Case Report

DOI: 10.7860/JCDR/2026/88221.24410

Anaesthesia Section

Perioperative Anaesthetic Considerations in Neurofibromatosis Type 1 with Periampullary Neuroendocrine Tumour Undergoing Whipple's Procedure: A Case Report

ABHIGNA GUMMALLA[1], VIVEK CHAKOLE[2], BHAGYESH SAPKALE[3]

ABSTRACT

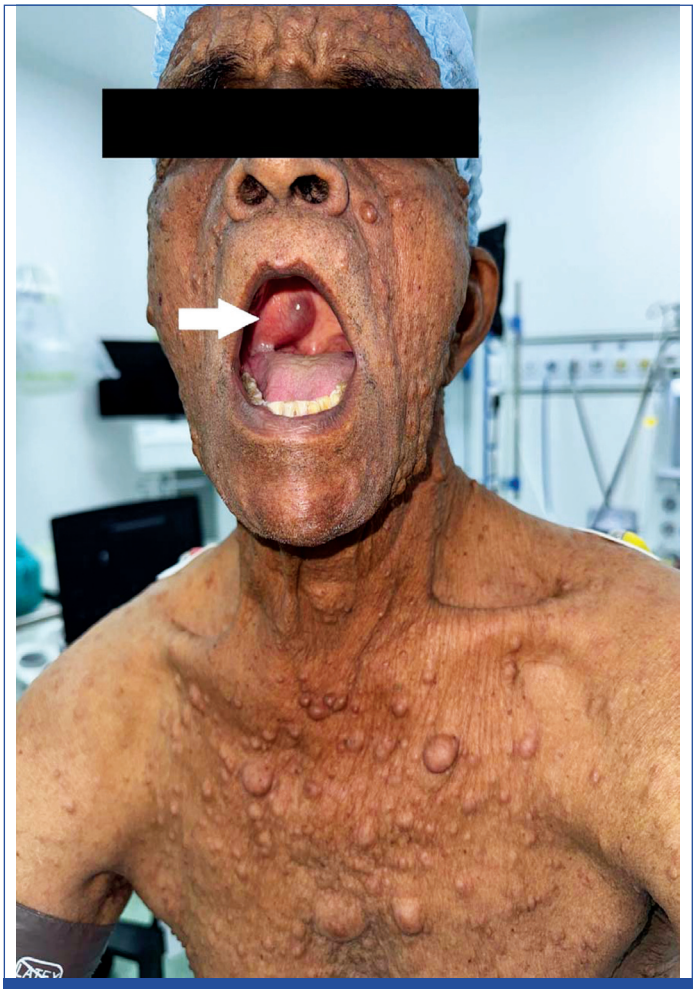
Neurofibromatosis type 1 (NF1) is an autosomal dominant multisystem disorder which is associated with benign and malignant tumours, skeletal deformities, airway lesions as well as endocrine abnormalities such as phaeochromocytoma posing significant anaesthetic challenges. Gastrointestinal involvement usually manifesting as periampullary Neuroendocrine Tumours (NETs), although uncommon may also require major surgical intervention. The present case report describes a 65-year-old male having known NF1 diagnosed with a well-differentiated periampullary NET scheduled for elective pancreaticoduodenectomy. Preoperative evaluation further showed multiple cutaneous, intraoral neurofibromas without any clinical evidence of difficult airway or carcinoid syndrome. Cardiovascular assessment was also unremarkable as well as biochemical parameters were within acceptable limits. Anticipating potential airway and haemodynamic concerns, invasive arterial and central venous monitoring were instituted and combined general anaesthesia along with epidural analgesia was administered. The intraoperative course of the patient remained haemodynamically stable also recovery was uneventful. The case highlights the importance of meticulous preoperative assessment, vigilance for occult endocrine pathology as well as individualised perioperative planning to ensure safe anaesthetic management in NF1 patients who are undergoing major abdominal surgery.

