Indian Journal of Clinical Anaesthesia 2024;11(2):258–260

Content available at: https://www.ipinnovative.com/open-access-journals

Indian Journal of Clinical Anaesthesia

Journal homepage: www.ijca.in

Case Report

Devic's disease: A devil's trap for the Anesthesiologist: A case report

Roshin Binoy[1]*, **Bincy V Thomas**[1], **Sheela Verghese**[1]

[1]*Dept. of Anaesthesiology, Government Medical College, Kottayam, Kerala, India*

ARTICLE INFO

Article history:
Received 14-03-2024
Accepted 26-04-2024
Available online 03-06-2024

Keywords:
Neuromyelitis optica spectrum disorder
Chronic pain
Muscle relaxants
General anesthesia

ABSTRACT

Neuromyelitis optica spectrum of disorder (NMOSD) is a rare autoimmune inflammatory relapsing astrocytopathy due to immunoglobulin against aquaporin 4 (AQP4) receptor. It is characterized by demyelination of the spinal cord and optic nerve. The vulnerability of demyelinated neurons to local anesthetics and increased response to neuromuscular blocking agents make the choice of anesthesia challenging. The rarity of the disease has made the literature scarce, especially when it comes to those undergoing surgeries and their anesthetic implications. We report the case of a 54-year-old patient with NMOSD who underwent modified radical mastectomy for carcinoma breast under general anesthesia. The use of multimodal analgesia for pain management and the avoidance of muscle relaxants resulted in an uneventful perioperative period in this patient.

1. Introduction

Neuromyelitis optica spectrum disorder (NMOSD), otherwise known as Devic's disease, is an inflammatory autoimmune disease that affects the spinal cord, optic nerve, and brainstem. It is a rare disease with prevalence estimates that range from 1-4 per 100,000, with Asians and Africans being disproportionately affected when compared to people of other ethnic backgrounds.[1]

The ideal choice of anesthesia for these patients remains controversial. The usage of muscle relaxants is a dilemma as there are very few case reports supporting their use. The propensity of developing chronic pain in these patients underlines the need for effective perioperative pain management. There exists little literature on the anesthetic management of patients with NMOSD. The existing literature on regional and general anesthesia is mostly on cesarean delivery management.

Here we report the case of a 54-year-old lady with pre-existing NMOSD who underwent modified radical mastectomy for carcinoma breast under general anesthesia without muscle relaxation.

2. Case Description

A 54 years old lady weighing 65 kg with a height of 155cm, diagnosed with left-sided carcinoma breast stage yc T0N0M0, was posted for left-sided modified radical mastectomy. She had completed eight cycles of neoadjuvant chemotherapy with Adriamycin and cyclophosphamide. Her comorbidities included type two diabetes mellitus for the past fifteen years and hypothyroidism for the past ten years for which she had been on medications. She developed quadriparesis with tonic spasms on the right side of her body four years back and was diagnosed to have NMOSD. The diagnosis was supported by positive serum Aquaporin 4(APQ4) Immunoglobulin(IgG) and the longitudinal involvement of more than two spinal cord segments. She was treated with pulse doses of

* *Corresponding author*.
E-mail address: roshinb@gmail.com (R. Binoy).

https://doi.org/10.18231/j.ijca.2024.049

methylprednisolone and rituximab following which she became symptomatically better. She had two previous relapses, which were treated with steroids and rituximab. Her last relapse was two years back.

Preanesthetic evaluation was unremarkable except for reduced power of grade four in bilateral lower limbs. Preoperative investigations were normal except for raised thyroid stimulating hormone levels of 15.08 m IU/L and elevated glycosylated hemoglobin levels of 11.5%. The dose of thyroxine was adjusted and glycemic control was attained preoperatively by adjusting the insulin regimen. The patient and the family were counseled regarding the need for general anesthesia, and the possibility of disease relapse in the postoperative period, and written informed consent was obtained.

