Regional Anesthetic Technique for a Patient with Neurofibromatosis Type 1 and an Unusual Appearance of the Lower Extremity Nerves

Suvar T* and Mehaffey
Department of Anesthesiology, University of Arkansas for Medical Sciences, USA

*Corresponding author: Suvar T, University of Arkansas for Medical Sciences, USA

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Abstract

Neurofibromatosis type 1 (NF1) is predominantly a neurocutaneous disorder, but can affect different organ systems; numerous considerations must be made in a NF1 patient who will undergo anesthesia. We report a case of a soft tissue biopsy in a 27-year-old Caucasian male with a history of NF1 complicated by multiple cervical spinal fusion surgeries with significant airway concerns. The decision was made to proceed with a regional anesthetic technique with moderate sedation.

Objective: This case demonstrates challenges associated with regional anesthesia and demonstrates safe anesthetic techniques for a surgical procedure in an NF1 patient.

Case Report: A 27-year-old patient with neurofibromatosis 1 had developed a rapidly growing mass in his right foot that required biopsy. History of multiple cranio-cervical-thoracic fusions, scoliosis, and limited neck extension raised concerns for a difficult airway.

Conclusion: The procedure was successfully managed with regional anesthesia and moderate sedation, with no subsequent complications.

Introduction

Neurofibromatosis type 1 (NF1) is predominantly a neurocutaneous disorder, but can affect different organ systems; numerous considerations must be made in a NF1 patient who will undergo anesthesia. We report a case of a soft tissue biopsy in a 27-year-old Caucasian male with a history of NF1 complicated by multiple cervical spinal fusion surgeries with significant airway concerns. The decision was made to proceed with a regional anesthetic technique with moderate sedation.

Case Presentation

A 27-year-old male with a past medical history of neurofibromatosis type 1 presented to the clinic for a 3-month history of a rapidly growing mass located on the dorsum of his right foot. MRI was performed and revealed a 3.8 x 4 x 3.8 mass, appearing to involve the 2nd and 3rd metatarsals. Surgical biopsy of the mass was planned. On pre-operative assessment, patient had a Mallampati score of 2, but has had numerous cranio-cervical thoracic spine fusions due to neuromuscular scoliosis, with significantly limited neck extension rendering him a difficult endotracheal intubation. The decision was made to proceed with regional anesthesia combined with intravenous sedation. The patient was pre-medicated with 2mg of midazolam. We attempted a sciatic popliteal and adductor canal nerve blocks under ultrasound guidance. The procedure was successfully managed with regional anesthesia and moderate sedation.

Discussion

Neurofibromatosis type 1 (NF1) is an autosomal dominant neurocutaneous condition caused by an NF1 gene mutation, resulting in decreased activity or nonfunctional neurofibronin protein. It affects 1 in 2600 to 3000 individuals. Clinical manifestations include café-au-lait spots, axillary freckling, Lisch nodules, optic gliomas, and neurofibromas. Neurofibromas are the most common tumor found in NF1 patients. They can be cutaneous, plexiform, or nodular, and they can undergo malignant transformation with a risk of 5-13% [1].

NF1 may involve multiple organ systems, and therefore raise anesthetic concerns during surgical procedures. Neurofibromas may be present in the airway which can make intubation difficult [2].

Lovell et al. reports a case of cervical vertebrae dislocation that was discovered post surgery in an asymptomatic NF1 patient with multiple cervical neurofibromas. There must be consideration for a radiologic examination prior to anesthesia for NF1 patients with a history of cervical surgery to assess for any unknown cervical dislocations [3]. Vertebral deformities, like scoliosis present in our...
patient, or neurofibromas in the spinal cord may also complicate spinal or extradural blocks. Other considerations include undiagnosed neurofibromas in the lungs and GI tract, hypertension, renal artery stenosis, and pheochromocytoma [2].

Shahid and Sebastian [4], report a case of lower limb wound debridement in a 66-year-old NF1 patient successfully managed with femoral and sciatic nerve block. Another case was reported by Bagam et al [5], of a 60-year-old female with NF1 who needed dynamic hip screw fixation of left femur and intramedullary interlocking nail for the humerus. The surgical procedures were managed with a subarachnoid block and a brachial plexus block, respectively. Given this patient’s extensive history of multiple cranio-cervical-thoracic fusions surgeries, NF1 complicated by scoliosis, and neurofibromas extensively found throughout his body, a decision was made to proceed with a regional block as demonstrated by various case reports to be the safest anesthetic option. Our ultrasound scan of the popliteal fossa demonstrated multiple hypoechoic structures within the popliteal nerve. The common peroneal and tibial nerve branching was not easily identified as they should have been seen converging from the popliteal nerve. As seen in Figure 1, there were concerns for having adequate local anesthetic infiltration of the endoneurium as well as considerations for nerve damage and insult to the neuromas with an inadvertent puncture of the nerve by the 22G short-bevel needle. A more caudal approach was utilized to block the right superficial and deep peroneal nerves, and tibial nerve as seen in (Figure 2).

Numerous challenges are faced when administering anesthetics to patients with NF1; significant airway concerns in our patient lead to a regional technique with mild sedation as a safer alternative to general anesthesia for lower extremity surgery. While there were concerns for performing regional blocks at the saphenous and popliteal level, a more distal approach can be used in the presence of neurofibromas in major peripheral nerves.

Written Consent Statement

The authors of this case report here by state that the involved patient provided written permission for the report to be published.

References