症 例

Shy-Drager Syndrome Operated under Epidural Anesthesia

Ryu Okutani*, Taeko Fukuda**
Kazuhide Fu*, Shigeo Maeda**

Shy-Drager syndrome (SDS) is a rare chronic progressive disease and a multiple system atrophy (MAS) which occurs in middle-aged men. This syndrome is characterized by antonomic nerve failure causing orthostatic hypotension, cerebellar disorder, extra-pyramidal symptoms and mental retardation[1]. A patient with SDS is susceptible to cardiovascular collapse during anesthesia and surgery due to the marked breakdown of autoregulation of autonomic nerve response. We report the preoperative examinations used for diagnosis of a SDS and anesthetic managements using epidural anesthesia for a SDS patient who underwent elective surgery.

A Case Report

A 50-year-old man (weight, 70kg ; height, 165cm) was scheduled for elective surgery for repair of a left inguinal hernia. At the age of 40 years', he had undergone surgery for the repair of bilateral inguinal hernia under spinal anesthesia. At that time, he had not shown any symptoms of SDS, such as wide fluctuation in blood pressure depedent upon postural change, yawning, nausea, or increased sweating. Symptoms related to SDS, sudh as orthostatic hypotension, stagger when walking, anemia, gait disturbance, and urinary incontinence, were noted during the 2 years prior to the admission for re-operative surgery of inguinal hernia at this time.

*Department of Anesthesiology, Hyogo Prefectural Nishinomiya Hospital
**Department of Anesthesiology, Hyogo College of Medicine

A standing test was performed to evaluate autonomic nerve function. His blood pressure was 134/91 mmHg (heart rate ; 70bpm) in the supine position, but fell to 60/44mmHg (heart rate ; 69bpm) after 1 min of standing, and to 42/20 mmHg (heart rate ; 70bpm) after 2 mins of standing and he lost consciousness ; the test was then terminated. Immediately after the supine position was resumed, his blood pressure recovered to 135/88mmHg, and he was regained consciousness without treatment by drug administration. On the Aschner test, no significant changes were found in his heart rate and blood pressure.

The variation or R-R interval on ECG responded to respiratory load showed no increase ; namely 1.6% at rest (normal reange, 2.7-5.1) and 1.6% during respiratory loading (normal reange, 5.2-9.0).

Plasma catecholamine levels at rest were within the normal ranges ; norepinephrine was 86 pg/ml and epinephrine 42pg/ml in plasma. No increases in these levels in response to postural change to standing form supine positon were found. The daily variation in blood pressure (circadian blood pressure pattern) was characterized by nocturnal stability and diurnal variation, while the heart rate remained unchanged even when blood pressure varied. Pressor response to various vasoconstrictive drugs was not evaluated in this patient. Pituitary, adrenal cortex, and thyroid functions were within normal limits. Based on the results of these examinations, our patient was diagnosed as having of SDS. No vasoactive or hormonal drugs

were administered preoperatively.

Anesthetic Procedure

The patient was brough to the operating room with no premedication. Pulse oximetry and electrocardiogram were placed and blood pressure was measured directly and continuously via a left radial artery catheter inserted under local anesthesia before induction of anesthesia. The variation of blood pressure due to postural change was confirmed before the start of operation in the operating room. Namely, the systolic blood pressure of 152mmHg in the supine position fell to 111mmHg by tilting the head up and to 125mmHg by taking lateral position. Heart rate, however, did not change during these postural changes (72bpm).

An epidural puncture was performed with 17G epidural needle by median approach at lumbar 2/3 interspace in the right lateral decubitus position, and then a catheter was inserted a length of 5cm to head. Analgesia from Th7 to L4 was obtained by injection of 8ml of 2% mepivacaine 10mins after the injection. Fifteen mins later, decrease in systolic arterial blood pressure from 143mmHg to 100mmHg was noted, and which was treated successfully to 134mmHg with a rapid infusion of 200ml of lactated Ringer's solution. The pulse rate remained unchanged at 72bpm throughout this episode. Additional 3ml of 2% mepivacaine was injected to epidural space 65mins after the first injection. No blood pressure changes were obseved at this time. The radical operation for inguinal hernia completed by 85mins without difficulty. The infused lactated Ringer's solution was 1100mℓ in total, urine out put was 190mℓ, and blood loss was 120g. His consciousness was consistently clear, and analgesia at the end of surgery was from Th6 to L4.

Discussion

Idiopathic orthostatic hypotension (IOH) is a progressive autonomic failure in the absence of peripheral neuropathy or neurological disordes[1)2)]. While, Shy and Drager[3)] described the distinct syndrome of orthostatic hypotension accompanied with neurologic dysfunction, termed multiple system atrophy (MSA)[4)]. Sympatho-adrenal orthostatic hypotension (SOH), which occurs in patients without neurologic or sympatho-adrenal abnormalities, is attended by marked tachycardia[5)].

While there is usually an abnormality of sympathetic nervous system function in SDS patients, therefore, one with SDS has a functional deficit in blood pressure regulation of central nerve system[6)]. This generalization is supported by the results of determination of plasma norepinephrine level[7)8)]. In our patient, the plasma catecholamine levels were in the normal range at rest, but remained unchanged on standing position.

Important aspects of the anesthetic management of SDS patients are the preoperative assessment of the degree of autonomic dysfunction and the selection of anesthetic procedure which will not induce cardiovascular collapse during anesthesia and surgery. Parameters useful in the evaluation of autonomic nerve function include performance on Shellong's tilting test (active tilting test), diurnal variation in R-R variation of R-R interval on ECG, the levels of plasma catecholamines, especially that of norepinephrine, and the levels of urinary eliminated 11-OHCS[9)].

