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CLINICAL INFORMATION

Anesthesia in a patient with Stiff Person Syndrome☆

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Abstract Stiff Person Syndrome (SPS), typified by rigidity in muscles of the torso and extremities and painful episodic spasms, is a rare autoimmune-based neurological disease. Here we present the successful endotracheal intubation and application of TIVA without muscle relaxants on an SPS patient.

A 46 years old male patient was operated with ASA-II physical status because of lumber vertebral compression fracture. After induction of anesthesia using lidocaine, propofol and remifentanil tracheal intubation was completed easily without neuromuscular blockage. Anesthesia was maintained with propofol, remifentanil and O_2/air mixture. After a problem-free intraoperative period the patient was extubated and seven days later was discharged walking with aid.

Though the mechanism is not clear neuromuscular blockers and volatile anesthetics may cause prolonged hypotonia in patients with SPS. We think the TIVA technique, a general anesthetic practice which does not require neuromuscular blockage, is suitable for these patients.

PALAVRAS-CHAVE
Anestesia;
Anestesia venosa total;
Síndrome da pessoa rígida

Anestesia em paciente com síndrome da pessoa rígida

Resumo A síndrome da pessoa rígida (SPR), caracterizada pela rigidez dos músculos do tronco e extremidades e episódios de espasmos dolorosos, é uma doença neurológica autoimune rara. Apresentamos o ocaso de intubação endotraqueal bem-sucedida e aplicação de AVT sem relaxantes musculares em um paciente com SPR.

☆ This case report was presented as a poster at Turkish Anesthesiology and Reanimation Association 46th National Congress in Antalya, Turkey, in 2012.

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Paciente do sexo masculino, 46 anos de idade, estado físico ASA-II, submetido à cirurgia devido à fratura por compressão da coluna lombar. Após a indução da anestesia com lidocaína, propofol e remifentanil, a intubação traqueal foi concluída com facilidade, sem bloqueio neuromuscular. A anestesia foi mantida com propofol, remifentanil e mistura de ar/O_2. Após o período intraoperatório que transcorreu sem intercorrências, o paciente foi extubado e, sete dias depois, recebeu alta, deambulando com ajuda.

Embora o mecanismo não esteja claro, bloqueadores neuromusculares e anestésicos voláteis podem causar hipotonia prolongada em pacientes com SPR. Acreditamos que a técnica de AVT, uma prática de anestesia geral que não requer bloqueio neuromuscular, é adequada para esses pacientes.

Introduction

Stiff Person Syndrome (SPS), typified by rigidity in muscles of the torso and extremities and painful episodic spasms, is a rare autoimmune-based neurological disease.[1] It is thought that autoantibodies that attack the glutamic acid decarboxylase (GAD) enzyme necessary for physiopathologic synthesis of GABA may be responsible. In the higher centers without the effect of GABAergic inhibitors hyperactivity of the motor neuron system may cause consequent progressive muscle rigidity.[2] Treatment includes medication to increase GABA activity and immunosuppressants. Due to some anesthetic medications interacting with GABA receptors, anesthesia in SPS patients requires special attention.[3] We present a successful endotracheal intubation and TIVA application without muscle relaxants in an SPS patient.

Case report

A 46-year-old male patient was diagnosed with SPS 7 years previously. Treatment with diazepam (15 mg day^{-1}), baclofen (30 mg day^{-1}) and prednisolone (20 mg day^{-1}) reduced symptoms but for the previous two months the patient complained of increased painful spasms especially in the lower extremities. The patient could walk with help, but a thoracolumbar CT scan was done to investigate an increase in a two-year lower back pain complaint. The scan revealed a compression fracture at lumbar 2–5 level and an operation was planned. The patient's history only included hypertension. Physical examination revealed increased tone in the lower extremities. On consultation with an internal expert preoperative 20 mg prednisolone (single dose) im was advised. Extra medication used included bisoprolol, lansoprazole, tramadol, calcium and vitamin D supplements. With these findings the patient was accepted as ASA risk group II.

