Anesthesia for a Patient with Recessive Dystrophic Epidermolysis Bullosa

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Two different anesthetic methods were employed for a patient with recessive dystrophic epidermolysis bullosa (R-DEB).

One was plexus brachial block in combination with ketamine infusion. The other was general anesthesia with N_2O-O_2-halothane via a face mask. In the former, no particular problem developed. In the later, however, some blisters were newly formed on the region where the anesthesist's fingers were attached to hold a face mask.

Although mask anesthesia was considered to be not always suitable for patients with DEB, we chose it because tracheal intubation may cause more serious damage to the upper airway leading to airway obstruction. (Key words: recessive dystrophic epidermolysis bullosa, monitoring. anesthesia)

(Tsukamoto N, kobayashi E, Kasuda H et al.: Anesthesia in a patient with recessive dystrophic epidermolysis bullosa. J Anesth 3: 223–226, 1989)

Dystrophic epidermolysis bullosa (DEB) is a form of diseases characterized by blisters formation and subsequent scarring of the skin and the mucous membranes following minimal trauma. This autosomal recessive disorder described first by Fox in 1879 is a rare disease (1:300000 births). However, DEB presents considerable problems to the anesthesist: tracheal intubation causes airway obstruction by blisters formation on the upper airway, and the attachment of the monitoring apparatus may make blisters. The successful anesthetic management implies minimal contact with the skin and the mucous membranes. This presents a challenge to the anesthesiologist to maintain patency of the airway during application of a face mask. We describe here such a case.

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Case Report

A 21 year-old man with skin disorder was admitted to Jichi Medical School Hospital for plastic and orthopedic surgery. Erosions were present on the skin of his legs at birth. He had had blisters and erosions after minor mechanical trauma repeatedly. Subsequetly they altered to scars. His nails defected from his fingers and toes in the first one year of life. Flexion contracture of the knees and the elbows, and deformity of fingers and toes occurred at 1.5 years of age and advanced gradually. Pseudosyndactyly of the fingers with club shaped hand developed at 15 and 19 years of age (fig. 1). There was no history of skin disease in the family. Skin biopsy made by a dermatologist revealed subepidermal blisters and cleavage beneath the basement membrane (fig.2). Electronmicroscopy demonstrated blisters between the lamina densa and the dermis. These findings

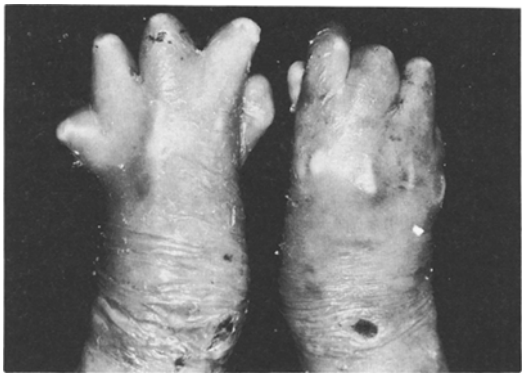

Fig. 1. Pseudosyndactyly

indicated recessive dystrophic epidermolysis bullosa (R-DEB). The patient underwent two operations for pseudosyndactyly. His body weight was 38 kg and his body height was 170 cm. There were many blisters and erosions on the whole body. Erosions were seen on buccal mucosa, soft palate and pharynx, too. There was blister containing blood on the mucous membrane of the upper esophagus. It was difficult to open his mouth sufficiently due to scar formation. Thirteen teeth had already been extracted. All his rest teeth were carious. Hemoglobin was 8.3 g/dl, indicating iron deficiency anemia on admission. He was given iron therapy, and hemoglobin rose to 12.2 g/dl on the day of the first operation. Other laboratory data were within normal limitis.

The first operation was performed to put a skin flap on his left wrist from his right leg for the purpose of release of flexion contracture, resulting in widening range of movement of his wrist. The patient was premedicated with atropine sulfate 0.4 mg and hydroxydine 50 mg intramuscularly. Supraclavicular block of the brachial plexus was performed with a 3.2 cm, 23-gauge needle. Seventeen ml of 2% mepivacaine was injected through the needle. The nerve block was perfectly successful. Skin harvesting from his leg was performed under ketamine anesthesia, at a rate of 1 mg/min contineously following a bolus of 50 mg. Droperidol was also intermittently injected in a bolus of 5 mg for sedation (total dose; 10 mg). Operation time was two hours and a half. Monitors were restricted to blood pressure measurement and ECG. The method of palpation of the radial artery was used for assessing blood pressure without a precubital stethoscope. A blood pressure cuff was well padded with soft cotton.A tourniquet was used to reduce bleeding. A tourniquet was padded with soft band and soft cotton, too. Electrodes of ECG were lubricated by lidocaine jelly. A rectal temporature probe, esophageal temporature probe and urine catheter were not used. Neither blister nor erosion was formed on the skin region through which the needle was inserted for nerve block, and to which the cuff was attached. Some erosions were formed on the regions to which electrodes of ECG were attached despite lidocaine jelly for the purpose of minimizing frictions.

