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Ultrasonic adenotonsillectomy for the treatment of obstructive sleep apnea in a child with hemophilia A

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Abstract

Background: Adenotonsillectomy is now approved to be a primary treatment for obstructive sleep apnea syndrome (OSAS) in children. On the other hand, this surgery has historically been contraindicated in hemophilia. We show a case with OSAS and severe hemophilia A, in which adenotonsillectomy was successfully performed using an ultrasonic instrument. *Case history*: A 7-year-old boy with hemophilia A was diagnosed with OSAS by polysomnography (PSG). Since his adenoids and faucial tonsils remarkably hypertrophied, adenotonsillectomy was indicated. The preoperative screening tests exhibited that activated partial thromboplastin time was prolonged to 89.8 s and factor VIII was 1.1%. Fifty units per kilogram of the recombinant antihemophilic factor (Recombinate™, Baxter, USA) were administered preoperatively to obtain more than 100%, and 25 units/kg were given every 6 h for 12 days after surgery to maintain 50%. The ultrasonic scalpel (Harmonic Scalpel™, Ethicon Endosurgery, USA) was introduced into the operation for a complete hemostasis. As a result, intraoperative bleeding was successfully controlled to 8.5 g. He has been free from the disease after surgery. *Conclusion*: Adenotonsillectomy in a hemophilic child can be challenged using an ultrasonic instrument if the close collaboration of otolaryngologists and pediatricians is established.

Keywords: Ultrasonically activated scalpel; Adenoidectomy; Tonsillectomy; Recombinant antihemophilic factor; Bleeding disorder; Postoperative hemorrhage

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1. Introduction

Adenotonsillectomy is now approved to be a primary treatment to overcome obstructive sleep apnea syndrome (OSAS) in children [1,2]. On the other hand, this surgical procedure has historically been absolutely or relatively contraindicated in patients with an inherited bleeding disorder such as hemophilia. Therefore, a strategy for the treatment of OSAS in hemophiliacs has not yet been established, although a few attempts were reported [3–5].

We present here a case with a coexistence of OSAS and severe hemophilia A. In this case, adenotonsillectomy was successfully performed using an ultrasonic instrument. The sufficient perioperative replacement of factor VIII restrained troublesome postoperative hemorrhage.

2. Case history

A 7-year-old boy was found to have problems with snoring, restlessness during sleep and occasional cyanosis, and has been suffering from daytime somnolence since 6 months. He was referred to Kochi Medical School Hospital and overnight polysomnography (PSG), including oral and nasal airflow, thoracoabdominal respiratory effort and O_2 saturation, were performed in the 9-h period. PSG revealed mild to moderate OSAS and the apnea–hypopnea index (AHI) was 17.0. O_2 desaturation was obviously synchronizing with hypopneic events. Since the palatine tonsils looked remarkably hypertrophied and nasopharyngoscopic examination found adenoid tissue moderate-sized, adenotonsillectomy was indicated to solve OSAS. However, he had been diagnosed with severe hemophilia A, and the preoperative screening tests exhibited that activated partial thromboplastin time was prolonged to 89.8 s and factor VIII activity was 1.1%. To accomplish this surgical treatment of a hemophiliac safely, two aspects of strategy were considered. One was how the factor VIII could be maintained to achieve complete hemostasis. The other was how intraoperative bleeding could be locally controlled.

2.1. Surgical procedure

The ultrasonically activated scalpel (Harmonic Scalpel™, Ethicon Endosurgery, Cincinnati, OH, USA) was introduced into this operation to obtain a complete hemostasis (Fig. 1). After a Davis mouth gag was inserted, small rubber catheters were passed transnasally to gently retract the soft palate. A 4-mm 70° rigid endoscope was used transorally to visualize the nasopharynx for more adequate removal of adenoid tissue. A Harmonic Scalpel™ was operated transorally. This device has several kinds of blades; we used a Katana blade (HC 145) and a ball coagulator (HBC 05) for the adenoidectomy and a curved shears (CS 14C) for the tonsillectomy.

