Anesthetic Management of a Patient with Hallermann-Streiff Syndrome

Masafumi TASHIRO, Kazufumi OKAMOTO and Tohru MORIOKA

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Hallermann-Streiff syndrome is an uncommon congenital anomaly featuring oculo-mandibulo-cranial malformation with hypotrichosis. Originally described by Hallermann[1] in 1948 and Streiff[2] in 1950, the term "bird face" was applied because of the peculiar facial appearance. Since this anomaly is accompanied by microstomia, mandibular hypoplasia, high arched palate, and a large tongue in relation to the oral cavity, the difficulties in maintaining a patent airway become a problem for anesthetic management. We describe the anesthetic management of a patient with Hallermann-Streiff syndrome during emergency cornea transplantation.

Case Report

A 48-year-old woman, 44 kg in weight and 147 cm in height, with a diagnosis of Hallermann-Streiff syndrome, was admitted in anticipation of transplantation of the right cornea. The patient had been suffering from corneal cataract and glaucoma of the right eye for about 18 years. Although she had had a trabeculotomy for glaucoma of the right eye 15 years ago, degeneration of the cornea had gradually progressed to loss of sight.

Characteristic physical features present in this patient included hypotrichosis, mandibular hypoplasia, microstomia, dental anomalies, "Parrot-beak" like nose[3], and small eyes, as shown in figures 1 and 2. She had no motor or intellectual dificits. Laboratory studies were unremarkable except for monofocal premature ventricular contractions on ECG. An emergency transplantation of the cornea was scheduled and general anesthesia was requested by the opthalmologist when a donated cornea was obtained.

In view of the particular difficulties in maintaining an open airway with this syndrome, a fiberopticscope-guided awake tracheal intubation seemed to be the best choice. Although the patient had the typical mandibular deformity, her tongue size in relation to the oral cavity was not as large as we had expected. After discussion among the concerned anesthesiologists, we decided to attempt a conventional orotracheal intubation after a rapid induction of anesthesia with thiamylal and succinylcholine chloride. From our experience, we judged that we would be able to maintain an open airway with a mask during anesthetic induction and be able to insert the endotracheal tube. A fiberoptic bronchoscope was prepared if needed.

Atropine sulfate, 0.5 mg i.m., and hydroxydine chloride, 40 mg i.m., were administered 30 min before induction of anesthesia. In the operating room, the patient was monitored with an automated blood pressure device, ECG, and pulse oximeter. In anticipation of difficulties in keeping the airway open, we prepared a transparent mask (#5240, Vitalsigns Inc.) that fitted well with her face, a Macintosh laryngoscope with a No. 2 blade

Department of Anesthesiology, Kumamoto University Medical School, Kumamoto Japan

Address reprint requests to Dr. Tashiro: Department of Anesthesiology, Kumamoto University Medical School, 1-1-1 Honjo, Kumamoto City, 860 Japan

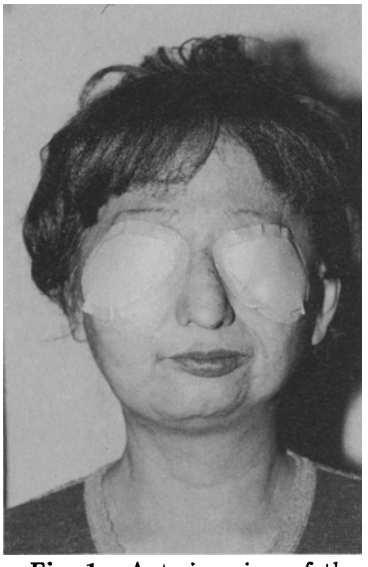

Fig. 1. Anterior view of the patient with Hallermann-Streiff syndrome.

