Case Report

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Anesthetic Experience in a Patient with Stiff-Person Syndrome - Case Report

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ABSTRACT

Stiff-person syndrome is a rare disorder characterized by muscle rigidity and episodic spasms. In this report, we present a case of stiff-person syndrome that underwent successful general anesthesia. According to our experience, we concluded that propofol, fentanyl and atracurium should be preferred in these patients during anesthesia.

Keywords: Stiff-person syndrome, general anesthesia

ÖZET

Stiff person sendromlu bir hastadaki anestezi deneyimlerimiz

Stiff Person sendromu kas rijiditesi ve epizodik spazmlarla seyreden nadir bir hastalıktır.
Bu olgu sunumunda stiff person sendromlu bir hastadaki başarılı anestezi girişimimizi sunmayı amaçladık. Propofol, fentanil ve atrakuryumun stiff person sendromlu hastalarda güvenle kullanılabileceğini düşünüyoruz..

Anahtar kelimeler: Stiff person sendromu, genel anestezi

Introduction

Stiff-person syndrome (SPS) is a rare disorder, characterized by rigidity of the muscles in the extremities and trunk and episodic spasms. Continuous contraction of agonist and antagonist muscles causes involuntary movements. Apart from global muscle stiffness, results of neurologic examination, computed tomography, and magnetic resonance imaging of the brain are normal.

The etiology of SPS is unknown; however, in most patients titers of anti-glutamic acid decarboxylase (GAD) antibodies are found to be high. GAD antibodies have been associated with decrease of GABA A receptor-associated protein; hence the stability and the surface expression of the GABA A receptor decreases (1). A reduction in brain levels of GABA has been demonstrated with magnetic resonance imaging in patients with SPS (2).

We present a case report concerning the anesthetic experience in a woman with SPS.

Case Report

A 55-year-old woman was admitted to our neurology department with inability to walk. According to her medical records, she had been examined by psychiatry and physical therapy departments previously because of stiffness and muscle spasm. Neurological examination and cerebral computed tomography examination were normal. Because of the presence of unexplained stiffness in her previous surgery and general anesthesia, the neurology department suspected SPS, and laboratory findings were positive for GAD antibodies. Her symptoms were controlled with intrathecal baclofen and diazepam (50 mg/day), but her left leg became rigid from the pelvic region. The patient was recommended for orthopedic surgery and scheduled for a release operation.

On the day of surgery, 5 mg diazepam premedication was given. The operating room was quiet and heated before the patient's arrival. Anesthesia was induced with propofol (2 mg/kg), and fentanyl (1 µg/kg). After administration of atracurium

(0.3 mg/kg), the trachea was intubated and anesthesia was continued with sevoflurane (1-2 vol%) and oxygen/N2O mixture (50:50). In the following 2.5 hours, fentanyl (50 µg) was administered. After discussion with the orthopedic surgeon we were quite convinced that muscle tension was adequate for release surgery. At the end of the procedure, the patient, who had a TOF ratio of 4/4 and a tidal volume of 5 ml/kg, was extubated without problem. In the operation, M. tensor fasciae latae and M. gluteus medius were released by the orthopedic surgery team. After this operation and uneventful recovery, the patient was referred to the physical therapy department, and 2 months later, she started to walk without a walker or a cane.

Discussion

In 1956, Moersh and Woltman reported a series of 13 patients with progressive fluctuating muscular rigidity and spasm. They called this entity the stiff-man syndrome, but today's authors prefer to call it stiff-person syndrome (3).

There are a number of case reports about SPS and anesthesia. Johnson and Miller observed muscle weakness in a patient with SPS when baclofen was combined with inhaled desflurane or isoflurane, vecuronium and succinylcholine (4). Obara et al. reported a patient with SPS who underwent 3 operations under general anesthesia without any problems. They used isoflurane, fentanyl and vecuronium (5). Haslam and Price used atracurium with uneventful recovery in a 60-year-old patient with SPS (6). In the case report by Bouw et al., inhaled anesthetic (isoflurane) was responsible for prolonged muscle relaxation at the end of the procedure especially when combined with baclofen (7). The authors used morphine, sufentanil, atracurium, and isoflurane. But there are not enough data available about the relationship between baclofen and neuromuscular blocking or volatile anesthetic agents.

According to our experience, we concluded that using sevoflurane, propofol, fentanyl, and atracurium for general anesthesia in SPS patients should be the preferred approach.

Contribution Categories	Name of Author
Follow up of the case	Y.C.A.
Literature review	Y.C.A.
Manuscript writing	Y.C.A.
Manuscript review and revisation	T.A.

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