Anesthetic management of a parturient with Stiff person syndrome for urgent cesarean delivery

B.T. Boettcher,[a] M. Muravyea,[a] C. Kuo,[a] C. Drexler,[a] P.S. Pagel[b]
[a]*Department of Anesthesiology, Medical College of Wisconsin, Milwaukee, WI, USA*
[b]*Anesthesia Service, Clement J. Zablocki Veterans Affairs Medical Center, Milwaukee, WI, USA*

ABSTRACT
Stiff person syndrome is a rare neurologic disorder with an estimated incidence of 1:1 000 000. The underlying pathophysiology is truncal and proximal limb muscle stiffness resulting from continuous co-contracture of agonist and antagonist muscle groups concomitant with superimposed episodic muscle spasms. Loss of gamma-aminobutyric acid-mediated inhibition creates chronic excitation manifested by tonic agonist-antagonist muscle contraction. To date, only three case reports referred indirectly to the anesthetic management of parturients with Stiff person syndrome. The authors describe their management of a parturient with Stiff person syndrome who underwent urgent cesarean delivery under epidural anesthesia.

Keywords: Stiff person syndrome; Glutamic acid decarboxylase; Gamma-aminobutyric acid; Cesarean delivery; Epidural anesthesia

Introduction

Stiff person syndrome (SPS), also known as Moersch-Woltman syndrome, is a rare neurologic disorder first described in 1956.[1] Truncal and proximal limb muscle stiffness resulting from continuous co-contracture of agonist and antagonist muscle groups concomitant with superimposed episodic muscle spasms are the primary clinical manifestations of SPS.[2] The disease has a reported incidence of 1:1 000 000, usually presents in the fourth to sixth decade of life, affects women more than men (ratio 2:1), and is temporally progressive. The diagnosis is often delayed because of the disorder's rarity and nonspecific initial presentation. Muscle spasms are debilitating, fluctuate in timing and severity, and occur spontaneously or in response to stimuli including loud noises, unexpected contact, pain and anxiety.[3]

Stiff person syndrome is probably an autoimmune disease, as two-thirds of patients have glutamic acid decarboxylase 65 isoenzyme (GAD 65) autoantibodies, but positive serology is not required for the diagnosis. Glutamic acid decarboxylase 65 mediates the rate-limiting step in gamma-aminobutyric acid (GABA) synthesis.[4] Loss of GABA-mediated inhibition creates chronic excitation manifested by tonic agonist-antagonist muscle contraction.[5] Stiff person syndrome is associated with other autoimmune disorders including type I diabetes mellitus and thyroiditis.[6,7]

The anesthetic implications of SPS have been infrequently reported.[8–13] To date, only three case reports referred indirectly to the anesthetic management of parturients with SPS.[14–16] The authors describe their management of a parturient with SPS who underwent urgent cesarean delivery under epidural anesthesia.

Case report

A 30-year-old, 171 cm, 102 kg (body mass index 34.9 kg/m^2), nulliparous patient with SPS was admitted at 24 + 3 weeks of gestation for observation because of intrauterine growth restriction and hypertension with concerns for preeclampsia. The initial treatment plan included fetal monitoring, a course of betamethasone for fetal lung maturation, and magnesium for neuroprotection if delivery was imminent. A 24-hour urine protein sample supported the diagnosis of preeclampsia. The patient also had essential hypertension treated with labetalol, and the dose was increased because her arterial blood pressure was elevated on admission (150–160/80–90 mmHg).

Stiff person syndrome was diagnosed at the hospital's neuromuscular disease clinic approximately six months before the current admission. The patient described life-long stiffness that had become progressively worse and contributed to numerous falls. She reported frequent muscle "contractures" and "locking-up" of her lower and upper extremities, often without apparent cause. Her spine was relatively spared of these symptoms. She could not perform fine motor skills and

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Correspondence to: Brent T. Boettcher DO, Department of Anesthesiology, Medical College of Wisconsin, 8701 Watertown Plank Road, Milwaukee, WI 53226, USA.
E-mail address: bboettcher@mcw.edu

required assistance for many activities of daily living. Serologic assay for anti-GAD 65 antibodies was negative. The patient was attempting to conceive during the initial neurologic evaluation and as a result, refused treatment with benzodiazepines and immunoglobulin due to concerns of fetal toxicity.