Adenoid tissue was initially shaved in the direction from the oropharynx to the nasopharynx, mostly using a Katana blade at a power level of 3 (Fig. 2A). Thereafter, residual tissue, especially choanal adenoid and peritubal tonsil, was cleared off using a ball coagulator at a power level of 2. After the adenoidectomy, the blade was changed to a

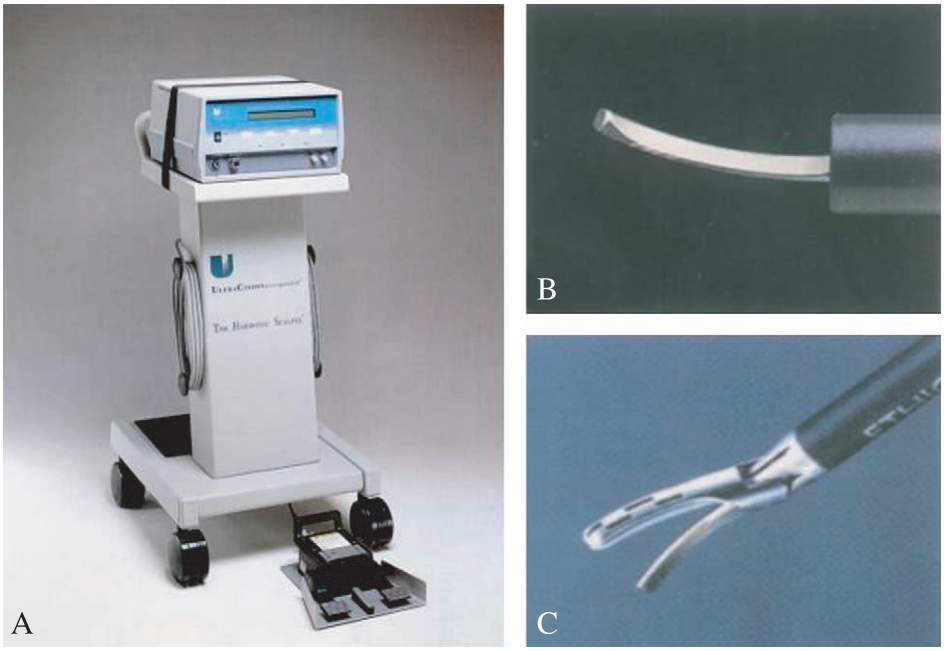

Fig. 1. Harmonic Scalpel™. (A) Generator. (B) Katana. (C) Curved shears.

curved shears. Following a usual soft palate incision and an exposure of tonsillar capsule, the palatine tonsil was dissected extracapsularily using this blade at a power level of 3 (Fig. 2B). No further hemostatic treatment was required. As a result, the amount of intraoperative bleeding was successfully controlled to 8.5 g, and the duration of the operative procedure was 20 min.

2.2. Use of recombinant antihemophilic factor

The absence of inhibitors and the satisfactory response of factor VIII rise were confirmed preoperatively. Fifty units per kilogram of the recombinant antihemophilic factor (Recombinate™, Baxter, Chicago, IL, USA) were administered 30 min prior to surgery to obtain more than 100% activity of factor VIII. Postoperatively, 25 units/kg of Recombinate™ were given every 6 h over 1 day to maintain 100%. From the second postoperative day, the same dose of Recombinate™ was infused every half a day to obtain more than 50% activity on the basis of its half-life of 14 h. However, small bleeding from the left tonsillar fossa was observed tentatively on the fourth day. At that time, factor VIII level was registering less than 35%, and this might mean unexpected consumption. Thereafter, 25 units/kg of Recombinate™ were given every 6 h and no episodes of hemorrhage were encountered till the sixth day. On the seventh day, infusion changed to every 8 h according to local healing conditions but small bleeding recurred in the same location, thereby signaling earlier infusion was needed than at the scheduled time. Although factor VIII level revealed 49.9% at that time, we decided to administer

