SSI-YAN-KAÏ Nicolas
MACCS 1ère année Anesthésie-Réanimation
UCL Mont-Godinne,
Présenté le 25/03/2015.

STIFF PERSON SYNDROM (SPS)

1 – ABSTRACT

Stiff person syndrom is a rare and progressive neurologic disorder, with symptoms of rigidity and spasms affecting axial and proximal muscles, and slowing ambulation. An autoimmune mechanism is found in most cases, with detection of antibodies to GAD or non-organ-specific autoantibodies associated with neoplasms. Sometimes, this disease is idiopathic because no evidence of autoantibody production is found. We here describe the anesthetic management of a man with idiopathic stiff person syndrome.
A 64 years old male patient, who had been suffering from SPS for 2 years, consulted for cervical magnetic resonance imaging (MRI) under general anesthesia in order to exclude an irritating spine, that could cause his cervical pain. We chose total intravenous anesthesia for managing this patient, using a laryngeal mask to control the airway. The procedure lasted for approximately 1 hour and was uneventful. The chosen anesthetic agent was propofol, and an effect-site concentration of 4 µg/mL was maintained. At the end of MRI, the patient recovered good spontaneous ventilation, and his laryngeal mask was removed easily. There was no adverse event monitored in the recovery room.

Comment [Bonhomme 1]: Please define abbreviations at first use

2 – KEY WORDS

Idiopathic stiff person syndrome – general anesthesia – volatile anaesthetic agents – propofol – TIVA – muscle relaxants.

3 – INTRODUCTION

Stiff person syndrome is a rare and progressive neurologic disorder, with symptoms of rigidity and spasms affecting axial and proximal muscles, and slowing ambulation. An autoimmune pathogenesis is suspected, and would lead to GABAergic neuron inhibition within the brain and spinal cord. There are no recommendations about anesthetic care in those patients, but several cases have already been reported. We here report the anesthetic management of a patient suffering from SPS, and undergoing magnetic resonance imaging (MRI) under general anesthesia.

4 – CASE REPORT

A 64 years old male patient, who had been suffering from SPS for 2 years, consulted for cervical MRI, in order to exclude an irritating spine that could cause cervical pain. He also suffered from chronic high blood pressure, had a past medical history of pneumonia, 3 pulmonary embolisms 9 years ago, and 3 ischemic strokes 6 years ago. He was allergic to seafood and grasses. His medications included Asaflow, Sintrom, Medrol, Imuran, Baclofen and Tramadol. He had undergone foraminectomy 3 years ago to cure a left sciatalgy with no improvement under medical treatment.
The early post-operative period after that surgery went well. Three weeks after, he started complaining of abdominal and back pain. Those symptoms worsened and, after investigation, the

Comment [Bonhomme 2]: Please use generic names

diagnosis of an idiopathic stiff person syndrom limited to abdominal muscles was made. His treatment first consisted in diazepam, rapidly replaced by Baclofen and corticosteroids. Imuran was started one year later, in face of the absence of improvement.
When he came to his preoperative consultation, he showed impaired ambulation with severe rigidity and pain in lumbar, abdominal and cervical region. There was no sign of difficult intubation or ventilation. His cardiorespiratory system and his blood testing were normal. We decided to maintain his treatment without premedication.
Total intravenous anesthesia (TIVA) was our preferred technique to manage this patient during cervical MRI, because we had found several successful case reports described with this technique. General anesthesia was induced with propofol at a target concentration of 3 µg/mL, which was rapidly increased to 4 µg/mL. Mask ventilation was easy, as well as laryngeal mask size 5 insertion. Anesthesia was maintained using propofol at the same target of 4 µg/mL. At the end of MRI examination, and after cessation of propofol infusion, the patient recovered good spontaneous ventilation rapidly, and his laryngeal mask was removed easily. There was no adverse event monitored at the recovery room.

Comment [Bonhomme 3]: Please use generic names.

5 – DISCUSSION

Stiff syndrom person was first described in 1956 by Moersch and Woltman from the Mayo clinic. Its prevalence is estimated to be 1/1 000 000. It is twice more common in females, and appears around the third decade. It is clinically characterized by progressive muscle rigidity, and painful spasms affecting the axial and limb musculature.
SPS is said to have an autoimmune mechanism since it occurs in conjunction with a variety of autoimmune diseases such as type 1 diabetes mellitus, vitiligo, or pernicious anemia. In addition, polyclonal and oligoclonal IgG antibody elevations have been observed in the cerebrospinal fluid of the majority of patients with SPS. These antibodies were found to target and inhibit GABAergic neurons and their nerve terminals.
Treatment of SPS is first symptomatic, including medications such as benzodiazepines (diazepam, GABA-A agonist) or, if necessary, Baclofen (GABA-B agonist). When the disease progresses, corticosteroids, azathioprin, or intravenous immunoglobulins may be necessary. Plasma exchange can also be considered **[1]**.

