Anesthetic Management of a Patient with Tuberous Sclerosis for Selective Arterial Embolization

- A case report -

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A 39-yr-old female patient, diagnosed with tuberous sclerosis 10 years ago, presented for selective arterial embolization under general anesthesia. The symptoms of the patient were lower abdominal pain and gross hematuria. Renal arteriography showed multiple bilateral renal angiomyolipomas and ruptured aneurysms in right kidney. She had also pulmonary lymphangiomatosis, hepatic angiomyolipomas, and multiple subependymal nodules without mass effect in brain. We report the anesthetic management of this case with tuberous sclerosis for selective arterial embolization under general anesthesia. (Korean J Anesthesiol 2005; 49: S 47-50)

Key Words: anesthesia, angiomyolipoma, selective arterial embolization, tuberous sclerosis.

Tuberous sclerosis (Bourneville’s disease) is a genetic disorder with an overall prevalence of about 1 in 29000, 1 in 15000 for those less than 5 years of age, and a birth incidence as high as 1 in 10000, making it one of the most common autosomal dominant disorders.1) The clinical expression is so variable that relatively mild forms of the disease may be difficult to recognize.

Renal involvement in individuals with tuberous sclerosis includes angiomyolipomas, which are considered pathognomonic when they are multiple and bilateral.2) Normal parenchymal preservation can be accomplished by selective arterial embolization. Anesthetic management may be influenced by the presence of cardiovascular, neurological and renal involvement of tuberous sclerosis. The anesthetic management of patients with tuberous sclerosis has not been reported in Korea. Therefore, we report a case of anesthetic management of a patient with tuberous sclerosis for selective arterial embolization.

CLINICAL FEATURES

A 39-yr-old woman with tuberous sclerosis, diagnosed 10 years previously, was admitted to our hospital due to sudden onset of lower abdominal pain and hematuria. She was not under regular medical treatment. Her siblings were not examined but are reported to be in good health. Her son was diagnosed with tuberous sclerosis. Physical examination revealed cutaneous evidence of tuberous sclerosis (adenoma sebaceum, cervical skin tags, and subungual fibromas). Her muscle tone and reflexes in all limbs were normal. Laboratory studies revealed hemoglobin of 8.6 g/dl, normal urea and electrolytes, and packed RBCs in urinalysis.

A computed tomography of the abdomen and pelvis was performed, and angiomyolipoma and hemangioma in the liver and bilateral renal angiomyolipomas were found (Fig. 1). Renal arteriography was performed under local anesthesia (Fig. 2). The radiologist tried embolization of the drainage vessels by coils and large sized gelform particles, but embolization material is drained by means of the draining veins due to their large size.

The electrocardiography showed QT prolongation. The 2-D echocardiography showed that the ejection fraction was 60-65%
and estimated pulmonary arterial pressure was about 60 mmHg. A chest radiograph showed non-specific finding, and arterial blood gas analysis while administering oxygen 2 L/min with nasal prong showed pH 7.45, PaO$_2$ 83.4 mmHg, PaCO$_2$ 30.5 mmHg, SaO$_2$ 96.6%, HCO$_3$ 20.7 mmol/L, and chest computed tomography revealed lymphangiomatomatosis in tuberculous sclerosis. Brain magnetic resonance of imaging showed subependymal nodules associated with tuberculous sclerosis.

Selective arterial embolization was scheduled to stop right renal bleeding. Atropine 0.5 mg and midazolam 3mg was administered intramuscularly for premedication 30 minutes before general anesthesia. On arrival in the angiography room, ECG monitoring, pulse oximetry, and non-invasive blood pressure were established. Blood pressure was 220/120 mmHg and heart rate 70 beats/min. Anesthesia was then induced with fentanyl 100 mcg, propofol 120 mg, lidocaine 60 mg, and atracurium 25 mg intravenously. After loss of eyelash reflex, isoflurane 1.5% in oxygen 5 L/min was administered via a facial mask. Direct laryngoscopy was performed 3 min after administration of atracurium. Ventilation was controlled artificially with 67% oxygen and 33% nitrous oxide so that the peak inspiratory pressure remained below 15 cmH$_2$O and the end tidal carbon dioxide concentration was maintained 35 to 40 mmHg, and SaO$_2$ was maintained at 98-99%. Arterial catheter was inserted into left radial artery, and a pulmonary artery catheter (Swan-Ganz CCOMbo, Edwards®, USA) was inserted into the right internal jugular vein.

During anesthesia, esmolol was administered to maintain systolic blood pressure 150 to 170 mmHg. CVP was maintained 6 to 10 mmHg. Pulmonary arterial pressure was 50 to 55 mmHg, and pulmonary arterial occlusion pressure was 16-22 mmHg. Arterial blood gas analysis in 67% oxygen inhalation showed pH 7.32, PO$_2$ 122.9 mmHg, PCO$_2$ 39 mmHg, SaO$_2$ 98.3%, HCO$_3$ 19.5 mmol/L.

Embolization was performed with the use of metallic coils and ethanol mixed with iodized oil (Lipiodol). Temporary balloon

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**Fig. 1.** Abdominal computed tomography showing multiple bilateral renal angiomyolipomas.

