Anesthesia in Shy-Drager Syndrome

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The Shy-Drager syndrome (SDS) is characterizes by autonomic dysfunction, accompanied by additional central neurologic symptoms. The main clinical manifestations of the autonomic dysfunction include orthostatic hypotension, vesicorectal dysfunction, hydrohidrosis and extrapyramidal syndrome. During anesthesia, the cardiovascular instability due to autonomic dysfunction represent a potential danger. The present paper reports the anesthetic course of a patient with SDS undergoing abdonimal surgery.

Case History

A 59 year-old man (weight 59 kg, height 169 cm) was scheduled for total gastrectomy for gastric cancer. The patient had exhibited tremor, muscular rigidity and bradykinesia for 4 years prior to admission. He was treated with parasympatholytic drugs and L-Dopa until admission under a diagnosis of Parkinson's syndrome. Other neurologic symptoms consisting of a speech disorder, dizziness, dysphagia, and ataxia of the upper and lower extremities had recently become more severe. On admission, marked orthostatic hypotension was observed with blood pressure of 140/80 mmHg lying and 90/50 mmHg standing with no change in the heart rate of 70 bpm. Abscence of sweating, sexual impotence, and gait ataxia were also present. A Valsalva maneuver produced hypotension without rebound hypertension or tachycardia. Tendon reflexes were brisk and symmetric, but a right Babinski sign was present. The chest X-ray and electrocardiograph were normal and laboratory examination disclosed no abnormality. A CT scan of the brain showed mild cerebellar atrophy and suspectable olivopontocerebellar atrophy (OPCA). However, the variety of symptoms and signs including autonomic dysfunction, cerebellar ataxia and involuntary movement were most consistent with a diagnosis of SDS.

The patient was premedicated with atropine 0.3 mg and hydroxyzine 25 mg intramuscularly 60 min before induction of anesthesia. Anesthesia was induced slowly with incremental doses of thiopentone to a total of 100 mg and supplemented with pentazocine 30 mg. Tracheal intubation was facilitated with suxamethonium 40 mg. Anesthesia was maintained with 0.4–0.6% enflurane and nitrous oxide in oxygen. He was ventilated by a respirator after paralysis with pancuronium. Fifteen minutes after induction of anesthesia, the blood pressure progressively fell from 160/90 to 60/30 mmHg. This hypotension was successfully treated with rapid fluid administration and continuous dopamine in-

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fusion at a rate of 5 $\mu g \cdot kg^{-1} \cdot min^{-1}$. The pulse rate remained constant at 70 bpm throughout the episode. Subsequently, blood pressure and heart rate were stable throughout the 2 hr surgery after infusion of dopamine. The patient's hemodynamics was easily maintained in the postoperative period with conventional fluid therapy.

In addition, we measured the intraoperative levels of plasma catecholamines. The plasma catecholamine levels by high-performance liquid chromatography were determined before, during and after anesthesia. The catecholamine levels before anesthesia were 148 $pg \cdot ml^{-1}$ for noradrenaline and 49 $pg \cdot ml^{-1}$ for adrenaline, but after induction the both catecholamine levels decreased to 125 $pg \cdot ml^{-1}$ and 34 $pg \cdot ml^{-1}$, respectively.

Discussion

It is well documented that the main problem with anesthesia for patients with SDS is the maintenance of cardiovascular stability[1-4]. In our case, the arterial blood pressure decreased after induction of anesthesia and required a dopamine infusion and fluid administration.

There are many possibilities for hypotension during anesthesia for patients with SDS. These patients may have relatively fixed heart rates which are unable to increase cardiac output against stress, because it is considered to be the loss of cardiovascular reflexes which commonly compensate for physiologic disturbances. Also, the patients with SDS are extremely sensitive to anesthetic agents[4]. In our case, a small dose of thiopentone (100 mg) occurred hypotension after induction of anesthesia. Profound hypotension induced by thiopentone has previously been reported in a patient with SDS[1,4,5]. Even if the dose of the anesthetic agent used has a minimal effect on the cardiovascular system, these patients exhibit extreme sensitivity to the anesthetic agent. Thus, agents causing minimal depression of cardiac output should be chosen for induction of anesthesia. Ketamine is one of the most useful anesthetic agens in patients with SDS, because anesthetic doses usually cause a profound increase in arterial pressure due to central sympathetic stimulation and parasympathetic inhibition[7]. It has also been reported that fentanyl and N_20 anesthesia provide cardiovascular stability in patients with SDS[3,6].

The use of regional anesthesia is controversial for patients with autonomic dysfunction. The choice of regional anesthesia is not theoretical, because more severe hypotension would be commonly anticipated following either epidural or spinal anesthesia[6].

If pressor agent are necessary for maintenance of blood pressure, direct acting sympatheomimetic agents are effective and a very low dose should be used initially to avoid an unpredictable hypertensive response to peripheral vasculature.

The basal plasma catecholamine concentrations were within the normal range and did not elevate above preoperative values with induction of anesthesia. The catecholamine levels were not accurate after infusion of dopamine, because our measurement could not distinguished between noradrenaline and adrenaline, and dopamine. In patients with SDS, the basal levels of catecholamines have been reported to be within the normal range or reduced, and no change was reported during anesthesia and operation[6,9]. The suppression of catecholamine release during anesthesia may not simply reflect suppressed sympathetic function resulting from anesthesia. Stores of norepinephrine at the nerve ending may be absent or reduced[6,8,9,10]. Exercise and stress cause norepinephrine release in normal man, however, postural change

and physical activity fail to cause an increase in plasma levels of norepinephrine in patients with SDS[1,3].

In addition to profound hypotension, abnormal respiratory patterns are also an important problem. The respiratory depression have been reported in patients with SDS undergoing anesthesia[4]. Thus, extreme caution should be required with the use of some drugs which possess respiratory depressant character.

In summary, hypotension was experienced after induction of anesthesia in patients with SDS. The plasma catecholamine levels were not elevated by tracheal intubation. We recommended that an induction agent with less potent effects on the cardiovascular system than that of thiopentone should be used.

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