Keywords: Airway management, Epidural analgesia, Multisystem disorder, Pancreaticoduodenectomy, Phaeochromocytoma

CASE REPORT

The 65-year-old male presented to the Department of Surgery having urinary complaints inclusive of increased frequency, burning micturition, nocturia as well as difficulty in passing urine for three months. He was a known case of NF-1 diagnosed in childhood. During evaluation for symptoms of three months duration, he was found to have a periampullary lesion and was scheduled for elective pancreaticoduodenectomy (Whipple's procedure). The patient had no history of nausea or vomiting, bleeding of gastrointestinal tract, abdominal distension, fever, loss of consciousness, seizures, injury to head, bleeding of Ear, Nose and Throat (ENT), incontinence of bowel or bladder. There was also no history of diabetes mellitus, hypertension, coronary artery disease, respiratory illness as well as previous anaesthetic exposure. The patient had no significant past surgical history also no known drug allergies. The patient denied dyspnoea, dysphagia, change in voice. No symptoms suggestive of carcinoid syndrome such as flushing, diarrhoea, bronchospasm, or cardiac involvement were present.

Preanaesthetic evaluation demonstrated a moderately built, nourished male with height of 167 cm, weight of 53 kg, Body Mass Index (BMI) of 19 kg/m^2 as well as Body Surface Area (BSA) of 1.57 m^2. On general examination of patient, he was conscious, oriented, and cooperative with a Glasgow Coma Scale (GCS) score of E4V5M6 (15/15). Baseline vital parameters were also stable having a pulse rate of 84/min, blood pressure of 130/90 mmHg, respiratory rate of 16/min, afebrile status and oxygen saturation of 100% on room air. Airway assessment of the patient showed a mouth opening of three finger breadths, Mallampati class II, adequate thyromental distance, and unrestricted neck movements. Multiple neurofibromatous lesions were noted over soft palate on oral cavity examination [Table/Fig-1] thereby raising possibility of a difficult airway, airway assessment revealed a Modified Mallampati Class II although no clinical predictors of difficult mask ventilation or intubation were evident.

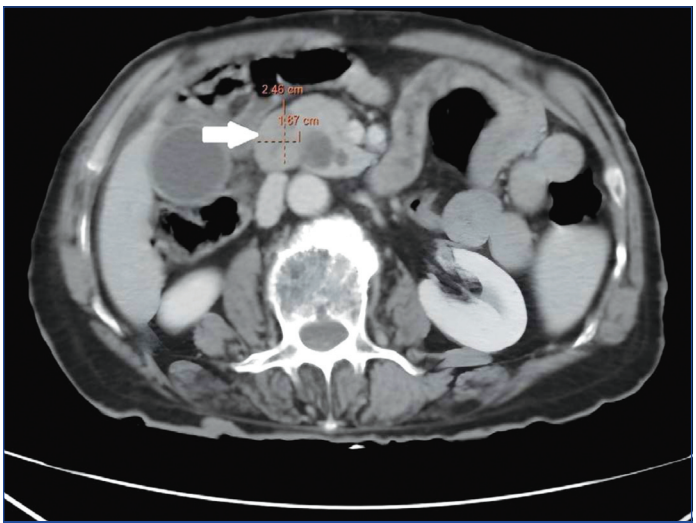


[Table/Fig-1]: Neurofibromatous lesions involving the soft palate as shown with white arrow.

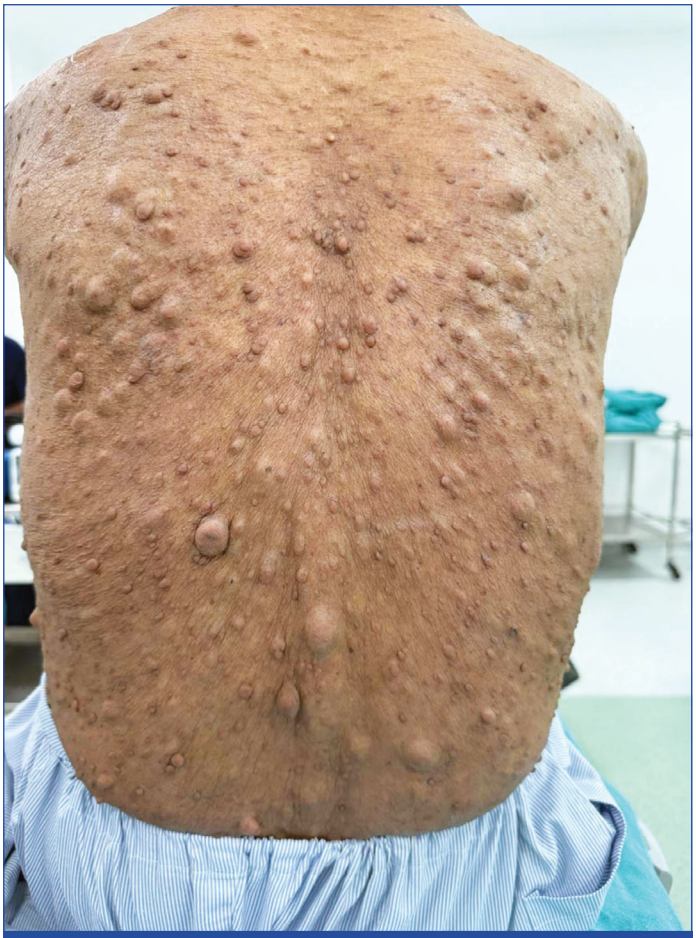
Systemic examination was unremarkable. Cardiovascular examination also revealed normal heart sounds (S1, S2), no presence of murmurs. Respiratory examination showed bilaterally equal air entry without any added sounds. Examination of Central Nervous System (CNS) also revealed no focal neurological deficits. Abdominal examination was soft, non-tender with no guarding or rigidity or distension as well as normal bowel sounds were present in all four quadrants.

