SCIENTIFIC JOURNAL
CEREM-GO
DOI 10.37951/2675-5009.2025v6i16.168
ISSN 2675-5009
e25168
SCIENTIFIC ARTICLE - CASE REPORT

ANESTHETIC MANAGEMENT IN A PATIENT WITH SUSAC SYNDROME: A CASE REPORT

GABRIEL PEIXOTO NASCIMENTO[1], DIRCEU CASTRO PACHECO[1], GUSTAVO SIQUEIRA ELMIRO[1], GIULLIANO GARDENGHI [1,2]

1. Clínica de Anestesia, Goiânia/GO, Brasil.
2. Hospital ENCORE, Aparecida de Goiânia/GO, Brasil.

ABSTRACT

Susac's Syndrome (SS) is defined as an occlusive microangiopathy of autoimmune etiology, characterized by the involvement of arterioles in the cerebral, retinal, and cochlear vascular beds. The classic clinical triad manifests as encephalopathy, visual deficits, and hearing loss. Given its relative rarity and variable clinical presentation, SS frequently poses a diagnostic challenge, resulting in underdiagnosis or misdiagnosis. Currently, the predominant therapy for SS involves immunomodulation with corticotherapy. This case report describes the clinical course of a 36-year-old female patient who underwent inhalational general anesthesia for elective laparoscopic cholecystectomy. Although invasive intracranial pressure monitoring was not implemented, the anesthetic management proved effective for the patient in question, highlighting the importance of a detailed clinical and neurological evaluation in the perioperative period, even in procedures considered to be of low neurological risk.

Keywords: Anesthesia general, Anesthesia intravenous, Susac syndrome, Cholecystectomy laparoscopic, Primary immunodeficiency diseases, Anesthesia.

INTRODUCTION

Thalassemias Susac Syndrome (SS) is defined as an autoimmune endotheliopathy characterized by a clinical triad of encephalopathy (with or without focal neurological signs), retinal artery occlusions, and hearing loss[1]. The pathogenesis of SS involves a microangiopathy affecting the precapillary arterioles of the retina, inner ear, and brain parenchyma.[2]

This disease predominantly affects young adults, with an average age of symptom onset around 18 years and a female-to-male ratio of 3.5:1.[3] SS can present with a wide spectrum of symptoms, including headache, mental confusion, personality and behavioral changes, ataxia, loss of balance, and dysarthria. In some cases, isolated vision loss or hearing loss may be the initial symptom.[4]

Due to its relative rarity and heterogeneous clinical presentation, the syndrome is often underdiagnosed or misdiagnosed, usually being identified only after the exclusion of other neurological, psychiatric, ophthalmological, and auditory conditions.[5] Characteristic findings on magnetic resonance imaging and audiometry contribute significantly to establishing the diagnosis.[4]

Current therapeutic recommendations for SS are based on expert-developed guidelines,

longitudinal follow-up of various patient cohorts, and recommendations extrapolated from other severe autoimmune diseases with similar immunopathogenesis. In this context, immunosuppression constitutes the cornerstone of treatment, with therapy intensity being adjusted primarily according to the severity of central nervous system involvement.[6]

Due to its rarity and the diagnostic challenge it still presents, there is a lack of studies in the medical literature addressing the management of this disease in the context of anesthesiology. In light of this, the present scientific article aims to report the anesthetic management of a patient with SS who underwent general anesthesia.

CASE REPORT

A 36-year-old female patient with a diagnosis of SS underwent elective videolaparoscopic cholecystectomy. On pre-anesthetic clinical evaluation, she presented with hearing impairment, did not respond to verbal commands, was hyperactive, and minimally cooperative. Additionally, she was overweight and classified as Mallampati class III.

Anesthetic induction was performed with 250 micrograms (mcg) of fentanyl, 150 milligrams (mg) of propofol, and 80 mg of succinylcholine administered intravenously, followed by direct laryngoscopy, which revealed a Cormack-Lehane grade 2A view. Orotracheal intubation was performed using a 7.0 mm cuffed endotracheal tube. Additional neuromuscular relaxation was achieved with 30 mg of rocuronium. General inhalational anesthesia was maintained with 2% sevoflurane. Additionally, adjuvant medications included 8 mg of ondansetron, 10 mg of dexamethasone, 2 grams of dipyrone, and 40 mg of parecoxib.

