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Anesthetic management of a patient with McCune-Albright syndrome accompanied by polyostotic fibrous osteodysplasia

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Introduction

Patients with McCune-Albright syndrome (MAS) have polyostotic fibrous dysplasia, dermal pigmentation (café-au-lait spots), and endocrine abnormalities, including sexual precocity, hyperthyroidism, Cushing's disease, acromegaly, hyperprolactinemia, and hyperparathyroidism [1]. Recently we conducted emergency anesthesia on a patient with MAS who had sexual precocity, osteodysplasia, and acromegaly. Because of their religious beliefs, her parents refused to allow her to receive blood transfusions.

Case report

A 16-year-old girl, 148 cm tall and weighing 42 kg, required emergency hospitalization for acute abdomen. The gynecological diagnosis was an ovarian hemorrhage requiring immediate surgery. On admission, the patient was found to have a macrosomial face, with a prominent frontal region and a superciliary arch (Fig. 1). The patient was using a wheelchair because of fibrous osteodysplasia in the left femur (Fig. 2). There was no pigmentation in the skin.

Laboratory findings indicated slight anemia and inflammation: white blood cell count 8100 μl^{-1}; hemoglobin 11.5 $g \cdot dl^{-1}$; hematocrit 33.6%; C-reactive protein 3.33 $mg \cdot dl^{-1}$ creative kinase 37 $U \cdot l^{-1}$. Thyroid and parathyroid functions were normal: thyroid-stimulating hormone 0.59 $\mu U \cdot ml^{-1}$; T3 3.05 $ng \cdot dl^{-1}$; T4 1.05 $ng \cdot dl^{-1}$; Ca 8.8 $mg \cdot dl^{-1}$; P 3.0 $mg \cdot dl^{-1}$; calcitonin 24 $pg \cdot ml^{-1}$. Urinalysis and chest X-ray examination detected no abnormalities. She had a history of asthma since childhood, and at the time of admission she was receiving aminophylline (4 $mg \cdot kg^{-1}$, i.v.).

The patient was able to open her mouth up to her own 1.5 finger breadth. Her parents were Jehovah's Witnesses and refused a blood transfusion for her.

Anesthetic process

As premedication, the patient received 0.5 mg atropine sulfate i.m. 30 min before entering the operation room. Anesthesia was induced with 100 mg propofol and 50 mg suxamethonium. After sufficient muscular relaxation had been attained, tracheal intubation was attempted first with a Macintosh laryngoscope. Because the device failed to show the glottis clearly, we replaced it with a McCoy laryngoscope. A 7.0-mm inside diameter endotracheal tube was successfully introduced. Anesthesia was maintained with sevoflurane, nitrous oxide, oxygen, and vecuronium until the end of the operation. The patient's heart rate, blood pressure, Sp_{O2}, $P_{ET_{CO2}}$, and rectal temperature remained stable, with no sign of arrhythmia.

Immediately after induction of anesthesia, 500 ml of a hydroxyethyl starch (HES) was intravenously infused at a relatively high rate, followed by lactated Ringer's solution (10 $ml \cdot kg^{-1} \cdot h^{-1}$). The right ovary was successfully repaired without blood transfusion.

Discussion

Since the reports by McCune et al. [2] and Albright et al. [3] in 1937, McCune-Albright syndrome (MAS) has been a rare disease in Japan. As mentioned above

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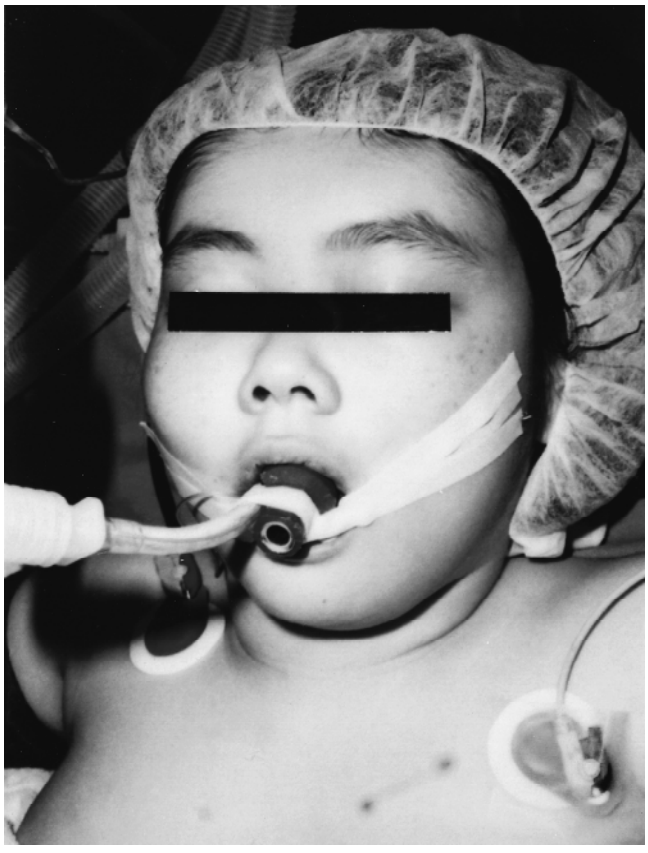

