Epidural Anesthesia for a Patient with Acute Idiopathic Pandysautonomia

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In patients with autonomic dysfunction, the cardiovascular instability is a common problem during anesthesia. A significant decrease in blood pressure without a compensatory tachycardia following thiopental induction or inhalation of volatile anesthetics has previously been reported to occur in patients with dysautonomia[1,2], Shy-Drager syndrome[2,3], or familial dysautonomia[4]. Because these patients already have sympathetic denervation, it is assumed that regional anesthesia may not cause any further cardiovascular perturbation due to sympathetic blockade. Therefore, we speculate that regional anesthesia would provide an advantage such as cardiovascular stability in these patients during anesthesia. To date, however, there has been no report describing epidural anesthesia for patients with dysautonomia. We describe a case in which a 37-year-old patient with acute idiopathic pandysautonomia (AIPD)[5,6] had a stable anesthetic course with thoracic epidural plus light general anesthesia.

Case Report

A 37-year old woman, 39 kg, was admitted to the hospital with a renal stone, and scheduled to undergo the renal lithotomy. She had a history of orthostatic syncope, and numbness of face, fingers, and toes. And she was found to have photophobia, hypesthesia of the oral cavity and tongue, dullness of extremities, and absence of sweating. The diagnosis of acute idiopathic pandysautonomia (AIPD) had been made during a previous admission at her age of 36 following several autonomic function tests including Valsalva maneuver, mental arithmetic test, cold pressor test, hyperventilation test, direct sweat test with pilocarpine and reflex sweat test. She also demonstrated marked increase in blood pressure (from 116 mmHg to 161 mmHg systolic) during the infusion of small dose of norepinephrine (0.1 μg/kg/min). Biopsy her sural nerve showed axonal degeneration. One year after she showed clinical recovery except for persisting orthostatic hypotension.

Pre-operatively, her laboratory data, chest X-P and electrocardiogram were within normal limits, however, she demonstrated a significant orthostatic hypotension (120 mmHg systoric during supine position to 70 mmHg by standing). She did not receive premedication. Her supine preoperative blood pressure was noted to be 94/50 mmHg.

On the arrival in the operating room an intravenous cannula for iv infusion and a radial arterial cannula for continuous measurements of blood pressure were inserted under local anesthesia. Pressure-inducing maneuver such as coughing[7], produced no apparent alteration in R-R interval and a Valsalva maneuver showed a response with no overshoot in blood pressure and no bradycardia after the

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release of the maneuver.

After sterile prepartion and draping of the patient in the right lateral decubitus position, 2 ml of 1% lidocaine was injected intradermally and subcutaneously at the T10–11 intervertebral space. Then a 16-gauge Tuohy needle was inserted into the epidural space using the hanging-drop technique and an epidural catheter was inserted for continuous blockade. She then was placed in the supine position and 10 ml of 1.5% lidocaine without epinephrine was injected through the catheter. After the injection of anesthetic into the epidural space, her blood pressure decreased gradually from 96/64 to 86/50 mmHg during the first 20 min but further decrease was not observed and heart rate was stable around 90 beats per min (BPM). The analgesic level of T4–12 was obtained at 10 min after the epidural injection. Pressor-inducing maneuvers described above were carried out again and revealed the same results. To minimize the stress due to the kidney position, we induced general anesthesia with 5 mg of diazepam and 100 μg of fentanyl. Because she still responded to verbal commands, 100 mg of thiopental was then administered additionally. Her blood pressure fell transiently from 88/46 to 80/40 mmHg with no alteration of heart rate. Pancuronium bromide, 4 mg, was injected to facilitate insertion of an endotracheal tube. She was then ventilated mechanically by a mask with N_2O 67% and O_2 33% for 5 min and endotracheal tube was placed without additional intravenous agent. After tracheal intubation, blood pressure increased slightly from 88/40 to 94/56 mmHg, but heart rate was almost fixed at 92 BPM.

During the operation lasting for 60 min, blood pressure was almost stable between 120/80 and 86/42 mmHg and did not show any further decrease in spite of the repeat injection of 1.5% lidocaine 6 ml and 4 ml at 45 min and 90 min after the initial dose, respectively. Heart rate decreased slightly from 84 to 78 BPM and remained constant at around 78 BPM throughout the operation. She received an epidural injection of morphine hydrochloride, 2 mg, diluted with normal saline 5 ml for postoperative pain relief at 15 min before the surgical termination. Neuromuscular blockade was reversed by 2 mg of neostigmine following 1 mg of atropine at the end of the operation. The patient soon regained her spontaneous respiration, thereafter her trachea was extubated. Neither atropine nor neostigmine altered heart rate which remained constant at 82 BPM. Fifteen minutes after the extubation, her blood gas analysis showed pHa of 7.42, Pa_{CO_2} of 46.2 mmHg, Pa_{O_2} of 271.8 mmHg, base excess of 4.2 mEq/L under spontaneous breathing with a oxygen mask. The postoperative analgesic level, which determined 35 min after the last injection of lidocaine, was between T10 and L1. Her body temperature decreased from 36.3°C to 35.8°C during the surgery. Postoperatively, changes of her vital signs were carefully observed and she made an uneventful recovery.