**Fig. 2.** Right renal angiography before selective arterial embolization. There are two aneurysms in mid and lower portion, and they showed dauer sacs, suggested as ruptured aneurysms.

**Fig. 3.** Right renal angiography after selective arterial embolization. There is devascularization of the tumors with embolization using coils and ethanol/Lipiodol mixture.
occlusion catheter was located on the proximal portion of the aneurysm feeding artery. Ethanol/Lipiodol was injected until the drainage vein was opacified under occlusion balloon inflation. After the injection, the balloon was left inflated for approximately 3 minutes. Postembolization angiography showed the complete embolization of aneurysms initially. But a few minutes later, recanalization of aneurysms was visualized with decreased feeding arteries diameter than initial evaluation, so the radiologist used metallic coils to embolize two intratumoral aneurysms in this patient, and thus satisfactory stasis was achieved (Fig. 3). After embolization, we moved the patient to the recovery room, and observed her while administering oxygen 5 l/min with facial mask. The patient complained of dyspnea, and postoperative arterial blood gas analysis while administering oxygen 2 L/min with nasal prong showed pH 7.40, PO2 77.4 mmHg, PCO2 32.1 mmHg, SaO2 95.1%, HCO3 19.5 mmol/L. Two days later, the symptoms of this patient (gross hematuria, abdominal pain) were relieved. Dyspnea had disappeared and arterial blood gas analysis in room air showed pH 7.44, PO2 111.6 mmHg, PCO2 22 mmHg, SaO2 98.4%, HCO3 14.5 mmol/L.

**DISCUSSION**

We describe a woman of 39 years with tuberous sclerosis. This case represents the anesthetic management with tuberous sclerosis for selective arterial embolization.

Tuberous sclerosis is a systemic disorder characterized by benign hamartomatous tumors that involve multiple organ systems. Although traditionally recognized as a neurological and dermatological manifestation, renal disease is a frequent manifestation of tuberous sclerosis and the leading cause of death in adults with tuberous sclerosis. Renal involvement in patients with tuberous sclerosis begins at infancy, and the most common lesion is angiomylipoma.

Our patient had no history of seizure and had normal intellect, but brain magnetic resonance of imaging showed subependymal nodules. Roach et al. reported that neurological presentation of tuberous sclerosis occurs typically in children with seizures and intellectual impairment. However approximately 50% of patients who fulfill the diagnostic criteria have normal intellect and 15% remain free from seizures.

Cardiac rhabdomyoma, although rare, is the commonest benign cardiac tumor associated with tuberous sclerosis. Obstruction of the left ventricular flow tract may lead to cardiac failure and arrhythmias such as Wolff-Parkinson-White syndrome. The 2-D echocardiography of the patient showed no cardiac tumor, normal cardiac function, and pulmonary hypertension.

Lung involvement due to tuberous sclerosis was present in our patient, although it is considered rare (< 1%). In this case, the patient had a pulmonary lymphangiomyomatosis in the chest computed tomography and a relatively high (67%) concentration of inspired oxygen was used to maintain oxygen saturation.

We monitored pulmonary artery pressure and pulmonary artery occlusion pressure because the patient had a high estimated pulmonary artery systolic pressure (60 mmHg) and there was the risk that systemic vascular absorption of ethanol may have induced pulmonary vasoconstriction. Ethanol has been used as an angiographically administered agent for vascular embolization of tumors and anesthesiologists should be aware of the possibilities of systemic hypotension, increased pulmonary vascular resistance, and myocardial toxicity under general anesthesia.

The anesthetic management of the patient with pulmonary hypertension are avoidance of increases in pulmonary arterial pressure and pulmonary vascular resistance, avoidance of changes in right ventricular preload, and maintenance of left ventricular afterload and right ventricular contractility. Propofol has been used as an induction agent without problems in patients with pulmonary hypertension, and isoflurane also can be used to maintain anesthesia in patients with pulmonary hypertension. Treatments of pulmonary hypertension during anesthesia include inhaled nitric oxide, milrinone, dipyridamole, and inhaled prostaclin.

The kidneys can be involved in three different ways in tuberous sclerosis: angiomyolipomas, cystic disease, and renal cell carcinomas. The typical angiomyolipoma is a mixture of smooth muscle, fat, and abnormal blood vessels, the relative proportions of which vary widely. Dysmorphic vessels in angiomyolipomas can have microaneurysms or macroaneurysms. These aneurysms may rupture and bleed resulting in significant morbidity with pain, loss of renal unit, subsequent renal failure and possibly death, although angiomyolipomas do not cause renal failure by themselves. Selective arterial embolization was performed to control acute hemorrhage temporarily, and has been used with increasing frequency to manage not only symptomatic tumors but also asymptomatic tumors with the risk of hemorrhage.

We suggest that anesthesiologists have to be aware of the clinical features of tuberous sclerosis and all the organs that could be involved, such as the brain, skin, heart, eyes and kidneys, to ensure appropriate monitoring for complications.
REFERENCES