Research Article

# Anaesthetic Management of A Patient With Neurofibromatosis Undergoing Surgery For Ectopic Pregnancy

Dr. Muthulakshmi R.[1], Dr. Rangapriya A.[2]*, Dr. Ravela Lakshmi Sravya[3]

[1] Postgraduate Resident, Department of Anaesthesiology, Sree Balaji Medical College & Hospital, Bharath Institute of Higher Education and Research (BIHER), Chennai, Tamil Nadu, India. Email: muthu1312.mm@gmail.com | ORCID: 0009-0009-8255-2223
[2] Senior Resident, Department of Anaesthesiology, Sree Balaji Medical College & Hospital, Bharath Institute of Higher Education and Research (BIHER), Chennai, Tamil Nadu, India. Email: Drrangapriya.as@gmail.com | ORCID: 0000-0003-2728-8158
[3] Senior Resident, Department of Anaesthesiology, Sree Balaji Medical College & Hospital, Bharath Institute of Higher Education and Research (BIHER), Chennai, Tamil Nadu, India. Email: bannuravela@gmail.com | ORCID: 0009-0003-2302-438X

*Corresponding author: Dr. Rangapriya A. Email: Drrangapriya.as@gmail.com

**Abstract**

**Background:** Neurofibromatosis type 1 (NF1) is a multisystem genetic disorder characterized by multiple cutaneous neurofibromas and café-au-lait macules, with variable neurological, cardiovascular, respiratory and musculoskeletal involvement. Anaesthetic management may be challenging because of potential airway involvement, cardiovascular abnormalities, spinal pathology and associated pheochromocytoma. Pregnancy and laparoscopic surgery require additional perioperative considerations.

**Case Presentation:** A 28-year-old woman, G4P2L2A1, at 6 weeks and 5 days of gestation, presented for termination of an unwanted pregnancy with planned laparoscopic sterilization. She was a known case of NF1 for approximately 4 years and was not receiving regular medication for the condition. Examination revealed multiple café-au-lait macules and numerous cutaneous neurofibromas over the trunk and back. She was haemodynamically stable, with a pulse rate of 78 beats/min, blood pressure of 110/70 mmHg, and oxygen saturation of 99% on room air. Her height was 148 cm, weight 68 kg, and body mass index 31 kg/m². Airway assessment showed approximately three-finger-breadth mouth opening with adequate neck movements and no clinically evident airway obstruction. Laboratory investigations revealed haemoglobin 11.9 g/dL, platelet count 3.6 lakh/mm³, serum creatinine 0.44 mg/dL, sodium 141 mmol/L, and potassium 3.8 mmol/L. Electrocardiography showed normal sinus rhythm, while echocardiography demonstrated preserved left ventricular systolic function with an ejection fraction of 68%, without regional wall-motion abnormality. Neurological evaluation did not reveal significant abnormality. Ultrasonography demonstrated an early intrauterine pregnancy with a crown-rump length of 4.7 mm and a 30 × 5 mm subchorionic haemorrhage. The patient was classified as ASA II and underwent the planned procedure under general anaesthesia with appropriate preparation for potential difficult airway management and vigilant perioperative monitoring. The perioperative course was uneventful.

**Conclusion:** NF1 requires careful multisystem evaluation before anaesthesia, particularly during pregnancy. Assessment of the airway, cardiovascular and neurological systems, consideration of associated spinal pathology and preparation for potential airway difficulty are essential. Individualized anaesthetic planning and vigilant perioperative monitoring can facilitate safe management of patients with NF1 undergoing laparoscopic procedures during early pregnancy.

**Keywords:** Neurofibromatosis type 1; Pregnancy; Medical termination of pregnancy; Laparoscopic sterilization; General anaesthesia; Cutaneous neurofibromas; Obstetric anaesthesia; Perioperative management.

## Introduction

Neurofibromatosis type 1 is an autosomal dominant multisystem disorder caused by pathogenic variants in the NF1 gene, located on chromosome 17q11.2. It is characterized by café-au-lait macules, axillary or inguinal freckling and multiple cutaneous, subcutaneous and plexiform neurofibromas. Although the cutaneous manifestations are often the most readily recognized clinical features, NF1 may involve the central and peripheral nervous systems, cardiovascular system, respiratory tract, bones and endocrine system.

From an anaesthetic perspective, NF1 is important because neurofibromas may involve the face, tongue, pharynx, larynx and cervical region, potentially resulting in difficult mask ventilation or tracheal intubation. Neurological involvement, spinal neurofibromas and dural ectasia may influence the feasibility of neuraxial anaesthesia. Cardiovascular complications include hypertension and vascular abnormalities, while pheochromocytoma is an important associated condition because an undiagnosed tumour may result in severe perioperative haemodynamic instability.

