Brazilian Journal of Anesthesiology 2022;72(6):826–828

Sociedade Brasileira de Anestesiologia

Brazilian Journal of ANESTHESIOLOGY

CASE REPORT

Neuraxial block anesthetic technique in a patient with SCN8A encephalopathy: case report

Eric Guimarães Machado [a,*], **Isis da Rocha Costa Billé** [a], **Mariana Moraes Pereira das Neves Araújo** [b,c], **José Francisco Nunes Pereira das Neves** [b], **Gilson Lorena Maués** [b], **Marco Felipe Bouzada Marcos** [d], **Fernando de Paiva Araújo** [b,c]

[a] *Santa Casa de Misericórdia de Juiz de Fora, Serviço de Anestesiologia, Juiz de Fora, MG, Brazil*
[b] *Hospital Monte Sinai, Serviço de Anestesiologia, Juiz de Fora, MG, Brazil*
[c] *Hospital Universitário de Universidade Federal de Juiz de Fora (UFJF), Serviço de Anestesiologia, Juiz de Fora, MG, Brazil*
[d] *Hospital Escola Luiz Gioseffi Jannuzzi, Valença, RJ, Brazil*

Received 17 June 2020; accepted 26 August 2020
Available online 4 February 2021

KEYWORDS
Encefalopatias;
Raquianestesia;
Canais de Sódio

Abstract Mutations in SCN8A gene lead to changes in sodium channels in the brain, which are correlated with severe epileptic syndrome. Due to the rarity, there are few studies that support anesthesia in that population. The present study aims to report alternatives to inhalation anesthesia at epileptic encephalopathy.
Case report: Male, 4 years old, with SCN8A encephalopathy with surgical indication of orchidopexy. Neuroaxis block was performed and dexmedetomidine was used as a pre-anesthetic and sedation. The anestheticsurgical act was uneventful.
Conclusion: The association of neuraxial block and dexmedetomidine proved to be a viable alternative for surgery in patients with SCN8A encephalopathy.

Introduction

Mutations on gene SCN8A lead to changes in voltage-dependent sodium channels in the brain that have recently been associated with severe epileptic syndrome, although a well-defined clinical presentation is still being investigated. SCN8A encephalopathy is a rare syndrome with

* Corresponding author.
E-mail: ericgmac@hotmail.com (E.G. Machado).

https://doi.org/10.1016/j.bjane.2020.08.013

onset in childhood, described by the presence of recurrent drug-resistant seizures, developmental delay, and frequent epileptiform activity on the Electroencephalogram (EEG).[1,2]

Given the disease is rare, there are no studies in the literature favoring a particular anesthesia management for these patients. According to Pal et al.,[3] mice with SCN8A gene mutations are more sensitive to sevoflurane and isoflurane, inhalation agents commonly used in pediatric anesthesia. Thus, the present case report aimed to describe alternatives to the use of inhalation anesthesia in this group of individuals.

Case report

Consent to report the case with educational purposes was authorized and signed by the family. The patient was a 4-year-old male, 16 kg, candidate for orchidopexy and circumcision. The patient presented SCN8A encephalopathy with major developmental and behavioral impairment and was being followed-up at Instituto Estadual do Cérebro Paulo Niemeyer (Rio de Janeiro, RJ) for 2 years, with the diagnosis confirmed by whole exome sequencing in 2014 (the only case confirmed of the syndrome in Brazil to date). The patient was on regular use of oxcarbamazepine 6% (600 mg.day^{-1}), sodium divalproate (375 mg.day^{-1}) and cannabidiol (144 mg.day^{-1}). Despite using anticonvulsant medication, the patient had countless daily episodes of seizures. Physical examination was unremarkable and complementary tests within normal values.

Dexmedetomidine (2 μg.Kg^{-1} by nasal route) was administered as preanesthetic medication, 30 minutes before entering the operating room. At that time, the patient presented a short-duration seizure episode that repeated during positioning on the operating table. In the operating room, the patient was monitored with electrocardiogram, pulse oximetry and noninvasive blood pressure. After venous cannulation with 22G catheter, hydration with sodium chloride solution 0.9% was started. Sedation was maintained with continuous intravenous infusion of dexmedetomidine (1.0 μg.Kg^{-1}.h^{-1}) and the patient received O_2 supplement by nasal catheter (3 L.min^{-1}). With the patient in the sitting position, spinal anesthesia was performed with a single puncture using a pediatric Quincke 25G type needle and administration of 10 mg of 0.5% hyperbaric bupivacaine.

