Case Report

Segmental Thoracic Spinal Anesthesia- An Alternative Anesthesia Technique for Cholecystectomy in a Pre-Diagnosed Case of Congenital Adrenal Hyperplasia: A Case Report

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ABSTRACT

Introduction: Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder of defective steroidogenesis. Perioperative care of the patient with CAH should be focused on concerns related to deficiency and excessive production of specific hormones secreted by the adrenal cortex as well as on chronic steroid therapy. Cholecystectomy can be done under various anesthetic techniques; however, the administration of general anesthesia (GA) is the most common of them. Open cholecystectomy has been successfully managed under regional anesthesia. Surgery and anesthesia-related stress response could be responsible for the unsatisfactory outcome in patients with CAH in which cortisol secretion is already hampered. Thus, modulation and reduction of this stress response during the perioperative period can significantly alter postoperative outcomes in such patients. The choice of anesthetic technique with minimal provocation of the stress response is the mainstay of using thoracic segmental spinal anesthesia for cholecystectomy in this patient. This case report describes the successful anesthetic management of an adult patient with CAH under thoracic segmental spinal anesthesia posted for open cholecystectomy.

Key Words: Cholecystectomy, Cortisol, Congenital adrenal hyperplasia, Segmental thoracic spinal anesthesia.

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INTRODUCTION

Congenital adrenal hyperplasia (CAH) is an autosomal recessive disorder of defective steroidogenesis, most commonly due to 21-hydroxylase deficiency resulting in the variable deficiency of the end products cortisol and/or aldosterone[1]. Impaired synthesis of cortisol leads to chronic elevation of adrenocorticotropic hormone (ACTH) via a negative feedback mechanism causing overstimulation of the adrenal cortex, which stimulates excessive adrenal androgen production[2]. The signs and symptoms of CAH are mainly due to adrenocortical insufficiency, hyperandrogenism, and side effects of long-term exogenous steroid therapy used in the treatment of the condition.

Cortisol, the primary stress hormone, manages metabolism, immune response, and inflammation, and triggers the fight-flight response. Surgery and anesthesia both activate the systemic stress response, including increased cortisol production[3]. In patients with CAH, cortisol secretion may fail in response to surgical stress response. A severe deficiency of cortisol at the time of stress can cause life-threatening low blood pressure, low blood glucose, hyponatremia, and hyperkalemia[4]. Cholecystectomy can be done under various anesthetic techniques; however, the administration of general anesthesia (GA) is the most common of them. Open cholecystectomy has been successfully managed under regional anesthesia[5].

The stress response is a significant risk factor that can be responsible for unsatisfactory outcomes in patients with known metabolic disorders[6]. Thus, modulation and reduction of this stress response during the perioperative period can significantly alter postoperative outcomes in such patients. The choice of anesthetic technique with minimal provocation of the stress response is the mainstay of using thoracic segmental spinal anesthesia for cholecystectomy in this patient. This case report describes the successful anesthetic management of an adult patient with CAH under thoracic segmental spinal anesthesia posted for open cholecystectomy.

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CASE REPORT

A 30-year-old female patient weighing 62kg was scheduled for open cholecystectomy. The patient was diagnosed with congenital adrenal hyperplasia due to 21-hydroxylase deficiency at age 3 when she first visited the hospital to evaluate and treat an abnormally enlarged clitoris. For that, she underwent a reduction clitoroplasty under general anesthesia. She was on irregular follow-ups. Due to the relative lack of medical care, she had received, previous laboratory values and medical records were unavailable. Her medication included a tablet of prednisone 10mg orally once a day for the last three years. Her preoperative physical examination was notable for extensive masculinization but the systemic examination was unremarkable. Preoperative serum cortisol level was 2mcg/dl, lower than the normal limits. All other laboratory investigations complete blood count, serum electrolytes, random blood sugar, and serum creatinine were within normal limits. Premedication was done with a tablet of alprazolam 0.25mg, a tablet of ranitidine 150mg, and a tablet of prednisone 10mg one night before the surgery. Preoperative hemodynamic parameters were a heart rate of 92 beats per minute, non-invasive systolic blood pressure of 123/66mmHg, respiratory rate of 14 breaths per minute, and oxygen saturation of 99% on room air.