The patient had type I diabetes mellitus initially diagnosed at age 15, which was managed with a subcutaneous insulin pump. Her diabetes was well controlled (HbA1C 5.2%). A previous gastric emptying study identified gastroparesis, but the patient denied symptoms of aspiration or regurgitation. She reported blunted awareness to hypoglycemic events and a stocking-and-glove peripheral neuropathy. Her renal function was normal. She had asthma requiring occasional albuterol use, allergies to several medications including sodium citrate and citrate fruits (severe dermal and gastrointestinal tract ulcers), and intolerance to metoclopramide.

Physical examination revealed bilateral flexion contractures of the fingers, elbows and shoulders. The patient's lower extremities remained extended and abducted in the sitting position on an examination table. Her motor strength was intact throughout and reflexes were normal bilaterally. Exaggerated lumbar lordosis and neuraxial stiffness were absent. A Mallampati grade II airway examination with normal thyromental distance was observed. Anesthesia records from our institution documented that a grade 1 direct laryngoscopy view was present during previous tracheal intubations.

The obstetrical and anesthesiology teams, a neurology consultant and the patient concurred that cesarean delivery was most appropriate because the possibility for lower extremity muscle spasms during induction of labor made vaginal delivery inadvisable. In-depth discussions were conducted between the patient and the care team about her anesthetic and surgical options. Considering the patient's SPS and preeclampsia, a multidisciplinary consensus decision was reached to provide slowly titrated epidural anesthesia for cesarean delivery. The patient and the consultants agreed that general anesthesia would be used in the event that emergent delivery was required.

Twelve days after admission, a cesarean delivery was urgently scheduled because of recurrent fetal heart rate decelerations. In addition, the patient's hypertension had become more refractory to treatment, and reversal of end-diastolic umbilical arterial blood flow velocity was observed. The patient received famotidine 20 mg, and her insulin infusion pump was temporarily discontinued. The patient was taken to the operating room, and with the obstetricians present, the patient was positioned in the right lateral position before lumbar epidural catheter placement. A fluid bolus of lactated Ringer's solution was administered. Good communication before and during the procedure reduced the patient's anxiety and minimized the risk of stimulated muscle spasms. Fetal heart rate was continuously monitored; conversion to general anesthesia was planned if fetal heart rate decelerations recurred. Moderate sedation was not used to avoid neonatal respiratory depression. A 17-gauge Tuohy needle was inserted at the L4–5 interspace after local anesthetic infiltration and the epidural space was identified on the first attempt using saline for loss of resistance. A 20-gauge multi-orifice epidural catheter was inserted 6 cm into the epidural space. The patient was then positioned supine with left uterine displacement. An epidural test dose (1.5% lidocaine with 1:200 000 epinephrine 3 mL) was negative for tachycardia, transient neurologic findings (tinnitus, perioral numbness) and lower extremity weakness. A total of 20 mL of 2% lidocaine with 1:200 000 epinephrine and fentanyl 5 μg/mL was administered in 5 mL increments over 15 min to achieve a T6 block as determined by sensory dermatomal pinprick assessment. The patient did not develop muscle spasms and the fetal heart rate remained reassuring throughout the procedure.

The uneventful cesarean delivery was performed through a classical incision. A 430 g male infant was delivered with Apgar scores of 4, 5, and 6 at 1, 5, and 10 min, respectively; the baby was transferred to the neonatal intensive care unit for further care. The mother received two 5-mL epidural doses of 0.5% bupivacaine 45 min after the initial block because of the duration of the procedure. She also received intravenous fentanyl 50 μg because of discomfort during removal of the placenta and manual exploration of the uterine cavity to evaluate a small area of hemorrhage noted on the superior aspect of the placenta. A 20 U/L oxytocin infusion was started after the placenta was delivered. The patient remained hemodynamically stable and did not suffer muscle spasms during the operation. As is our standard practice, we discussed neuraxial opioids or patient-controlled analgesia as options for postoperative pain control with the patient before surgery. She preferred and received patient-controlled analgesia (intermittent bolus hydromorphone). Intravenous ketorolac and oral acetaminophen were used as adjunctive medications for postoperative analgesia. The patient had no further SPS symptoms or muscle spasms throughout her hospitalization. The remainder of her course was uncomplicated, and she was discharged on the fourth postoperative day. The premature infant had a prolonged intensive care unit stay, but was eventually discharged home.

Discussion

We chose epidural anesthesia (lidocaine with epinephrine and fentanyl) for our patient with SPS and preeclampsia undergoing urgent cesarean delivery because general anesthesia may be associated with delayed emergence and prolonged muscle weakness in patients with SPS. This approach provided suitable