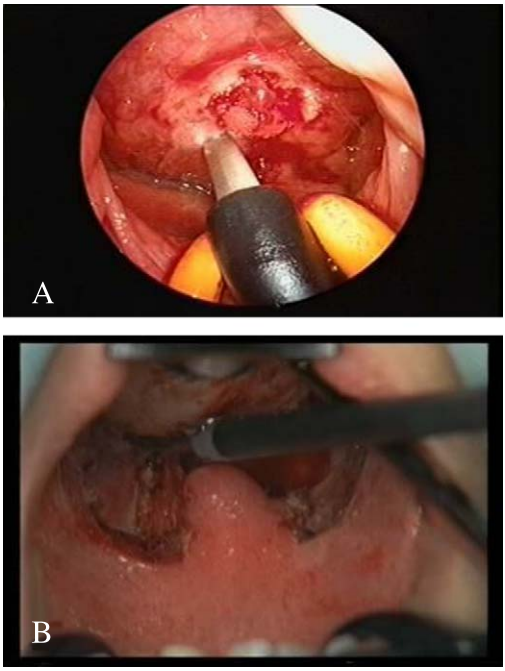

Fig. 2. Intraoperative findings. (A) Adenoid tissue was shaved mostly using a Katana blade. (B) Tonsillectomy was performed using curved shears.

Recombinate™ every 6 h again and maintained this interval until the 12th day. We diminished frequency to three times on the 13th and 14th day, and twice on the 15th day consecutively. On the 16th day, no more Recombinate™ was given, and the patient was discharged on the 18th day.

3. Discussion

In the past few reports [4,5], tonsillectomy was satisfactorily performed in two children with severe hemophilia A under the protocol of infusion of factor VIII concentrate. In them, however, the detail of administration was not discussed and adenoidectomy was avoided because of its difficulty in controlling bleeding [5]. In addition, the risk of viral infection in concentrate factor was closed up as a medicosocial problem. Recently, the recombinant antihemophilic factor was produced for viral safety and has become widely used for treatment of hemophilia [6–8]. Here we report a first case of a hemophiliac undergoing an adenotonsillectomy with a replacement of recombinant factor.

Since adenotonsillectomy was considered major, life-threatening surgery due to the risk of easily bleeding, we infused 25 units per kilogram of Recombinate™ twice a day postoperatively to maintain >50% activity on the basis of a half-life of 14 h [7,9]. Pathological hemorrhage, however, was encountered on the fourth day and factor VIII

Table 1
Proposal guideline for replacement of antihemophilic factor

Type of hemorrhage	Factor VIII level	Dose (units/kg)	Frequency
Major surgery, life-threatening hemorrhage (e.g., CNS, GI, airway)	>100% for 24 h	50	2
	>50% for 5–7 days	25	2
	>30% for additional 5–7 days	15	1–2
Adenoidectomy and/or tonsillectomy	>100% for 24 h	50	4
	>50% for 10 days	25	4
	>30% for additional 5 days	25	2–3

level revealed less than 35%. Furthermore, similar bleeding recurred on the seventh day, although three times a day of infusion resulted in the bottom level of 49.9% then. These facts may show much more was consumed than predicted, and therefore administration every 6 h was considered necessary, at least for 10 days (see Table 1).

In considering adenoidectomy, local control of primary bleeding is very important. We applied the Harmonic Scalpel™ to this operation for this purpose. Since 55,000 Hz ultrasound vibration and low-heated operation cause tissue degeneration, this instrument facilitates sealing of a severed vessel (smaller than 3 mm in diameter), and thereby has the potential to reduce bleeding [10]. As a result, intraoperative bleeding was successfully controlled to 8.5 g, including during the tonsillectomy. In addition, this surgical instrument might contribute to shorter duration of antihemophilic factor administration because more rapid healing can be expected according to animal experiments [11].

4. Conclusion

We believe ultrasonic adenotonsillectomy with sufficient replacement of recombinant antihemophilic factor, in other words, technical modification by otolaryngologists and close relation to pediatricians (or hematologists), will throw light on the treatment of hemophiliacs.

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