Fig. 1. Photograph of the patient demonstrating the acromegaly

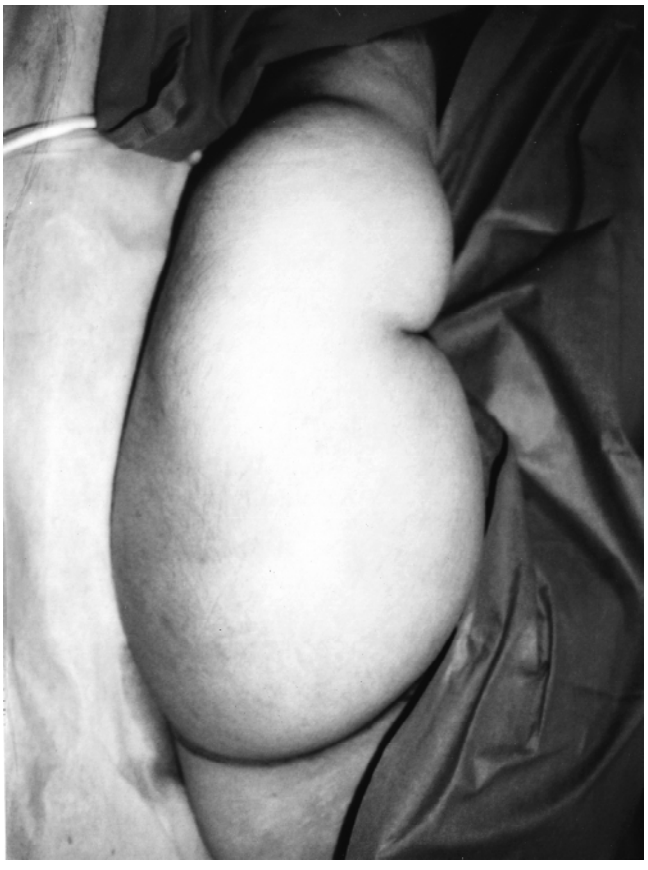

Fig. 2. Photograph demonstrating the fibrous osteodysplasia in the left femur

[1], it is accompanied by complex endocrine abnormalities. When an operation is intended for a target endocrine organ, it is necessary to employ appropriate symptomatic therapy and hormone supplementation before and after the operation.

The primary anesthetic problem in dealing with MAS is difficulty in tracheal intubation. In this disease, osteodysplasia develops in the proximal femur, pelvis, and cranium/face [4]. Among the concomitant endocrine abnormalities, acromegaly, as a result of premature or excessive secretion of somatotropin, produces a relatively small lower jaw and a large tongue, which impede visualization of the larynx with a laryngoscope. Trismus is frequently caused by the restricted mobility of the fibrotic mandibular joint. At our preoperative visit, the degree of the patient's mouth opening and tongue protrusion was rated as Class III according to Mallampati's classification [5]. In addition to this classification, we use as indices for difficulty of intubation the protrusion or lack of the upper incisors, the use of dentures, mobility of the jaw and cervical vertebrae, and thickness of the neck (buffalo hump). EL-Ganzouri et al. [6] stated that any one of these factors can result in a risk during intubation. At the time of endotracheal intubation, the condition of the patient's pharynx was rated as Grade III according to Cormack's classification [7]. By switching the laryngoscope from the Macintosh to the McCoy type, we succeeded in intubation, since the latter was reported [8] to ensure easier dilatation of the pharynx and a better visual field. Different techniques have been devised to cope with intubation difficulties, including the use of a bronchoscope, a laryngoscope equipped with an endoscope, a lamp-guided tube, and a tube guided by a laryngeal mask airway [9]. Langer et al. [10] reported the use of a thicker tube in patients with tracheas relatively thick for their age.

In cases of anticipated difficulty in intubation, care should also be taken in anesthesia induction. Nagura [11] has reported his method, characterized by a gradual increase in anesthetic depth; he stated that after ventilation is fully established, a muscular relaxant should be administered. The choice of muscular relaxants is also crucial. We used suxamethonium in the present patient because of its rapid onset and recovery. Rapacuronium [12], a recently developed nondepolarizing muscular relaxant, is as rapidly effective and recoverable as

suxamethonium. This drug is particularly useful when depolarizing muscular relaxants are to be avoided, so that the pyrexia from hyperthyroidism may not be misdiagnosed as malignant hyperthermia.

Another problem for us was the refusal of blood transfusion by the patient's family for religious reasons. During preoperative discussion, they refused any form of transfusion. We proposed the possibility of autologous blood transfusion by retrieving the patient's blood and reintroducing it to her during and after the operation, but they also rejected this proposal. In the present case, we exchanged written agreements prepared by the patient's family and resolved the problem in accordance with the hospital's ethical standards. Takaoka et al. [13] have suggested hypervolemic hemodilution as a means to facilitate the acquisition of the patient's agreement. To avoid blood transfusion, we started, simultaneously with the initiation of anesthesia, the rapid infusion of hydroxyethyl starch (HES), while closely observing the patient's circulatory system. In our opinion, this was quite effective in increasing extracellular fluid, diluting the blood, and thus reducing the loss of erythrocytes. To insure the patient's safety during the operation, the goal was to minimize blood loss and maintain the hemoglobin level above $8.5\,g \cdot dl^{-1}$, given that blood transfusion was not an option. With the use of an infusion of HES (500 ml) during the $1\frac{1}{2}$-h operation, the actual blood loss was 250 ml, and the hemoglobin level was maintained at $10.2\,g \cdot dl^{-1}$.

Throughout the emergency anesthesia for an MAS patient, we experienced many anesthetic problems, since the disease was associated with intubation difficulties and various endocrine abnormalities. To resolve these problems, a fully skilled anesthetist should select the most suitable method of anesthesia induction and prepare several endoscopical devices to cope with possible intubation difficulty, even when the operation does not involve endocrine organs.

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