Preoperative laboratory investigations were within normal limits. Liver function tests of the patient showed mildly elevated alkaline phosphatase (328 IU/L) as well as transaminases with a total bilirubin of 1.8 mg/dL. Viral markers {such as Hepatitis B surface Antigen (HBsAg) and Hepatitis C virus (HCV)} were also non reactive. Urine routine examination was within normal limits. Contrast-Enhanced CT (CECT) abdomen showed periampullary soft-tissue lesion measuring approximately 24×18 mm in size along with dilatation of the main pancreatic duct, common bile duct, and bilateral intrahepatic biliary radicles, suggestive of periampullary carcinoma as depicted in [Table/Fig-2]. The final diagnosis of a well-differentiated NET of the ampulla of pancreas was confirmed on histopathological examination of the resected specimen. However, tumour grading and Ki-67 proliferation index were not available. No significant lymphadenopathy was noted in the patient. Multiple subcutaneous soft-tissue lesions suggestive of neurofibromas were also observed as shown in [Table/Fig-3]. CT thorax was done which ruled out pulmonary metastases. Two-dimensional echocardiography (2D-echo) demonstrated good left ventricular systolic function with an ejection fraction of 60%, concentric left ventricular hypertrophy as well as grade-I diastolic dysfunction without any valvular abnormalities, pulmonary hypertension. Based on these findings he was assessed as American Society of Anaesthesiologists (ASA) physical status II.

After taking appropriate informed consent of the patient, he was taken to the operation theatre where standard monitors inclusive of Electrocardiogram (ECG), non invasive blood pressure, pulse oximetry and capnography were attached. Peripheral intravenous access was secured using an 18G cannula. Under proper aseptic precautions, invasive arterial monitoring was established adequately through the left radial artery using a blind technique. A 7-French central venous catheter was secured in right internal jugular vein under ultrasound guidance. An epidural catheter was placed at the T7-T8 intervertebral level under aseptic precautions. Correct placement was confirmed using a test dose of 3 mL lignocaine with adrenaline. No epidural-related complications were observed. Premedication of patient included usage of i.v. glycopyrrolate 0.2 mg, midazolam 0.04 mg/kg also fentanyl 2 µg/kg. Glycopyrrolate was administered as the patient had a normal baseline heart rate and no clinical evidence of catecholamine-secreting tumour.



[Table/Fig-2]: CECT abdomen showing periampullary lesion (white arrow) with biliary and pancreatic duct dilatation.



[Table/Fig-3]: Clinical photograph showing multiple cutaneous neurofibromas.

General anaesthesia was further induced with i.v. propofol 2 mg/kg then neuromuscular blockade using vecuronium 0.1 mg/kg. The trachea was intubated with the help of direct laryngoscopy with an 8.0 mm cuffed endotracheal tube, correct placement was confirmed by performing bilateral chest auscultation, capnography. Anaesthesia was maintained using combination of intravenous and sevoflurane also intermittent epidural top-ups of 0.25% bupivacaine (10 mL) for intraoperative analgesia, haemodynamic stability. The intraoperative blood loss was estimated approximately 900 mL. Urine output 1000 mL along with insensible losses were estimated at 1400 mL. A total of 3000 mL of crystalloids (Ringer's lactate and normal saline) as well as one unit of packed red blood cells (350 mL) were administered to the patient. No vasopressor or inotropic support was required during procedure.