Several anesthetic techniques have been reported for management of a patient with SDS. General anesthetics using such as halothane, enflurane[10)], sevoflurane[11)], and ketamine have been used effectively for SDS, accompanied with well controlled hemodynamics. However, some vasopressors are necesary. Moreover, cases of vocal cord paralysis in a SDS patient after general anesthesia were recently reported[12)13)]. In these cases, abductor vocal cord paralysis was presented, thus the potential for respiratory difficulty should be kept in mind after extubation. Since these patients are highly sensitive to anesthetics, respiratory depression in patients with SDS induced by general anesthesia has been reported[14)]. On the other hand, spinal or epidural anesthesia has been reported to induce marked hypotension[15)], because of the lack of compensation for cardiovascular autoregulation defect. Therefore, spinal and epidural anesthesia were

considered not to be an appropriate anesthetic method. In spite of such, the reasone we selected epidural anesthesia instead of general anesthesia for this patient was that the site of operation was in the low abdominal field. We planned that reduction of blood pressure due to epidural anesthesia would be treated by infusion of Ringer's solution and when the incresing effect of blood pressure would be insufficient by fluid infusion exogenious cathecholamines, especially, alpha-stimulant would be carefully administed. Fortunately, the lowered blood pressure was treated by volume-loading sololy.

Using of elastic leg bandages for prevention of blood pooling[16], in venous vessels, and using of vasopressin[17], L-threo-DOPES (a norepinephrine prodrug)[18], and somatostatin analog[18)19)], for increase of vascular resistance should be considered.

We experienced the anesthetic management for surgical repair of an inguinal hernia complicated with SDS syndrome which occurred 2 years before surgery. The fact that the selected epidural anesthesia did not cause problematic decrease of the blood pressure during surgery indicated that the epidural anesthesia could be one of options as anesthesia for patients with SDS syndrome.

References

1) Wagner H H. : Orthostatic hypotension. Bull Johns Hopkins Hosp **105** : 322～359, 1959
2) Bradbury S, Eggleston C. : Posturral hypotension: A report of three cases. Am. Heart J **1** : 73～86, 1925
3) Shy G M, Drager G A. : A neurological syndrome associated with orthostatic hypotension. A clinical-pathologic study. Arch Neurol **2** : 511～527, 1960
4) Bannister R, Sever P, Gross M. : Cardiovascular reflexes and biochemical responses in progressive autonomic failure. Brain **100** : 327～344, 1977
5) Demanet J C. : Usefulness of noradrenaline and tyramine infusion tests in the diagnosis of orthostatic hypotension. Cardiology **61** (Suppl) : 213～224, 1976
6) Kontos H A, Richardson D W, Norvell J E. : Norepinephrine depletion in idiopathic orthostatic hypotension. Ann. Intern. Med **82** : 336～341, 1975
7) Polinsky R J, Kopin I J, Ebert M H, et al : Pharmacologic distinction of different orthostatic hypotension syndromes. Neurology **31** : 1～7, 1981
8) Polinsky R J, Goldstein D S, Brown R T, et al : Decreased sympathetic neuronal uptake in idiopathic orthostatic hypotension. Ann. Neurol **18** : 48～53, 1985
9) Koike Y, Sakurai Y, Senda Y, et al : Diurnal variation in autonomic functions-Comparative study of Shy-Draper syndrome and normal control. Auto Nerv. System (Jiritushinkei) **26** : 531～536, 1989
10) Okuda T, Wakita K. : Anesthesia in Shy-Drager syndrome. J. Anesth **6** : 500～502, 1992
11) Fujibayasi T, Takakura Y, Kawakami H, et al : Management of sevoflurane in a patient of Shy-Drager syndrome. Jpn. J. Clin. Anesth **13** : 1423～1424, 1989
12) Drury P M E, Williams E G N. : Vocal cord paralysis in the Shy-Drager syndrome. Anaesthesia **46** : 466～468, 1991
13) Kew J, Gross M, Chapman P. : Shy-Drager syndrome presenting as isolated isolated paralysis of vocal cord abductors. Br. Med. J **300** : 1441, 1990
14) Sweeney B P, Jones S, Langford R M. : Anaesthesia in dysautonomia: further complications. Anaesthesia **40** : 783～786, 1985
15) Stirt J A, Frantz R A, Gunz E F, et al : Anesthesia, cathecholamine, and hemodyanamics in autonomic dysfunction. Anesth. Analg **61** : 701～704, 1982
16) Harioka T, Miyake C, Toda H, et al : Anesthesia for Shy-Drager syndrome; Effects of elastic bandage, phenylephrine, and IPPV. Jpn. J. Anesthesiol (Masui) **38** : 801～804, 1989
17) Bevan D R. : Shy-Drager syndrome: A review and a description of anaesthetic management. Anaesthesia **34** : 866～873, 1979
18) Sakoda S, Suzuki T, Higa S, et al : Treatment of orthostatic hypotension in Syh-Drgper syndrome with D, L-threo-3, 4-dihydroxypenylserine: A case report. Eur. Neurol **24** : 330～334, 1985
19) Weiss M, Shaeffer Z, Eizenstein Z. : Postural hypotension; Pressor effect of octreotide not mediated by norepinephrine. South. Med. J **83** : 1315～1317, 1990 (Circ Cont **15(2)** : 290～292, 1994)