The surgical procedure proceeded without complications and lasted 60 minutes. Monitoring included pulse oximetry, cardiac monitoring, non-invasive blood pressure, and temperature measurement. Intracranial pressure was not assessed before, during, or after anesthesia.

At the end of the procedure, neuromuscular blockade was reversed with 200 mg of sugammadex; however, the patient experienced difficulty resuming spontaneous ventilation, resulting in carbon dioxide (CO_2) retention. Arterial blood gas analysis was consistent with severe respiratory acidosis. Consequently, the first extubation attempt failed, and mechanical ventilation had to be prolonged to correct the respiratory-origin acid–base imbalance.

Approximately 30 minutes after the first extubation attempt, the patient was successfully extubated without complications. Her recovery took place in the post-anesthesia care unit within the surgical center, where she remained for two hours before being transferred to the ward.

DISCUSSION

Patients with neurological manifestations of SS are predisposed to elevated intracranial pressure (ICP)[7]. In this context, the use of non-invasive techniques—such as ultrasonographic assessment of the optic nerve sheath diameter—has shown diagnostic accuracy for detecting increased ICP in the intraoperative setting. However, this method was not employed in the anesthetic management of the patient in the present case.

Furthermore, the choice of anesthetic technique should be based on the presence or absence of intracranial hypertension[1]. In this regard, it is recommended to minimize the patient's exposure to stimuli that could increase ICP, such as sympathetic stimulation. Accordingly, direct laryngoscopy was performed only under an adequate anesthetic plane and after the

administration of fentanyl at a dose of 4 to 5 mcg/kg.

Pneumoperitoneum is known to induce reflex vasodilation, which can lead to ICP[8]. To mitigate this effect, peritoneal insufflation was performed gradually rather than abruptly, maintaining intra-abdominal pressure below 12 mmHg. Although the use of lower insufflation pressures may be considered, the most effective strategy for minimizing the impact on ICP would be the use of abdominal wall lift systems for performing videolaparoscopy[9].

Another measure aimed at preventing increased ICP was the reduction of arterial partial pressure of carbon dioxide ($PaCO_2$) during the period the patient remained under controlled mechanical ventilation, through hyperventilation. The decrease in $PaCO_2$ induces alkalosis, and the resulting increase in blood pH has a direct effect on cerebral arterioles, promoting vasoconstriction, reducing intracranial blood volume, and consequently lowering ICP[10]. However, at the time of extubation, the patient presented with carbon dioxide retention secondary to apnea, progressing to severe respiratory acidosis. Thus, the effect of hyperventilation on ICP may not have been beneficial during extubation.

Regarding the selection of anesthetic agents in cases of intracranial hypertension, intravenous propofol, used in total intravenous anesthesia, is known to induce cerebral vasoconstriction and reduce ICP[11], compared to inhalational agents. On the other hand, in the absence of intracranial hypertension, the use of propofol may not be advisable, as it may exacerbate cerebral, retinal, and cochlear hypoperfusion in the context of preexisting microangiopathy in SS[1].

Other interventions for ICP control, such as the administration of hypertonic saline and diuretics, were not implemented during the anesthetic management of this case. However, elevation of the head of the bed while in the supine position was performed, as it also represents an appropriate surgical positioning for the proposed procedure.

In conclusion, the anesthetic management described in this case proved to be effective in the anesthetic care of a patient with SS undergoing elective videolaparoscopic cholecystectomy. Although invasive ICP monitoring was not performed, measures were implemented with the aim of preventing its elevation, as detailed above.

However, there is a significant lack of studies addressing anesthetic management in patients with this syndrome, highlighting the need for future research on the topic.

Other interventions for ICP control, such as the administration of hypertonic saline and diuretics, were not implemented during the anesthetic management of this case. However, head elevation in the supine position was performed, as it also represents an appropriate surgical positioning for the proposed procedure.

CONCLUSION

The anesthetic management described in this case proved to be effective in the anesthetic care of a patient with Susac's syndrome undergoing elective laparoscopic cholecystectomy. Although invasive ICP monitoring was not performed, measures were implemented to prevent its elevation, as detailed. Nevertheless, there is a significant lack of studies addressing anesthetic management in patients with this syndrome, highlighting the need for further research on the subject.