Other than these medications, premedication was not given. Coincident with standard monitoring invasive blood pressure measurements were taken. Induction of anesthesia was done with midazolam 0.1 mg kg^{-1}, lidocaine 1.5 mg kg^{-1}, propofol 2 mg kg^{-1} and remifentanil 2 mcg kg^{-1}. Without using a neuromuscular blocker endotracheal intubation was easily achieved and the lungs were ventilated with an O_2/air mix. Central venous and bladder catheters were emplaced. Anesthesia was maintained by 60–100 mcg kg^{-1} min^{-1} propofol and 0.1–0.8 mcg kg^{-1} min^{-1} remifentanil infusion. Intraoperative hemodynamics remained stable, bolus anesthetics were not required and the patient was given 2 units of packed red blood cells for bleeding. Postoperative analgesia was intravenous 75 mg tramadol and 25 mg dexketoprofen. At the end of the operation after a problem-free extubation the patient was transferred to the surgical intensive care unit. With no follow-up problems the patient was moved to the wards the following day and 7 days later was discharged walking with aid again.

Discussion

Stiff Person Syndrome (SPS) was first described by Moersch and Woltman in 1956.[4] While the cause is unknown, the presence of GAD antibodies in the cerebrospinal fluid (60–70%) and coincidence with other autoimmune diseases such as diabetes mellitus and thyroiditis suggests an immunological basis. The syndrome is progressive, severe muscle rigidity and sudden onset spasms are common. It affects the lower extremities and vertebrae of nearly all patients. The symptoms may be triggered by psychological stress, sudden sounds or visual warnings and touch. Autonomic symptoms (tachycardia, hyperhidrosis, blood pressure changes, constipation, urinary retention, etc.) accompany spasms. The syndrome develops in middle age. It commonly occurs with autoimmune diseases and cancer (as paraneoplastic syndrome).[5] EMG shows that diazepame helps to reduce the agonist-antagonist simultaneous muscle contractions.[6] Treatment may be with benzodiazepines (GABA-A receptor agonists) which increase cortical and spinal inhibition, baclofen (GABA-B receptor agonist) and similar GABAergic agents and steroids, plasmapheresis or immunglobulins for immune modulation.[5,7,8] Our patient had been using diazepam, baclofen and prednisolone since his diagnosis in 2005.

There are various studies on anesthetic methods for SPS patients in the literature. Johnson and Miller[9] found muscle weakness required mechanic ventilation after using

thiopental, sufentanil, vecuronium and isoflurane for general anesthetic in an operation to implant an intrathecal baclofen pump in an SPS patient. Five months later the same procedure was completed without any problems under general anesthetic without using neuromuscular blockers and they advised avoiding the use of non-depolarising muscle relaxants in SPS patients.

Bouw et al.[10] noted prolonged hypotonia after general anesthetic 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 in the preoperative period may experience hypotonia due to the effects of volatile anesthesia agents on GABA receptors.

Obara et al.[11] presented a case study of an SPS patient who underwent thymectomy, appendectomy and an endoscopic sinus operation within one year. Diazepam, propofol, thiopental, fentanil nitrous oxide, isoflurane and vecuronium were used for general anesthetic. The patient woke from all three procedures in a short time with no problems extubating. However they do not mention whether or not the patient used baclofen or other GABAergic agents in the preoperative period.

Ledowski and Russell[3] used TIVA without neuromuscular blockers in an SPS patient undergoing an ENT operation and noted that the patient was discharged with no complications.

Regional anesthesia has been successfully used in SPS patients. Shanthanna[12] used combined spinal-epidural anesthesia for planned total knee arthroplasty in an SPS patient while Elkassabany et al.[13] used a paravertebral block for an inguinal hernia operation. However, they emphasized that as needle pain, fear and anxiety may trigger spasms it is important that detailed disclosure be made in the preoperative period and sufficient sedation be ensured during the procedure.

The literature shows that suitable conditions for tracheal intubation under general anesthetic can be provided without the use of neuromuscular agents.[14] A combination of hypnotic and opioid drugs was used. Though chest wall rigidity may be linked to high opioid doses especially in conscious patients[15] we encountered no such problem with our patient.

Though the mechanism is unclear the use of neuromuscular blockers and volatile anesthetics may cause prolonged hypotonia in SPS patients. We believe the TIVA technique provides suitable general anesthesia in these patients without the use of neuromuscular blockers.

Conflicts of interest

The authors declare no conflicts of interest.

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