tomography or magnetic resonance imaging, we chose general anesthesia because we could not rule out spinal cord neurofibroma. We made the necessary preparations for the possibility of difficult intubation preoperatively. We applied 2 mg/kg propofol, 0.6 mg/kg rocuronium in anesthesia induction and used sevoflurane in maintenance. By ensuring sufficient preoxygenation, we intubated the patient without any problems with an endotracheal tube with an internal diameter of 7 mm. No hemodynamic problem was detected during the operation. At the end of the operation, the patient was sent to recovery, where she was followed up without any problems.

## Discussion

Neurofibromatosis is an inherited disease with an autosomal dominant transition and is classified in 2 types. NF1 (von Recklinghausen disease) is the more common type and is characterized by benign neurofibromas (cutaneous neurofibroma) of the skin and brown skin patches (café-au-lait spots) [2, 3, 6, 7]. NF-2 occupies the central nervous system. The bilateral vestibular schwannomas leading to gradual hearing loss are characteristic [8]. Features, such as meningioma of the brain, cranial, spinal or peripheral nerve schwannoma and juvenile cortical cataracts, may also be present [9].

In NF-1 patients, neurofibromas pressing on the tracheobronchial system and located in the lung parenchyma, chest wall deformities with severe scoliosis and kyphosis are commonly detected [10, 11]. In NF-1, pheochromocytoma or hypertension associated with renal artery stenosis can be seen [8, 9]. Neurofibromas can also affect the gastrointestinal tract and can be carcinoid tumors in the duodenum [12, 13]. The presence of cervical neurofibroma, laryngoscopy, and tracheal intubation can cause spinal cord injury, and, therefore, radiographic examination of the neck is recommended prior to anesthesia application [14]. In NF-1, urethral obstruction of retroperitoneal neurofibromas can be seen, and genitourinary system disorders such as hydronephrosis are also common. This may cause difficulty in bladder catheterization [15]. In addition, it has been reported that NF-1 patients have increased sensitivity to nondepolarizing neuromuscular blocking drugs [16, 17].

In neurofibromatosis, macroglossia, abnormal formations in the tongue, presence of plexiform fibromas in the pharynx, larynx, and supraglottic region can prevent endotracheal intubation and cause upper airway obstruction during anesthetic induction [2, 4, 18]. For this reason, in the patient, dysphagia, dysarthria, presence of stridor, and voice changes should be questioned and these lesions should be questioned [2]. The involvement that causes facial malformations can cause facial asymmetry, and may contribute to facial mask and difficult ventilation [19].

General anesthesia is considered safer since the presence of intracranial neuromas or unknown spinal neuromas in neurofibromatosis may cause destructive complications, such as hematoma and stroke [20-22]. Gliomas, meningiomas, hydrocephalus, spinal tumors, and spina bifida have been identified in NF-1, and these findings prevent the use of regional anesthesia [18]. Esler et al. [23] reported that an undiagnosed NF-1 patient had difficulty in analgesia after an epidural block application and an epidural hematoma occurred in the patient. In addition, the presence of skeletal anomalies, such as kyphosis and kyphoscoliosis, in patients with neurofibromatosis may cause difficulties in regional anesthesia application [21]. There is consensus that it is correct to put on a regional anesthetic indication after proving that there are no neurofibromas in the central nervous system by using imaging methods such as computed tomography and magnetic resonance in patients with NF [5]. In addition, successful neuroaxial anesthesia applications have been reported [22-24].

In our patient, findings of upper respiratory obstruction and facial malformation were not available. CT can be used to detect upper airway neurofibromas. However, the CT imaging

method could not be performed in the preoperative period due to the pregnancy of the patient. Despite these risks, we preferred to perform general anesthesia because our patient had a history of uneventful operation. Her mallampati score was 2, and there was no central nervous system image of the patient. However, we took the necessary precautions against the possibility of difficult airway. Our patient remained peroperatively hemodynamically stable.

Due to airway management, respiratory and cardiovascular problems, central nervous system structure, and vertebral anomalies, it may be difficult to choose an anesthesia method in neurofibromatosis patients [22]. A careful systemic evaluation is required to decide the appropriate anesthetic method.

### Conclusion

When deciding on the anesthesia method to be applied in a pregnant patient with neurofibromatosis, we should thoroughly evaluate the patient preoperatively, perform the physical examination completely, and use imaging methods if possible. Necessary precautions should be taken by taking into consideration the possibility of difficult airway, problems encountered in regional anesthesia, and multisystem complications that may develop.

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