

Anesthesia in a Patient with Stiff Person Syndrome: About A Case

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Abstract

Case Report

Stiff person syndrome, characterized by stiffness in the muscles of the torso and extremities and painful episodic spasms, is a rare autoimmune neurological disease. Here we present successful endotracheal intubation. A 46-year-old man underwent surgery with ASA-III classification due to necrosectomy following fasciitis necrotizing. After induction of anesthesia using fentanyl propofol and rocuronium, tracheal intubation proceeded easily without neuromuscular blockade. Anesthesia was maintained with isoflurane and an O₂/air mixture. After an uneventful intraoperative period, the patient was extubated. The evolution was marked by death following a septic shock. Although the mechanism is unclear, neuromuscular blockers and volatile anesthetics can cause prolonged hypotonia in patients with stiff-man syndrome.

Keywords: Stiff person syndrome, anesthesia, immunotherapy.

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INTRODUCTION

Stiff person syndrome (SPS) is a rare and disabling disorder characterized by muscular rigidity with superimposed painful spasms involving axial musculature and limbs [1].

The stiff person syndrome is characterized by paraspinal and abdominal rigidity with exaggerated lumbar lordosis and superimposed spasms precipitated by movement, emotional upset, peripheral stimulation or auditory startle [2].

Most patients belonging to classical SPS have antibodies against glutamic acid decarboxylase (GAD), which is the enzyme essential for GABA synthesis.

Loss of GABAergic supply from higher centers and spinal interneurons causes hyperactivity of the motor neuron system and subsequent progressive muscle rigidity [3].

Treatment is mostly based on benzodiazepines, baclofen and immunosuppressants.

CASE REPORT

A 46-year-old man followed in internal medicine since November 2020 for stiff person

syndrome retained in the face of joint stiffness, muscle rigidity and the presence of anti-GAD antibodies put on immunotherapy (3rd bolus received on June 11, 2021).

After two months of the last bolus, there is an installation of a picture of necrotizing fasciitis for which he had to undergo an extended necrosectomy of the two lower limbs.

In Preoperative

The patient was informed about the nature of the surgery and the procedure as well as the anesthetic risk.

Patient with a Glasgow score of 13, HR at 98 bpm, BP at 130/70 mmHg, FR at 25 bpm and SaO₂ at 98% on room air.

On the local level, inflammatory plaque with bullous lesions of necrotizing appearance with resulting pus at the level of the two lower limbs.

Soft tissue ultrasound showed soft tissue infiltration and diffuse skin thickening.

Doppler ultrasound without signs of thrombophlebitis

ECG without abnormality

Put on antibiotics since his admission: ceftriaxone, aminoglycoside and metronidazole.

| | 09/12/2021 | 09/16/2021 |
|-------------------|------------|-------------|
| leukocytes | 46300 | 38010 |
| PNN / Lymphocytes | 44480/1190 | 33740/3400 |
| HB/HTC | 13 / 37.9 | 7.4/22.6 |
| pads | 50000 | 132000 |
| TP / TCA | 63.6/28.3 | 54.6 / 26.3 |
| Na/K | | 138/5.2 |
| Urea/ creat | | /5.1 |
| CRP | | 48.38 |
| AST/ALT | | 25/25 |

Intraoperatively

Induction of anesthesia was performed with fentanyl 2 mcg /kg, propofol 2.5 mg/kg, rocuronium 0.6 mg/kg. Endotracheal intubation was easily performed and the lungs were ventilated with an O2/air mixture at 6ml/kg.

Central venous, arterial and bladder catheters were placed.

Anesthesia was maintained with isoflurane, bolus anesthetics were not required.

Monitoring by curameter was done.

The patient presented hemodynamic instability made of hypotension improved by transfusion with two globular concentrates and introduction of norepinephrine at 0.48mg.

Analgesia was used to relieve postoperative pain.

At the end of the operation the TOF was 4 out of 4 before extubation which was uneventful, the patient was transferred to the surgical intensive care unit.

Postoperatively

Patient admitted to intensive care unit for postoperative monitoring.

Patient stabilized on noradrenaline 0.8 mg, and put on imipenem, amikacin, quinilone and vancomycin.

Evolution marked by the appearance of postoperative complication related to septic shock with multiorgan failure.

In 3rd daypostoperative, the patient suffered a cardiac arrest that did not recover.