Discussion

This patient had a well documented autonomic failure associated with severe orthostatic hypotension, absence of cardiovascular reflexes and sweating, and hyperreactivity to exogenous norepinephrine. Since central neurologic disorder was absent with this patient, she was diagnosed as acute idiopathic pandysautonomia (AIPD). The above mentioned defects would induce serious difficulties and complications during anesthesia and surgery. Because of the absence of autonomic responses, the induction of general anesthesia with thiopental or inhalation of potent volatile anesthetics produces profound fall in blood pressure without a compensatory tachycardia[1–4]. The clinical evaluation of the depth of anesthetia in a patient with autonomic dysfunction is difficult to assess because the clinical signs such as sweating, tachycardia, and decrease or increase in blood pressure are lacking. Thus profound cardiovascular perturbation and accidental overdose of anesthetics can be result in some instances. Furthermore, decrease in the respiratory sensitivity to carbon dioxide accumulation seems to exist in these particular patients during general anesthesia, especially in using enflurane[1]. Therefore, selection of anesthetic agent could be important for this particular patient.

The anesthetic choice, between general and regional, for patients with autonomic dysfunction has been discussed previously. Cohen et al.[3] selected epidural anesthesia with a reason that a partially "sympathectomized" person would have less hypotension with this technique than would a normal individual. However, because they ultimately failed to obtain satisfactory analgesia for surgery, certain advantages of epidural anesthesia might not be elucidated from their report. Recently, Beilin et al.[8] reported successful anesthetic management in patients with familial dysautonomia by moderate-dose fantanyl (50–1100 μg) anesthesia. While this technique could provide cardiovascular stability, necessity of subsequent postoperative ventilatory support lasting for hours (from 2 to 14 hours) may be disadvantageous. In our present case, although we used supplementation of nitrous oxide and small dose of fentanyl anesthesia to minimize the stress due to the kidney position, the anesthetic technique with thoracic epidural anesthesia was satisfactory in providing cardiovascular stability during the surgery.

Bevan[9] argued that the choice between general and regional anesthesia was less important than adequate cardiovascular monitoring and maintenance of blood pressure with adequate amount of intravenous fluids. Bevan[9] also suggested that the fall in blood pressure during anesthesia of patients with autonomic dysfunction was caused by the decrease in venous return due to intermittent positive pressure ventilation (IPPV). In our present case, the commencement of IPPV did not produce remarkable change in blood pressure, however, IPPV combined with inhalation of volatile anesthetics may cause profound decrease in blood pressure which needs the support of pressor agent such as phenylephrine or dopamine[3,4].

The absence of clinical signs of light anesthesia such as sweating, tachycardia, or dilated pupils make it difficult to evaluate the depth of anesthesia during surgery. In this instance, epidural anesthesia can provide for patients adequate analgesia which is confirmed before initiation of surgical stimulation. We used a non-depolarizing relaxant, pancuronium, to facilitate tracheal intubation, in view of possible implications of marked hyperkalemia or hyperpyrexia following succinylcholine use in patients with neurologic disease[10,11]. After the administration of pancuronium, because of its long acting property, we had to establish IPPV throughout the anesthesia with this patient. While our patient did not show decrease in blood pressure due to IPPV, spontaneous ventilation is more suitable for patients with autonomic disorder since there would be minimal decrease in cardiac output.

Epidural anesthesia, however, has been reported to suppress reflex cardiovascular function which maintain cardiovascular stability by the balance of sympathetic nerve and parasympathetic nerve activities in a moment[12,13]. Cardiac sympathectomy induced by cervical epidural anesthesia suppressed partially baroreceptor function, the responses in heart rate and blood pressure to Valsalva maneuver, coughing, swallowing and nasal stimulation by interrupting sympathetic efferent fibers innervating the heart[12,13]. In the present case, some of these test were already suppressed and those were not affected significantly by epidural anesthesia. Baroreflex sensitivities appear not to be affected by lidocaine infusion producing plasma lidocaine levels comparable with those observed during uncomplicated regional anesthesia[14]. Thus, epidural anesthesia is unlikely to produce the further deterioration in cardiovascular function even in patients with AIPD.

In summary, we have described a successful anesthesia management using epidural blockade for a patient with AIPD undergoing renal lithotomy. Epidural anesthesia may be the first choice for patients with autonomic dysfunction, because of pre-existing sympathetic denervation as well as absense of autonomic responses during surgery.

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