The anesthetic-surgical procedure was uneventful. At the end, the patient was sent to the Postanesthetic Recovery Unit (PACU), placed under the same intraoperative monitoring and no new seizure episodes were observed. He was discharged from the PACU 120 minutes after admission, presenting score of 10 on the modified Aldrete scale. Hospital discharge was on the postoperative day 1.

Discussion

Epilepsy is a common pediatric neurological disorder, and drug-resistant epilepsies are present in 30% of cases. Such severe forms include epileptic encephalopathies, responsible for several sequelae and cognitive and behavioral impairment. SCN8A encephalopathy is a recently described epileptic encephalopathy, caused by mutations on the SCN8A gene that codifies the sodium channel Nav1.6.[1]

SCN8A encephalopathy was first identified in 2012 and only 140 cases have been diagnosed since. Most individuals affected present drug-resistant seizures and inconsistent response to conventional anticonvulsants.[1]

Although developmental delay may start from birth in a child presenting SCN8A encephalopathy, in many cases, development is normal before seizures begin. The most common clinical presentations are intellectual impairment (varying from mild, moderate to severe) and motor abnormalities, which include hypotonia, ataxia, dystonia, hyperreflexia, and choreoathetoses. (1) Severe cognitive, motor, and behavioral impairment can be observed, as the condition progresses. (2) Sudden death during seizure has been reported in five individuals. Most patients reported in the literature were in the first two decades of life.[1] In the case reported, the child presented motor delay and important cognitive impairment.

There are reports in the literature showing that mice with gene SCN8A mutations assessed by EEG monitoring were more sensitive to volatile anesthetics (sevoflurane and isoflurane). Despite not increasing emergence and awakening time, in vivo exposure resulted increased theta wave activity on EEG, which correlates with general anesthesia depth.[3] There are no reports in the literature to date describing the anesthetic management of patients with SCN8A encephalopathy. Thus, pursuing anesthetic alternatives for these patients is required. In the present report, we chose to use neuraxial block associated with dexmedetomidine as pre-anesthetic medication and intraoperative sedation.

In pediatrics, spinal anesthesia remains controversial as the first-choice technique and its use have been limited to specialized pediatric surgery centers. However, the literature provides a rationale concerning safety of the technique.[4] The main limitation of the technique is the limited duration of the blocks in children, that can be overcome by associating adjuvant drugs such as clonidine, morphine, epinephrine, neostigmine,[4] and even intravenous dexmedetomidine.[5]

In the present case, the drug used for spinal anesthesia was 0.5% hyperbaric bupivacaine. Bupivacaine is a local anesthetic and, as such, blocks the sodium channel at axons reversibly, preventing spread of stimulus. Despite the patientś syndrome presented altered sodium channels, no adverse reactions were observed by using neuraxial local anesthetic, and the child responded to the technique within what is expected in individuals without SCN8A syndrome.

Dexmedetomidine is an α-2 agonist class drug and is used in pediatric patients as pre-anesthetic medication for intraoperative sedation and as an adjuvant drug in postoperative pain management.[5] Although there are no studies to support using the drug both in SCN8A encephalopathy or in the pediatric population in general, in the present case report, its use provided satisfactory results and no adverse effects.

Conclusion

The association of neuraxial block and dexmedetomidine as pre-anesthetic medication and intraoperative sedation proved to be a feasible alternative for surgeries below the diaphragm for patients with SCN8A encephalopathy.

Although the mutation found in this syndrome is on sodium channel blockers, using a local anesthetic – such as, in this case, bupivacaine – yielded the expected response, like that in other individuals in the same age group. Plus, dexmedetomidine proved to be efficient in promoting sedation during the procedure.

Conflicts of interest

The authors declare no conflicts of interest.

References

1. Meisler MH, Helman G, Hammer MF, et al. SCN8A encephalopathy: Research progress and prospects. Epilepsia. 2016;57:1027–35.
2. Malcolmson J, Kleyner R, Tegay D, et al. SCN8A mutation in a child presentingwith seizures and developmental delays. Cold Spring Harb Mol Case Stud. 2016;2:a001073.
3. Pal D, Jones JM, Wisidagamage S, Meisler MH, Mashour GA. Reduced Nav1.6 Sodium channel activity in mice increases in vivo sensitivity to volatile anesthetics. PLoS One. 2015;10:e0134960.
4. Gupta A, Saha U. Spinal anesthesia in children: A review. J Anaesthesiol Clin Pharmacol. 2014;30:10–8.
5. Mahmoud M, Mason KP. Dexmedetomidine: review, update, and future considerations of paediatric perioperative and periprocedural applications and limitations. Br J Anaesth. 2015;115:171–82.