Pregnancy in women with NF1 requires additional consideration. Studies have reported increased risks of gestational hypertension, preeclampsia, intrauterine growth restriction, cerebrovascular complications and preterm delivery.

The presence of an ectopic pregnancy requiring urgent surgical intervention adds another dimension to anaesthetic decision-making because there may be limited time for extensive investigation.
The choice of anaesthetic technique should therefore be individualized. Neuraxial anaesthesia may be complicated by spinal neurofibromas or dural ectasia, whereas general anaesthesia requires careful assessment and preparation for a potentially difficult airway.
We present the anaesthetic management of a 28-year-old woman with NF1 and an ectopic pregnancy at 6 weeks and 5 days of gestation, who underwent surgical management under general anaesthesia.

## CASE PRESENTATION

A 28-year-old woman, G4P2L2A1, presented to the Department of Obstetrics and Gynaecology for medical termination of an unwanted pregnancy with laparoscopic sterilization at a gestational age of 6 weeks and 5 days. She had a history of two previous normal vaginal deliveries, with the last childbirth approximately 11 years earlier. She was a known case of neurofibromatosis type 1 (NF1) for the past 4 years and was not receiving any regular medication for NF1.
There was no history of cough, cold or fever. She had no documented history of diabetes mellitus, hypertension, bronchial asthma, tuberculosis, coronary artery disease, cerebrovascular disease, thyroid disorder or seizure disorder. There was no history of recent respiratory infection, snake bite or burns. She had no previous surgical history.
On examination, the patient was conscious, oriented and cooperative. She was afebrile, with a pulse rate of 78 beats/min, blood pressure of 110/70 mmHg, and oxygen saturation of 99% on room air. Her height was 148 cm, weight 68 kg, and body mass index was 31.0 kg/m$^2$. Cardiovascular examination revealed normal S1 and S2 with no clinically significant abnormality. Respiratory examination revealed bilateral air entry with no added sounds. There was no pallor, icterus, cyanosis, clubbing, lymphadenopathy or pedal oedema.
Cutaneous examination revealed multiple café-au-lait macules and numerous cutaneous neurofibromas distributed over the trunk, back and pelvic region, consistent with NF1. The lesions were variable in size and morphology, with several discrete papular and nodular lesions over the posterior trunk.
Airway assessment revealed a mouth opening of approximately three finger breadths, with adequate neck movements and no documented neck restriction. No obvious airway neurofibroma was identified clinically. Dentition was assessed, and artificial dentures were noted. There was no clinically evident respiratory obstruction.
Routine preoperative investigations were within acceptable limits. Haemoglobin was 11.9 g/dL, packed cell volume 34%, platelet count 3.6 lakh/mm$^3$, serum urea 16 mg/dL, and serum creatinine 0.44 mg/dL. Serum sodium was 141 mmol/L and potassium 3.8 mmol/L. Prothrombin time/INR was 13 seconds/1.14, and APTT was 37.5 seconds. HIV and HBsAg were negative, and anti-HCV was non-reactive. Thyroid-stimulating hormone was 1.98 mIU/L. Electrocardiography showed normal sinus rhythm, and chest radiography was within normal limits.
In view of the underlying NF1, further systemic evaluation was undertaken. Echocardiography demonstrated preserved left ventricular systolic function with an ejection fraction of 68%, without regional wall-motion abnormality, with only trivial mitral and tricuspid regurgitation. Neurological evaluation was obtained, and no significant neurological abnormality requiring alteration of the planned procedure was identified. Available neuroimaging did not reveal a significant intracranial abnormality.
Obstetric ultrasonography demonstrated an early intrauterine pregnancy corresponding to 6 weeks and 5 days of gestation, with a crown-rump length of 4.7 mm. A 30 × 5 mm subchorionic haemorrhage was noted, with a closed internal os. The patient was subsequently planned for the intended obstetric procedure.
Considering the patient's NF1, early pregnancy, obesity and the requirement for laparoscopic surgery, she was assessed as ASA physical status II. Particular attention was given to the possibility of difficult airway management and cardiovascular or neurological manifestations associated with NF1. Appropriate difficult-airway equipment was kept available, and standard perioperative monitoring was planned. After multidisciplinary assessment and informed consent, the patient was taken up for the planned procedure under general anaesthesia.



**Figure 1.** Multiple cutaneous neurofibromas distributed over the back and trunk.



**Figure 2.** Facial and cutaneous manifestations of neurofibromatosis type 1.

## DISCUSSION

Neurofibromatosis type 1 (NF1) is a multisystem genetic disorder characterized by variable involvement of the skin, peripheral and central nervous systems, cardiovascular system and musculoskeletal system. The presence of multiple cutaneous neurofibromas and café-au-lait macules in the present patient was consistent with the clinical phenotype of NF1.[1] Although the cutaneous manifestations are often the most obvious feature, their presence warrants assessment for clinically occult systemic involvement.