The intraoperative course was uneventful, and haemodynamic parameters remained stable throughout the procedure. At the time of completion of surgery, neuromuscular blockade was reversed using i.v. neostigmine 0.05 mg/kg (2.65 mg), glycopyrrolate 0.01 mg/kg (0.5 mg) also patient was successfully extubated after fulfilling standard extubation criteria. He was shifted to postoperative care unit in a stable condition having adequate analgesia and stable vital parameters.

DISCUSSION

An autosomal dominant neurocutaneous disorder known as NF1 caused due to mutations in NF1 gene which further encodes neurofibromin (tumour suppressor protein) that negatively regulates Ras/MAPK signalling pathway [1]. The estimated incidence of NF1 is approximately 1 in 2,500-3,000 live births [1]. NF1 is characterised having café-au-lait macules, axillary or inguinal freckling, cutaneous-plexiform neurofibromas, Lisch nodules as well as an increased predisposition to both benign and malignant neoplasms [2]. NF1 is also associated having multisystem involvement which is inclusive of central and peripheral nervous system tumours, skeletal deformities,

Author	Patient profile	Surgery/Procedure	Anaesthetic issue encountered	Key findings	Management	Clinical implication
Theodosopoulou P et al., [5]	42-year-old female with known NF1	Open myomectomy	Severe refractory hypertension during induction (BP 220/120 mmHg)	Elevated plasma metanephrines, normetanephrines; 2 cm left adrenal mass	Surgery aborted; α-blockade (phenoxybenzamine) followed by β-blockade; subsequent adrenalectomy	Importance of screening for occult phaeochromocytoma in NF1 to prevent peri-induction hypertensive crisis
Wang J et al., [8]	44-year-old male with longstanding NF1	Glioma resection	Hypertensive crisis during induction (BP 310/140 mmHg)	Elevated catecholamines; large adrenal mass on CT	Phentolamine, esmolol, remifentanil; later adrenalectomy	Strong association of NF1 with occult phaeochromocytoma; mandates thorough endocrine evaluation
Aman A et al., [9]	Late 30s male with NF1 and kyphoscoliosis	Open reduction of hip dislocation	Inadequate, asymmetric spinal block	MRI: scoliosis, dysmorphic vertebrae, dural ectasia	Ultrasound-guided thoracic epidural anaesthesia	Neuraxial anaesthesia may be unpredictable in NF1 with spinal deformities; ultrasound improves safety
Taka H et al., [10]	34-year-old parturient with NF1 and preeclampsia	Repeat caesarean section	Prior failed spinal anaesthesia	MRI: dural ectasia, radicular cyst	Combined Spinal-Epidural Anaesthesia (CSEA) with epidural supplementation	Preoperative spinal imaging valuable; CSEA offers titratable alternative in dural ectasia
Mallikarjuna S et al., [11]	26-year-old G5P4L4, undiagnosed NF1	Emergency caesarean section	Avoidance of neuraxial anaesthesia due to suspected occult CNS lesions	Clinical diagnosis of NF1 with cutaneous and intraoral neurofibromas	General anaesthesia with rapid sequence induction	Emergency settings require caution due to possible occult spinal/ intracranial tumours
Present case	65-year-old male with known NF1 and periampullary NET	Elective pancreaticoduodenectomy (Whipple's procedure)	Anticipated airway and haemodynamic concerns	Multiple cutaneous and intraoral neurofibromas; stable endocrine profile	Invasive arterial and central venous monitoring; general anaesthesia with epidural analgesia	Importance of meticulous preoperative assessment, endocrine screening, and individualised anaesthetic planning in major abdominal surgery

[Table/Fig-4]: Reported anaesthetic challenges in NF1 patients including the present case; made by authors [5,8-11].