Informed written consent was taken. The patient was kept nil per oral for 8 hours for solid and 2 hours for clear fluid before surgery. In the operation theatre, intravenous cannulation was established with an 18G cannula. All standard monitors were attached. The patient was given an intravenous (IV) injection of ranitidine 50mg, an injection of ondansetron 4mg IV, and an injection of hydrocortisone 100mg IV. The patient was made to sit on the operation table with their elbows resting on their thighs to achieve flexion of the spine. Segmental thoracic spinal anesthesia was given through a midline approach under all aseptic conditions using 1ml of isobaric ropivacaine 0.75% mixed with 25µg of fentanyl at T10-11 interspace with a 26-gauge spinal needle after confirming its placement by the free flow of clear cerebrospinal fluid. The patient was placed in the supine horizontal position, and the level of sensory blockade was assessed by the pinprick method every minute. The level of sensory blockade from T 4-L1 was achieved in the 7th minute. The patient was able to flex his knees slightly and move his foot but straight leg raising was not possible. The surgery lasted for 1 hour and went uneventful. Throughout the surgery, all hemodynamic parameters were stable. There was no bradycardia, hypotension, or desaturation episode in the intraoperative and preoperative periods. The sensory blockade started regressing after 2 hours and the complete regression was after 3 hours. Injection paracetamol 1 gram IV infusion was given for pain management every 8 hours in the postoperative period. The patient was advised for an evening dose of 100mg hydrocortisone injection IV followed by oral prednisone 10mg from the next day once oral ships started. The patient was made to ambulate on the same day and urinary catheterization was not needed. Serum electrolytes and blood sugar levels checked postoperatively were found within normal limits. An inquiry about post-dural puncture headache 24 hours after surgery was made but our patient did not experience it.

DISCUSSION

Congenital adrenal hyperplasia refers to a group of genetically determined disorders that affect one of the enzymes that take part in cortisol synthesis inside the adrenal cortex. In normal healthy individuals, total serum cortisol ranges from 5-23mcg/dl in the morning hours and 3-13mcg/dl in the evening[7]. The treatment of CAH includes exogenous steroid therapy to replace the low level of hormone and to suppress the over-secretion of ACTH thereby reducing virilization. The amount of exogenous steroid used for the replacement therapy in CAH is not like the amount of physiological secretion of cortisol. During surgery like stressful situations, supraphysiological doses of steroid need to be administered to avoid the adrenal crisis[8]. As our patient was already having low serum cortisol levels despite daily prednisolone dosing, the risk of getting an adrenal crisis in the perioperative period would be very high. Accordingly, we administered 100mg of IV hydrocortisone at the start of surgery and the evening of the first postoperative day followed by a prednisolone daily routine dose from the next postoperative dose with the start of oral intake. The patient was closely monitored in the postoperative period in terms of hypotension and bradycardia as the patient would be at risk of adrenal insufficiency in the perioperative period also.

Most of the previous literature is focused on the choice of GA and adjustment in perioperative steroid therapy in the management of the patient with CAH posted for any surgery[9,10]. The goal of anesthetic management in this patient was to minimize the stress response caused by the anesthetic technique. This case report demonstrates that segmental thoracic spinal anesthesia is a reasonable alternative to the use of conventional GA during open cholecystectomy in patients with pre-existing complications, even though it is not frequently used, it has been demonstrated to provide better patient hemodynamic stability, lowers the incidence of the stress response, postoperative pulmonary problems and lessen the other side effects of GA. Segmental thoracic epidural anesthesia might be another anesthetic alternative for the management of such patients but difficulties associated with needle insertion, uncertain and imprecise placement of catheter particularly in the high and mid thoracic epidural space, and high risk of spinal cord injury would complicate its use.

Segmental thoracic spinal anesthesia has some limitations when used for abdominal surgeries like the risk of spinal cord injury, high spinal anesthesia level, and patient discomfort. The scenario detailed in this case report can assist anesthesiologists in broadening their horizons when treating complex cases, even though each patient is unique and there are many anesthetic options for addressing traditional instances.

CONFLICT OF INTERESTS

There are no conflicts of interest.

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