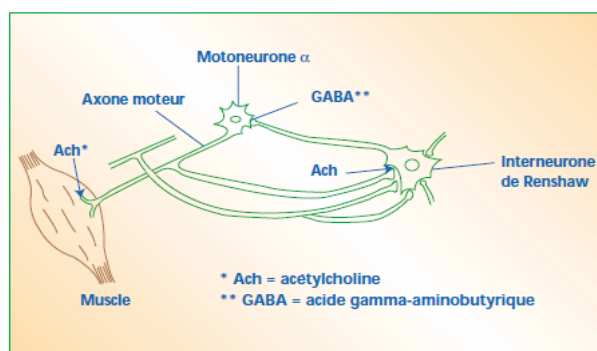
DISCUSSION

Stiff person syndrome is a rare condition characterized by progressive stiffness of the muscles of the trunk and the root of the limbs, on which are grafted painful spasms. The incidence is the same in men and women.

The association with autoimmune diseases, such as insulin-dependent diabetes (in 30% of cases) and thyroiditis is quite common with the presence of anti-GAD antibodies. The SPS can sometimes be integrated into a paraneoplastic framework.

The EMG shows continuous muscle activity, despite the patient's desire to relax.

Norepinephrine (catecholamines) and gamma-aminobutyric acid (GABA) control the activity of motor neurons. GABA is one of the main neurotransmitters in the brain with inhibitory activity. Glutamic _ sour decarboxylase (GAD) is necessary for the synthesis of GABA. The existence of anti-GAD autoantibodies decreases the level of GABA and, thus, the excess of norepinephrine at the synaptic junction causes this continuous contraction of the axial muscles and the limbs.



Among the effective treatments for stiffness, benzodiazepines (GABA-A receptor agonists) are the most widely used; they increase cortical and spinal inhibition [5]. EMG shows that diazepam helps reduce simultaneous agonist- antagonist muscle contractions [6].

Treatment may be with baclofen (GABA-B receptor agonist) and similar GABAergic agents and steroids, plasmapheresis, or immunoglobulins for immune modulation [5, 7, 8].

There are various studies on anesthesia methods for SPS patients in the literature.

Johnson and Miller [9] found that muscle weakness required mechanical ventilation after using thiopental, sufentanil, vecuronium, and isoflurane for general anesthesia during a pump implantation operation. Intrathecal baclofen in an SPS patient. Five months later, the same procedure was performed without any problems under general anesthesia without the use of neuromuscular blockers and they advised to avoid the use of non-depolarizing muscle relaxants in SPS patients.

Bouw *et al.*, [10] noted prolonged hypotonia after general anesthesia in an SPS patient operated on for colon carcinoma using propofol, sufentanil, atracurium, and isoflurane. Pharmacokinetic analysis showed that plasma concentrations of atracurium and sufentanil were well below therapeutic levels. They proposed that patients using baclofen during the preoperative period may suffer from hypotonia due to the effects of volatile anesthetic agents on GABA receptors.

Obara *et al.*, [11] presented a case study of an SPS patient who underwent thymectomy, appendectomy, and endoscopic sinus surgery within one year. Diazepam, propofol, thiopental, fentanyl nitrous oxide, isoflurane, and vecuronium have been used for general anesthesia. The patient awoke from all three procedures in a short time without any extubation problems. However, they do not mention whether or not the patient used baclofen or other GABAergic agents in the preoperative period.

Ledowski and Russell [4] used TIVA without neuromuscular blockers in an SPS patient undergoing ENT surgery and noted that the patient was discharged without complications.

Regional anesthesia has been used successfully in SPS patients. Shanthanna [12] used combined spino-epidural anesthesia for planned total knee arthroplasty in an SPS patient, while Elkassabany *et al.*, [13] used a paravertebral block for inguinal hernia surgery. However, they emphasized that, as needle pain, fear and anxiety can trigger spasms, it is important that detailed

disclosure is made in the preoperative period and that sufficient sedation is ensured during the procedure.

The literature shows that suitable conditions for tracheal intubation under general anesthesia can be provided without the use of neuromuscular agents. [14] A combination of hypnotics and opioid drugs has been used.

Although the mechanism is unclear, the use of neuromuscular blocking agents and volatile anesthetics can cause prolonged hypotonia in SPS patients.

CONCLUSION

Due to some anesthetic medications interacting with GABA receptors, anesthesia in SPS patients requires special attention.

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