The second operation was performed four

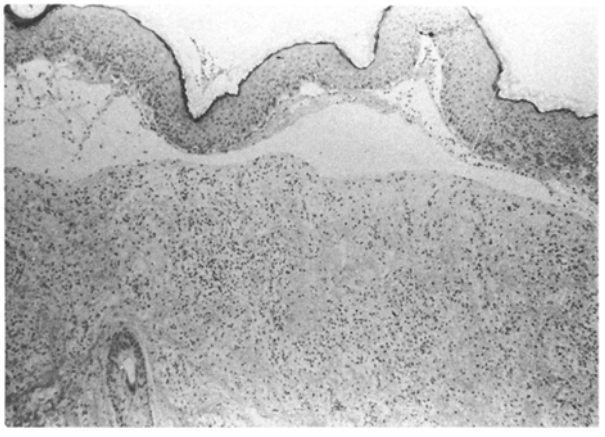

Fig. 2. Skin histology shows subepidermal bullae and cleavage beneath the basement membrane; magnification 25X

months later to release flexion contracture of his left fingers. His body weight was unchanged. Hemoglobin was 14.4 g/dl. Other laboratory data were almost within normal limits. His left fingers were covered with a pedicle flap. The flap of skin was raised from the abdomen, and under this flap his left hand was placed. The patient was not premedicated. Anesthesia was induced with 200 mg of thiopental and maintained with N_2O 4 L/min, O_2 2 L/min, and 1-1.5% halothane via a face mask connecting with a circle system. The skin with which the mask came into contact was protected with lidocaine jelly and soft cotton wadding. Muscle relaxant was not used. Spontaneous ventilation was maintained and not assisted manually. Twenty-two gauge catheter needle was inserted into the left dorsalis pedis artery and fixed to the skin with a silk stitch for contineous blood pressure monitoring. This catheter was also available for arterial blood gas analysis. Much more lidocaine jelly than in the first operation was put beneath electrodes of ECG. Blood pressure cuff was not used. Operation time was two hours and forty five minutes. It was very difficult to fit a mask to the patient's face and keep the airway patent by head tilt-jaw thrust maneuver, without giving friction to his skin. Immediately after anesthesia, some erosions were only on the region to which the anesthesist's fingers were attached.

Discussion

Epidermolysis bullosa (EB) is a form of diseases that comprises mucocutaneus blistering disorders after minimal mechanical trauma. The disease has been classified on clinical, pathological, genetic and biochemical criteria into four distinct categories: EB simplex, junctional EB, dominant dystrophic EB, and recessive dystrophic EB[2]. EB simplex and junctional EB are non scarring type. Dominant dystrophic EB and recessive dystrophic EB are scarring type. In non scarring types, examination by electronmicroscopy reveals changes above the basal lamina in the lower layers of the epidermis. In scarring types, the level of damage in the skin is in the upper dermis and elecronmicroscope reveals anchoring fibrils between the basal lamina and the dermis to be missing or broken[3,4]. Of these types, recessive dystrophic EB (R-DEB) is of special interest to anesthesiologists.

The case presented here was R-DEB that involved the mucous membrane. Therefore, R-DEB tends to affect not only the extremities resulting in fusion of digits but also the mouth, tongue, and esophagus. Microstomia, ankyloglossia, and esophageal strictures may be present. Severely dystrophic teeth are common. Anesthesia is required in patients with DEB for the purpose of correction of pseudosyndactyly, release of esophageal strictures, removal of scar tissue, or dental treatment. The risk of blisters formation must be reduced by appropriate anesthetic techniques.

Anemia, hypoproteinemia, electrolyte abnormalities, and dehydration are commom. Those abnormalities should be corrected before anesthesia.

In the anesthetic management we must protect a patient with DEB against minimal trauma. Shearing, friction and pressure to the skin and the mucous membrane result in blisters formation. Patients should be allowed to move themselves on to the operating table to minimizing trauma. The surface of the table should be smooth and be padded with wadding. All kinds of adhesives should not be used because they may cause blisters and erosions. Adhesive paper electrodes for ECG must be omitted. Much jelly should be put between the patient's skin and them. For blood pressure measurement, the technique of the direct intraarterial cannula sutured to the skin is recommended. A tourniquet over soft cotton padding should be used for the skin protection.

The most controversial problem for anesthesists is that tracheal intubation may cause blisters formation on the mucous membrane of the trachea, the larynx, the epiglottis and the pharynx, subsequently resulting in severe airway obstruction. There have been no previous reports of laryngeal or tracheal complications following tracheal intubation.

Although there is a report that intraoral surgey was done successfully by endotracheal technique, tracheal intubation should be avoided whenever possible, because airway obstruction due to blisters formation may occur after extubation[5]. It is very difficult to keep head tilt-jaw thrust maneuver without damaging the skin over the mandibula and the face. Therefore, mask anesthesia may not be recommended. If a mask is to be used, soft cotton wadding lubricated with jelly and steroid ointment should be applied to the skin. Regional conductive anesthesia is a good alternative, provided that the site of injection is free from skin lesions and infection[6].

Ketamine anesthesia has been used for patients with DEB[7]. The contineous infusion technique may be preferred since anesthetic condition is stable. However, ketamine may increase salivation and secretions from the respiratory tract, subsequently causing airway obstruction. Therefore, we must observe the patient's respiration carefully during ketamine anesthesia.

It has been suggested that there is a close association between porphyria cutanea tarda and epidermolysis bullosa[8]. Barbiturates are contraindicated in porphyria cutanea tarda (PCT). We should rule out PCT, whenever barbiturates are chosen as an anesthetic agent[9].

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