The plan of anesthesia was general anesthesia with supraglottic airway device I gel™ [Inter Surgicals Inc] without the use of muscle relaxants. The patient was wheeled into the operation theatre and the standard monitors were attached. An intravenous line was secured on the dorsum of the right wrist with an 18 gauge cannula and a balanced crystalloid solution was started. The patient was premedicated with intravenous midazolam 1.5 mg. After adequate preoxygenation, anesthesia was induced with intravenous propofol 150 mg, and the airway was secured with I gel™ size four. As a part of multimodal analgesia, the patient received the following drugs intravenously - fentanyl 120 mcg, preservative-free lidocaine 100mg, acetaminophen 1 gm, ketamine 40 mg, dexamethasone 8 mg and ultrasound-guided left sided serratus anterior plane block with 30mL 0.2% ropivacaine. Anesthesia was maintained with oxygen, nitrous oxide, and sevoflurane on pressure support ventilation ensuring a minimum alveolar concentration of one. The intraoperative period was uneventful and towards the end of the surgery, she received intravenous ondansetron 6 mg. I gel™ was removed when the patient was fully awake. Postoperatively, there were no fresh neurological deficits. She was shifted to the postoperative care unit (PACU) for monitoring and was shifted out when the PACU discharge criteria were met. She had an uneventful postoperative period. She has not reported any further relapses after surgery and is on regular follow-up.

3. Discussion

NMOSD is an autoimmune inflammatory disease affecting the aquaporin 4 (AQP4) receptors on astrocytes.[2] (Figure 1) It has a high female-to-male preponderance of 9:1 with a peak onset at 40 years of age. It is more prevalent among East Asians (3.5/100000) and Africans (10/100000) when compared to whites (1/100000).[3] Symptoms are due to the involvement of the optic nerve and spinal cord. It includes blurring of vision, loss of color vision, eye pain, weakness, numbness or loss of sensation, spasticity, and rarely symptoms like nausea and hiccup when it affects the area prostrema. The diagnostic criteria for NMOSD are given in Table 1. It has been described to have relapses that might last for months which come into remission with treatment. It has been found to have higher chances of relapse during peripartum and postpartum periods.

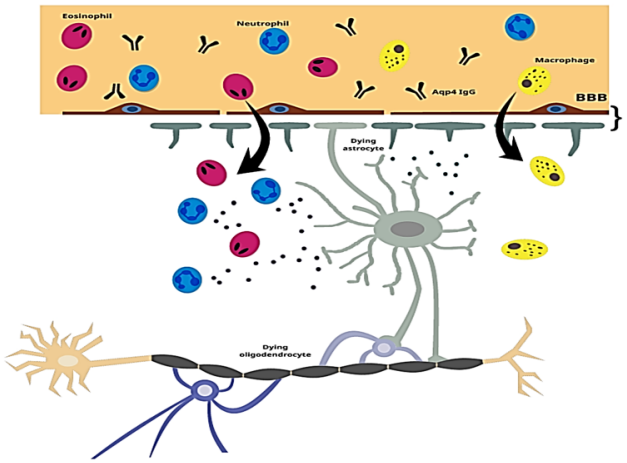

Figure 1: Pathogenesis of Neuromyelitis optica sprectrum of disorder

Increased susceptibility to neuromuscular blocking drugs leading to respiratory failure in the immediate postoperative period is a concern in these patients. There is a case report that documents incomplete recovery from neuromuscular blockade following general anesthesia for Caesarean section necessitating the requirement of postoperative bimodal positive airway pressure (BIPAP).[4] But S Dusitkasem reported that with adequate neuromuscular monitoring, muscle relaxants can be used without the risk of any post-operative pulmonary complications in a patient undergoing cesarean section under general anaesthesia.[5]

The demyelinated neurons are vulnerable to the action of local anesthetic agents. This makes central neuraxial blockade riskier in these patients. There are two case reports of exacerbation of NMSOD following spinal subarachnoid block.[6,7] The use of succinylcholine has been shown to cause hyperkalemia and hence has to be avoided.[8] These patients are susceptible to chronic pain, hence adequate pain relief should be provided with multimodal analgesia.[9]

Our concerns in the anesthetic management of this patient included avoiding post-operative pulmonary complications, preventing chronic pain from developing, and avoiding factors that cause a relapse of the disease. Hence we decided to avoid muscle relaxants and central neuraxial blockade for this patient. A multimodal analgesic regimen with serratus anterior plane block, ketamine, acetaminophen, lidocaine, fentanyl, and dexamethasone was used to manage acute pain and prevent the development of chronic pain. When considering the use of muscle