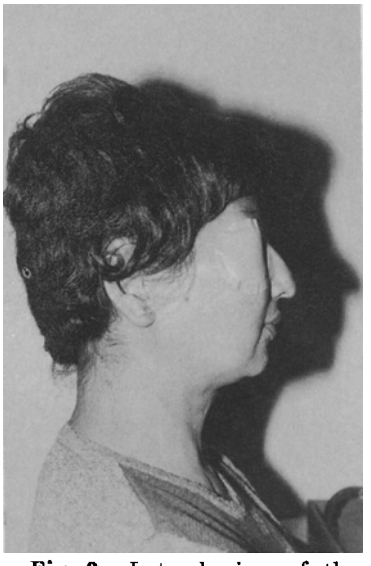

Fig. 2. Lateral view of the patient with Hallermann-Streiff syndrome.

(Penlon Co.), and a styletted endotracheal tube with an anterior curve at the tip. After about 5 min of preoxygenation, thiamylal, 50 mg i.v., was administered as a test, and then 150 mg was given for the induction of anesthesia. After confirming that mask ventilation was effective, succinylcholine chloride, 40 mg i.v., was administered for muscular relaxation. During the attempt at orotracheal intubation, although the posterior portion of the vocal cords was barely visible due to the microstomia, the curved tip of the styletted endotracheal tube was successfully advanced in front of the arytenoid into the trachea. General anesthesia was then maintained with enflurane and nitrous oxide in 40% oxygen with controlled respiration. The subsequent anesthetic course was uneventful. The endotracheal tube was removed without any difficulty when the patient was fully awake after the conclusion of the operation.

Discussion

The major clinical signs of Hallermann-Streiff syndrome are dyscephalia and bird face, bilateral congenital cataracts, and dental anomalies[4]. Other frequently associated findings include hypotrichosis, atrophy of the skin, bilateral microphthalmos, proportionate dwarfism, and mental retardation. It is suggested that this syndrome results from a developmental disorder that occurs at about the 5th or 6th week of embryonic life[5].

The most striking features, and a source of major concern for the anesthesiologist, are the upper airway deformities which make mask ventilation, laryngeal exposure and tracheal intubation difficult. Mandibular hypoplasia and a relatively large tongue compared to the oral cavity tend to produce airway obstruction during the induction of anesthesia. Abnormalities of the temporomandibular joints make opening the mouth difficult. Brittle teeth associated with dental anomalies, such as the remains of deciduous teeth, as well as arrangement anomalies present a high risk of fragmentation and aspiration of teeth during the induction of anesthesia.

In addition, nasotracheal intubation is difficult due to the high arched palate, nasal deformities called "Parrot beak", deviation of the nasal septum, and narrowing of the posterior nostril[6]. If a patient consents and

cooperates, an awake fiberopticscope-guided orotracheal intubation may be the procedure of choice since the operator can directly visualize the larynx during spontaneous respiration. However, more than 10% of the patients with this syndrome have been reported to have mental retardation[7]. The fiberopticscope-guided orotracheal intubation may not be useful for awake patients with mental retardation because of a lack of cooperation. In such cases, a fiberopticscope-guided intubation during controlled ventilation under general anesthesia is an alternative technique[8].

Oka et al.[9] reported a case of successful tracheal intubation using a conventional laryngoscope while maintaining spontaneous respiration under inhalation anesthesia. In the present case, we safely managed the airway by paying attention to the following points: (1) preparation of suitable instruments, such as a well-fitting face mask, a suitable laryngoscope, and a styletted endotracheal tube with a strong anterior curve at the tip; (2) sufficient preoxygenation to confirm maintaining an open airway for efficient ventilation during the careful administration of thiamylal, and then administration of a muscle relaxant; (3) maintenance of intubation until the patient was fully awake.

In summary, the anesthetic management of a 48-year-old female with Hallermann-Streiff syndrome is reported. The major problem in terms of anesthetic management is upper airway deformities making mask ventilation and endotracheal intubation difficult under general anesthesia. Awake intubation with a laryngoscope or a fiberoptic bronchoscope before induction of general anesthesia may be most suitable if the patient is cooperative. If endotracheal intubation is performed after the induction of general anesthesia, careful attention should be paid to maintaining an open airway and ventilation. Each alternative has advantages and management must be guided by the specific clinical circumstances.

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