Comment [Bonhomme 4]: Please use generic names.

Use of regional anesthesia in these patients can be advantageous, as it provides effective and deep analgesia without necessitating the use of muscle relaxants and inhalational agents. Indeed, those agents could interfere with the ongoing treatment. Several successful managements with regional anesthesia have already been reported. However, positioning and anatomical landmark spotting can reveal difficult. Puncture can also induce painful spams and rigidity **[2] [3]**.
In our situation, total immobility and muscle relaxation were needed, that's why general anesthesia was chosen.
Therapy with medications such as Diazepam or Baclofen must be persued during the perioperative period, because of the risks associated with withdrawal. Corticosteroids should also not be stopped. When necessary, plasma exchange or intravenous globulin therapy must be scheduled to help decreasing anti-GAD antibody levels **[2]**.
No cases of difficult intubation in a SPS patient has been reported. However, those patients are vulnerable to sudden stimulation and sounds, which could precipitate painful muscle spasms. That's why general anesthesia should be deepened before any airway manipulation. Premedication with midazolam can also be useful **[2]**.

Comment [Bonhomme 5]: Please use generic names

Comment [Bonhomme 6]: Please detail abbreviation at first use.

Prolonged hypotonia after general anesthesia with inhalational anesthetics has already been described, necessitating postoperative artificial ventilation for 2 hours. Isoflurane was suspected to potentiate the effects of Baclofen, and therefore induce this adverse effect **[4]**. That's why volatile anesthetics are no more recommended in these patients. Propofol TIVA has already been reported to

be effective and to minimize chances of hypotonia **[5][6][7]**.

Use of muscle relaxants may also potentiate hypotonia, and increase the risk of post-operative mechanical ventilation. Prolonged hypotonia under vecuronium has already been described **[8]**. There are no contraindications to the use of non-depolarizing relaxants in patients with SPS. However, it is recommended to avoid them as much as possible. In case they cannot be avoided, the smallest titrated doses of short acting relaxants should be used, and with close monitoring of neuromuscular function, keeping in mind that there could be no correlation between depth of muscle paralysis and observed hypotonia **[9]**.
Combined TIVA with high levels of morphinomimetics such as remifentanyl and propofol has already been used successfully to provide adequate relaxation and analgesia for intubation or surgery, without the use of any muscle relaxant.
In any event, close monitoring in recovery room remains recommended, because of the high risk of prolonged postoperative hypotonia in patient with SPS.

6 – CONCLUSION

Reports on anesthetic management in stiff person syndrome remain rare. There are currently no recommendations concerning their anesthetic management. We report a general anesthesia technique for managing a patient suffering with SPS and undergoing MRI. His initial Baclofen treatment was not discontinued preoperatively, in order to decrease the risk of withdrawal symptoms. Propofol was used as a sole agent through a target-controlled infusion preferably to inhalational anesthetic agents, to minimize the risk of prolonged postoperative hypotonia. No adverse event was recorded during the procedure and the postoperative period.

Comment [Bonhomme 7]: Please use generic names.

7 – REFERENCES

1 – Helfgott, S. Stiff-person syndrome. UpToDate, 2015.
2 – Shanthanna, H., Ferrandis, R., Goldkamp, J. Anesthesia recommandations for patients suffering from stiff man syndrome. Orphananesthesia, 2014.
3 – Shanthanna, H. Stiff Man Syndrome and Anaesthetic Considerations: Successful Management Using Combined Spinal Epidural Anaesthesia. J Anaesthesiol Clin Pharmacol., 2010 : 547–548.
4 – Bouw, J., Leendertse, K., Tijssen, M., Dzoljic, M. Stiff person syndrome and anesthesia : Case report. Anesth Analg, 2003 ; 97:486-7.
5 – Toscano, F., Vick, A., Shay, H., Delphin, E. Total Intravenous Anesthesia (TIVA) for Stiff-Person Syndrome. Open Journal of Anesthesiology, 2012, 2, 185-187.
6 – Ledowski, T., Russell, P. Anesthesia for stiff person syndrome : successful use of total intravenous anaesthesia. Anaesthesia, 2006 ; 61 : 714-726.
7 – Yagan, O., et al. Anesthesia in a patient with Stiff Person Syndrome. Rev Bras Anestesiol, 2014.
8 – Johnson, J., Miller, K. Anesthetic Implications in Stiff-Person Syndrome. Anesthesia and analgesia, 1995 ; 80 : 612-613.
9 – Sidransky, M., Tran, N., Kaye, A.D. Anesthesia considerations in stiff person syndrome. M.E.J. Anesth, 22 (2), 2013.