Table 1: Diagnostic criteria for Neuromyelitis optica spectrum of disorder (NMOSD)

	Diagnostic criteria for NMOSD1 with Aquaporin 4 Immunoglobulin (AQP4-IgG)
1	At least one core clinical characteristic (see below)
2	Positive test for AQP42-IgG3
3	Exclusion of alternative diagnoses
	Diagnostic criteria for NMOSD1 without AQP42-IgG3 or NMOSD1 with unknown AQP42-IgG3 status
1.	At least two core clinical characteristics plus all of the following 1. At least one among optic neuritis, acute myelitis with longitudinally extensive transverse myelitis lesions, or area prostrema syndrome 2. Dissemination in space 3. Magnetic resonance imaging requirements(MRI)[4], as applicable
2.	Negative tests for AQP42-IgG3 or testing unavailable
3.	Exclusion of alternative diagnoses
	Core clinical characteristics
1	Optic neuritis
2	Acute myelitis
3	Area prostrema syndrome: an episode of unexplained hiccups, nausea, or vomiting
4	Acute brainstem syndrome
5	Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD1-typical diencephalic MRI4 lesions
6	Symptomatic cerebral syndrome with NMOSD1-typical brain lesions

NMOSD[1] neuromyelitis optica spectrum ofdisorder, AQP4[2] aquaporin 4, IgG[3] Immunoglobulin, MRI[4] magnetic resonance imaging

relaxants versus central neuraxial blockade in patients who may benefit from either, it's crucial to carefully evaluate the risks and benefits associated with each option. Additionally, ensuring sufficient pain management is essential for addressing chronic pain associated with the disease.

4. Conclusion

The perioperative management of NMOSD is challenging because of the risk of acute and chronic complications. However, streamlining anesthetic management to prevent these complications can result in good patient outcomes.

5. Source of Funding

None.

6. Conflict of Interest

None.

References

1. Kasper DL, Braunwald E, Fauci AS, Hauser SL, Longo DL, Jameson JL. Harrison's Principles of Internal Medicine. 16th ed. New York, NY: McGraw-Hill Companies; 2005. p. 2444–70.
2. Ransohoff RM. Illuminating neuromyelitis optica pathogenesis. *Proc Natl Acad Sci U S A*. 2012;109(4):1001–2.
3. Hor JY, Asgari N, Nakashima I, Broadley SA, Leite MI, Kissani N, et al. Epidemiology of Neuromyelitis Optica Spectrum Disorder and Its Prevalence and Incidence Worldwide. *Front Neurol*. 2020;11:501.
4. Sadana N, Houtchens M, Farber MK. Anesthetic management of a parturient with neuromyelitis optica. *Int J Obstet Anesth*. 2012;21(4):371–5.
5. Dusitkasem S, Subsoontorn P. General anaesthesia for caesarean section in a patient with neuromyelitis optica spectrum disorder (NMOSD). *Int J Surg Case Rep*. 2019;60:72–4.
6. Facco E, Giorgetti R, Zanette G. Spinal anaesthesia and neuromyelitis optica: cause or coincidence? *Eur J Anaesthesiol*. 2010;27(6):578–80.
7. Hosseini H, Brugieres P, Degos JD, Cesaro P. Neuromyelitis optica after a spinal anaesthesia with bupivacaine. *Mult Scler*. 2003;9(5):526–8.
8. Kuberan A, Ramadurai R, Balasubramanian M, Dey A. Anaesthetic concerns in pregnancy associated with myelin oligodendrocyte glycoprotein encephalomyelitis. *ndian J Anaesth*. 2022;66(8):614–6.
9. Neuromyelitis Optica Spectrum Disorder (NMOSD). Accessed May 4, 2023. Available from: https://www.nationalmssociety.org/understanding-ms/what-is-ms/related-conditions/nmosd.

Author biography

Roshin Binoy, Junior Resident https://orcid.org/0009-0007-2863-1954

Bincy V Thomas, Assistant Professor

Sheela Verghese, Professor & HOD

Cite this article: Binoy R, Thomas BV, Verghese S. Devic's disease: A devil's trap for the Anesthesiologist: A case report. *Indian J Clin Anaesth* 2024;11(2):258-260.