cardiovascular abnormalities, endocrine tumours such as phaeochromocytoma [3]. Importantly, gastrointestinal manifestations are also increasingly recognised usually as NETs most commonly involving periampullary, duodenal regions [3]. These tumours are often well-differentiated which can present along with obstructive jaundice, non specific gastrointestinal symptoms [3]. From an anaesthetic perspective, NF1 also poses unique perioperative challenges. Airway difficulties can arise due to oropharyngeal, laryngeal neurofibromas while spinal deformities as well as dural ectasia can further complicate neuraxial techniques [4]. The risk of occult phaeochromocytoma needs proper preoperative evaluation to prevent catastrophic peri-induction hypertensive crises in patients [5,6]. The co-existence of NF1 along with a periampullary NET which requires major surgery such as pancreaticoduodenectomy further compounds into complexity of perioperative management [4,7]. Given the rarity of such association as well as potential for multisystem involvement influencing anaesthetic planning, reporting such cases contributes valuable insights into tailored perioperative strategies, risk mitigation in this unique type of patient population [6,7]. In the present case, a 65-year-old male with NF1 undergoing pancreaticoduodenectomy did not exhibit features of carcinoid syndrome or phaeochromocytoma. Airway examination showed intraoral neurofibromas raising concern for potential difficult airway, although intubation was achieved without difficulty. The perioperative course also remained haemodynamically stable without hypertensive crises or anaesthetic complications.

Theodosopoulou P et al., and Wang J et al., reported cases of severe peri-induction hypertensive crises in patients with NF1 due to undiagnosed phaeochromocytoma. In contrast to these reports, present patient did not demonstrate any perioperative haemodynamic instability or features suggestive of occult phaeochromocytoma [5,8]. Aman A et al., and Taka H et al., highlighted challenges with neuraxial anaesthesia in NF1 patients due to spinal deformities and dural ectasia, leading to unpredictable or inadequate blocks. In contrast, present patient had no spinal deformities or prior history of neuraxial block failure, and epidural anaesthesia was successfully administered without difficulty [9,10]. Mallikarjuna S et al., reported the use of general anaesthesia in an undiagnosed NF1 patient due to concerns regarding possible occult spinal or intracranial involvement [11].

The NF1 patients present multiple anaesthetic challenges including potential difficult airway due to upper airway neurofibromas, risk of airway obstruction, and difficulty in mask ventilation or intubation [4,11]. Neuraxial anaesthesia may be unpredictable due to spinal deformities, scoliosis, or dural ectasia leading to inadequate or asymmetric block [9,10]. Additionally, the presence of occult phaeochromocytoma poses a significant risk of severe perioperative haemodynamic instability [5,8]. Associated restrictive lung disease, skeletal abnormalities, and possible central nervous system involvement further complicate perioperative management [3,4]. These factors necessitate careful preoperative evaluation and individualised anaesthetic planning. Alternative anaesthetic strategies such as sole regional anaesthesia or combined spinal-epidural techniques have been described in NF1 patients [9,10]. However, in the present case, general anaesthesia with epidural analgesia was preferred considering the nature of major abdominal surgery, absence of spinal abnormalities, and need for optimal intraoperative haemodynamic control and postoperative analgesia. Reported anaesthetic challenges in NF1 patients including the present case are depicted in [Table/Fig-4] [5,8-11].

CONCLUSION(S)

The NF1 poses significant perioperative challenges due to its multisystem involvement, association with occult endocrine, airway abnormalities. The co-existence of NF1 with a periampullary NET requiring pancreaticoduodenectomy further increases anaesthetic complexity. Thorough preoperative evaluation usually for airway involvement and undiagnosed phaeochromocytoma is essential for preventing catastrophic intraoperative events. Careful planning of invasive monitoring, analgesic strategies, haemodynamic control ensures favourable outcomes. The present case highlights the importance of individualised anaesthetic management, multidisciplinary coordination in optimisation of perioperative safety in NF1 patients undergoing major surgery of abdomen.

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PARTICULARS OF CONTRIBUTORS:

1. Junior Resident, Department of Anaesthesia, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
2. Professor and Head, Department of Anaesthesia, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
3. Undergraduate Student, Department of Medicine, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Abhigna Gummalla,
Junior Resident, Department of Anaesthesia, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha-442107, Maharashtra, India.
E-mail: abhigna.gummalla@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Mar 06, 2026
- Manual Googling: Jun 13, 2026
- iThenticate Software: Jun 15, 2026 (5%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Feb 12, 2026**
Date of Peer Review: **Mar 26, 2026**
Date of Acceptance: **Jun 17, 2026**
Date of Publishing: **Sep 01, 2026**