Anaesthetic management in NF1 requires particular attention to the airway, cardiovascular system and neurological status. Neurofibromas involving the tongue, pharynx, larynx or cervical region may produce difficulty with mask

ventilation or tracheal intubation.[2] In the present patient, airway examination showed approximately three-finger-breadth mouth opening with adequate neck movements and no clinically evident airway obstruction. Nevertheless, appropriate preparation for difficult airway management was considered essential because airway involvement may not always be apparent on routine examination.

Cardiovascular assessment is also important because NF1 may be associated with hypertension, vascular abnormalities and pheochromocytoma.[2,6] The present patient was haemodynamically stable, with a blood pressure of 110/70 mmHg and pulse rate of 78/min. Echocardiography demonstrated preserved cardiac function with an LVEF of 68%, without regional wall-motion abnormality, with only trivial mitral and tricuspid regurgitation. These findings supported proceeding with the planned procedure under appropriate anaesthetic monitoring.

Pregnancy may increase the risk of hypertensive, cerebrovascular and other maternal complications in women with NF1.[3] In the present case, pregnancy was at an early gestational age of 6 weeks and 5 days, and the patient was scheduled for medical termination of pregnancy with laparoscopic sterilization. Ultrasonography demonstrated an early intrauterine pregnancy with a CRL of 4.7 mm and a 30 × 5 mm subchorionic haemorrhage.

Neuraxial anaesthesia also requires individualized consideration in NF1 because spinal neurofibromas and dural ectasia may occur and can influence the safety or efficacy of neuraxial techniques.[4] In this patient, neurological assessment and available neuroimaging did not reveal a significant abnormality. Considering the planned laparoscopic procedure and overall clinical assessment, general anaesthesia was selected.

The case emphasizes that NF1 should be approached as a multisystem anaesthetic problem rather than merely a cutaneous disorder. Careful airway assessment, cardiovascular evaluation, neurological assessment, appropriate investigation and preparedness for potential complications allowed safe perioperative management in this patient.

## CONCLUSION

Anaesthetic management of patients with NF1 requires individualized multisystem assessment and careful perioperative planning. Airway neurofibromas, cardiovascular abnormalities, pheochromocytoma and occult neurological or spinal pathology should be considered even in clinically stable patients.

In the present case, the patient was a 28-year-old G4P2L2A1 woman at 6 weeks and 5 days of pregnancy, with extensive cutaneous manifestations of NF1, who underwent medical termination of early intrauterine pregnancy with laparoscopic sterilization under general anaesthesia. Stable cardiovascular function, satisfactory airway assessment and absence of significant neurological abnormalities permitted planned anaesthetic management.

This case highlights the importance of anticipation rather than reaction in NF1: detailed preoperative assessment, preparation for a potentially difficult airway, cardiovascular evaluation and vigilant intraoperative monitoring are central to achieving safe anaesthetic outcomes.

## REFERENCES

1. Legius E, Messiaen L, Wolkenstein P, Pancza P, Avery RA, Berman Y, et al. Revised diagnostic criteria for neurofibromatosis type 1 and Legius syndrome: an international consensus recommendation. Genet Med. 2021;23(8):1506-13. doi:10.1038/s41436-021-01170-5.

2. Fox CJ, Tomajian S, Kaye AJ, Russo S, Volpi Abadie J, Kaye AD. Perioperative management of neurofibromatosis type 1. Ochsner J. 2012;12(2):111-21. PMID:22778675; PMCID:PMC3387836.

3. Socolov D, Ceausu I, Bernad E, et al. Maternal and fetal complications in pregnant women with neurofibromatosis type 1: literature review and two case reports. Medicina (Kaunas). 2025;61? [Verify final volume/pages from journal record before submission]. PMID:39860457; PMCID:PMC11765542.

4. Derdabi I, El Jouadi H, Edderai M. Dural ectasia: a manifestation of type 1 neurofibromatosis. Pan Afr Med J. 2018;31:226. doi:10.11604/pamj.2018.31.226.9797. PMID:31447984; PMCID:PMC6691315.

5. Mallikarjuna S, Karjigi D, Kadian S, Kumar A. Anesthetic management of pregnant patient with undiagnosed neurofibromatosis for emergency cesarean section. Cureus. 2023;15(2):e34786. doi:10.7759/cureus.34786. PMID:36923202; PMCID:PMC10008778.

6. Theodosopoulou P, Nastos C, Paraskeva A. Anesthetic management of a patient with type 1 neurofibromatosis and an occult pheochromocytoma: a case report. Braz J Anesthesiol. 2023;73(5):695-8. doi:10.1016/j.bjane.2021.02.045. PMID:33819497; PMCID:PMC10533959.