REFERENCES

1. Khandelwal A, Prasad C, Sokhal N, Chaturvedi A. Anesthetic management of a patient with Susac syndrome: A rare neurological disorder. Saudi J Anaesth. 2020;14(4):551-552.

2. Grygiel-Górniak B, Puszczewicz M, Czaplicka E. Susac syndrome-clinical insight and strategies of therapy. Eur Rev Med Pharmacol Sci. 2015;19:1729-35.

3. Jarius S, Kleffner I, Dörr JM, Sastre-Garriga J, Illes Z, Eggenberger E, Chalk C, Ringelstein M, Aktas O, Montalban X, Fechner K, Stöcker W, Ringelstein EB, Paul F, Wildemann B. Clinical, paraclinical and serological findings in Susac syndrome: an international multicenter study. J Neuroinflammation. 2014 Mar 8;11:46.

4. Zengin Karahan S, Boz C, Saip S, Kale N, Demirkaya S, Celik Y, Demir S, Kurne A, Ozbek SE, Terzi M. Susac Syndrome: Clinical characteristics, diagnostic findings and treatment in 19 cases. Mult Scler Relat Disord. 2019;33:94-99.

5. Marrodan M, Fiol MP, Correale J. Susac syndrome: challenges in the diagnosis and treatment. Brain. 2022;145(3):858-871.

6. Rennebohm RM, Egan RA, Susac JO. Treatment of Susac's syndrome. Curr Treat Options Neurol. 2008;10(1):67–74.
7. Dubourg J, Javouhey E, Geeraerts T, Messerer M, Kassai B. Ultrasonography of optic nerve sheath diameter for detection of raised intracranial pressure: a systematic review and meta-analysis. Intensive Care Med. 2011;37(7):1059-68.

8. Rosenthal RJ, Hiatt JR, Phillips EH, Hewitt W, Demetriou AA, Grode M. Intracranial pressure. Effects of pneumoperitoneum in a large-animal model. Surg Endosc. 1997;11(4):376-80.

9. Este-McDonald JR, Josephs LG, Birkett DH, Hirsch EF. Changes in intracranial pressure associated with apneumic retractors. Arch Surg. 1995;130(4):362-5.

10. Lundberg N, Kjalqüist A, Kullberg G, Pantén V, Sundbarg G. Non operative management of intracranial hypertension. In: Krayenbull H (ed.): Advances and Technical Standards in Neurosurgery. New York, Springer-Verlag; 1974. p. 3-59.

11. Petersen KD, Landsfeldt U, Cold GE, Pedersen CB, Mau S, Hauerberg J, Holst P. ICP is lower during propofol anaesthesia compared to isoflurane and sevoflurane. Acta Neurochir Suppl. 2002;81:89-91

MAILING ADDRESS

GIULLIANO GARDENGHI
CET – CLIANEST, R. T-32, 279 - St. Bueno, Goiânia - GO, Brazil
E-mail: coordenacao.cientifica@ceafi.edu.br

EDITORIAL AND REVIEW

Chief editors:

Waldemar Naves do Amaral - http://lattes.cnpq.br/4092560599116579 - https://orcid.org/0000-0002-0824-1138
Tárik Kassem Saidah - http://lattes.cnpq.br/7930409410650712- https://orcid.org/0000-0003-3267-9866

Authors:

Gabriel Peixoto Nascimento - http://lattes.cnpq.br/1553201526937403 - https://orcid.org/0000-0002-6607-7110

Dirceu Castro Pacheco - http://lattes.cnpq.br/9259558054810108 - https://orcid.org/0009-0009-5316-482X

Gustavo Siqueira Elmiro - http://lattes.cnpq.br/4765163399934337 - https://orcid.org/0000-0003-2113-8757

Giulliano Gardenghi - http://lattes.cnpq.br/1292197954351954 - https://orcid.org/0000-0002-8763-561X

Library Review: Izabella Goulart
Spell Check: Dario Alvares
Received: 08/05/25. Accepted: 12/05/25.. Published in: 23/06/2025.