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Anesthetic care of stiff person syndrome in the outpatient setting

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Abstract

Introduction: Stiff Person Syndrome is characterized as a neurological disease causing fluctuating muscle rigidity escalating to painful spasms. The presence of antibodies against glutamic acid decarboxylase, an enzyme in the pathway of the inhibitory neurotransmitter gamma-aminobutyric acid is the proposed cause. Central muscle groups are affected first but the disease can progress to include distal muscle groups yielding limited use of limbs.

Case report: A 55 year old male, with diagnosed stiff person syndrome, presented for ambulatory surgery. Obesity and parathyroid disease defined past medical history. All pre-operative laboratory testing fell within normal limits. Midazolam 2mg IV pretreatment preceded transfer to the operating room. Midazolam 2mg IV and intermittent doses of Ketamine totaling 60 mg IV over a 15 minute period maintained monitored anesthesia care. The patient remained comfortable and calm through the procedure and was transferred to the post-anesthesia care unit on nasal cannula oxygen for observation. No exacerbation of symptoms ensued.

Discussion: Many anesthetic agents are associated with enhancement of GABA-ergic inhibition making the care of a patient with stiff person syndrome challenging. Existing case reports are inconclusive in describing prolonged hypotonia following general anesthesia using inhalation gases and muscle relaxants. High stress can exacerbate an event. A previous case report showed that propofol could be used safely for a short procedure and may have the benefit of improved sequelae of stiff person syndrome following the procedure. However, because of the patients' airway ketamine was chosen. Patient comfort without residual effects of stiff person syndrome was achieved.

Keywords: Stiff person syndrome, ketamine, ambulatory surgery, ilioinguinal nerve block

Introduction

Moersch and Woltman originally described Stiff Person Syndrome in 1956 [1]. It is characterized as a neurological disease causing fluctuating muscle rigidity escalating to painful spasms. Pathogenesis has been attributed to an autoimmune disease, with loss of GABAergic interneurons in supraspinal locations [2]. The presence of antibodies against glutamic acid decarboxylase, an enzyme in the pathway of the inhibitory neurotransmitter gamma-aminobutyric acid (GABA), is the proposed cause. The lack of GABAergic tone is believed to produce painful spasms, which can develop insidiously, occur at random, or follow the insult of high stress states [3]. Central muscle groups are affected first but the disease can progress to include distal muscle groups yielding limited use of limbs [2].

Case report

A 55 year old male, with diagnosed Stiff Person Syndrome, presented for left ilioinguinal nerve block in the ambulatory surgery center secondary to chronic testicular pain attributed to stiff person syndrome. The patient was obese and had hyperparathyroidism. During airway examination he was found to be a mallampati class II but was at risk for difficult intubation secondary to body habitus and short neck. Medications included duloxetine, cyanocobalamine, diazepam, and intravenous immunoglobulin. The patient was allergic to tramadol and adhesives. Rare alcohol use defined social history. All pre-operative laboratory testing fell within normal limits. Midazolam 2mg IV pretreatment preceded transfer to the operating room. Standard ASA monitors were applied and 4 liters of oxygen via nasal cannula begun. Midazolam 2mg IV and intermittent doses of Ketamine totaling 60 mg IV over a 15 minute period maintained monitored anesthesia care. The patient remained comfortable and calm through the procedure and was transferred to the post-anesthesia care unit on nasal cannula oxygen for observation. No exacerbation of symptoms ensued.

Discussion

Many anesthetic agents are associated with enhancement of GABA-ergic inhibition making the care of a patient with Stiff Person Syndrome challenging for the anesthesia provider. Propofol being a popular drug of choice for short outpatient procedures is postulated to work through potentiation of GABA receptors. The effects of less GABA available in patients with Stiff Person Syndrome therefor may result in variable responses to drug administration. Existing case reports are inconclusive in describing prolonged hypotonia following general anesthesia using inhalation gases and muscle relaxants [4,5,6]. Ledowski *et al.*, described in a case report the successful use of total Intravenous anesthesia [7]. Shanthanna described in a case report

the successful use of combined spinal epidural [8]. Due to the rarity of the disease and the small number of case reports makes the choice of anesthetic challenging. Prior knowledge suggests that high stress can exacerbate an event [3]. It was imperative in this short outpatient procedure to ensure that the patient remained comfortable and the level of anesthetic depth enough to prohibit an exacerbating event. A previous case report showed that propofol could be used safely for a short procedure such as the one described and may have the benefit of improved sequelae of Stiff Person Syndrome following the procedure. However, because of the patient's restricted airway, ketamine was chosen. Patient comfort without residual effects of Stiff Person Syndrome was achieved, suggesting this option may be viable for future cases of Stiff Person Syndrome in the short procedure setting.

Competing interests
The authors declare that they have no competing interests.

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