

Difficult airway management of a child with Blepharophimosis Syndrome: Case report

Ahmet Selim Ozkan¹, Nihat Polat², Sedat Akbas¹, Zeynep Koylu¹, Mahmut Durmus¹

¹Inonu University, Faculty of Medicine, Department of Anesthesiology and Reanimation, Malatya, Turkey

²Inonu University, Faculty of Medicine, Department of Ophthaomology, Malatya, Turkey

Abstract

Blepharophimosis syndrome (BS) is a rare autosomal dominant syndrome characterized by eyelid malformations, and abnormal facial morphology. Flat and wide nasal roots, short neck, restricted mouth opening and neck extension, characteristic facial appearance, microcephaly and micrognathia make airway management of patients with BS important for anesthesiologists. Due to systemic pathologies and craniofacial anomalies, difficult airway may be seen in these patients and anesthesia management is important in patients with BS. Because of the difficult airway risk, it is important to determine the anesthetic method and make the necessary preparations. In this case report, we aimed to present difficult airway management of a patient with BS who underwent surgery for ocular disorders. This case report is the second case presented by successful airway management in general anesthesia practice.

Keywords: Difficult Airway; Blepharophimosis Syndrome; Child.

INTRODUCTION

Blepharofimozis syndrome (BS) is a rare autosomal dominant disorder affected FOXL2 gene in the 3q chromosome. BS is a complex eye lobe disorder characterized by ptosis, epicanthus inversus, and blepharophimosis. The narrowed horizontal valve space is the most obvious finding (1). In addition, broad and flat nasal roots, palate and ear anomalies, sporadic mental retardation, gonadal anomalies are common findings. In these patients, difficult airways may be seen due to systemic pathologies and craniofacial anomalies and therefore anesthesia management is important. Because of the difficult airway risk, it is important to determine the anesthetic method and make the necessary preparations. This case report is the second case presented by successful airway management in general anesthesia practice (2). We reported successful airway management during general management of children with BS planned for surgical repair of telecantus.

CASE REPORT

A 2-year-old, 15 kg, male patient admitted to our hospital due to an eyelid anomaly. It was learned that he was

diagnosed with BS 6 months ago from patient's parents and had difficulty breathing in sleep. Repair of right/left epicanthus with nasal wiring and bilateral Z plasty technique were planned and mental retardation, flat and wide nasal root, high arched plate, short neck and limited neck extension were detected in the preoperative physical examination of the patient (Figure 1).

In addition, Mallampati score was 3. Routine laboratory values and electrocardiogram (ECG) were normal. Informed consent was taken from the patient's family. The patient was taken to the operating room. Standard monitoring was performed and heart rate was 110 / min, blood pressure was 90/50 mmHg.

Premedication was applied with 0.5 mg / kg oral midazolam. Thirty minutes prior to surgery, Necessary equipment (supraglottic airway vehicles (i-gel, laryngeal mask airway), intubation tubes and laryngoscope blades at different sizes, videolaryngoscope and fiberoptic bronchoscope) were made available due to the expected difficult airway. Preoxygenation was performed with 100% O₂ for 5 min before induction of anesthesia. Anesthesia induction was achieved with an 8% sevoflurane and O₂ mixture, and vascular access was provided. Propofol 2.5 mg / kg IV and fentanyl 1 mcg / kg IV were administered

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Corresponding Author: Ahmet Selim Ozkan, Inonu University, Faculty of Medicine, Department of Anesthesiology and Reanimation, Malatya, Turkey, E-mail: asozkan61@yahoo.com

when manual mask ventilation was evident. Muscle relaxant was not applied to the patient. For airway control, laryngeal mask airway (LMA) no: 2,5 was applied but failed. After the failure of the second trial, it was decided to implement the i-gel supraglottic airway device (no: 2.5) and i-gel was successfully placed successfully at first attempt. Anesthesia was carried out using 2% sevoflurane in the 50% oxygen/air mixture. Any perioperative problem was encountered in the operation which lasted for seventy-five minutes.

Paracetamol 15 mg/kg IV was administered for postoperative analgesia. It was observed that the face was edematous because of surgery. At the end of the operation, the patient was extubated smoothly. The patient who was hemodynamic and respiratory stable was transferred to the service after 60 minutes in the postoperative care unit.



Figure 1. Appearance of patient with BS.

DISCUSSION

Blepharophimosis syndrome (BS) was first categorized into two types in 1921 (3). Type 1 is accompanied by premature overgrowth and infertility in women, while Type 2 has only facial and ocular findings (4). Other systemic involvements are rare in BS. Similar to BS, eye and airway problems are also present in Freeman-Sheldon syndrome, Dubowitz syndrome and Noonan syndrome. These syndromes may cause blepharophimosis and airway

anomalies as well as other systemic involvement (5-9). Patients with BS refer to the hospital for ophthalmic surgery in childhood and adolescence. In particular, flat and wide nasal roots, short neck, restricted mouth opening and neck extension, characteristic facial appearance, microcephaly and micrognathia make airway management of patients with BS important for anesthesiologists. Similarly, an anesthetic management of a patient with Dubowitz syndrome, which is difficult due to facial anomalies, has been reported, and successful endotracheal intubation has been reported using direct laryngoscopy (5). In airway management, the difficult airway is seen in 1.5-20% of the cases, with 2-3% serious difficulties encountered (10). Supraglottic airway devices are frequently used as an alternative to endotracheal intubation with direct laryngoscopy in difficult airway management. Especially the new generation supraglottic airway vehicles are preferred because of easy practice and high success rate of application. Young et al. (11) advised LMA because of blind and safely implementation in airway management of these patients.

Proseal LMA and Glidescope videolaryngoscope is also proposed as an alternative to fiberoptic bronchoscope (12). Baidya et al. (2) reported that cobra perilaryngeal airway (cobraPLA) was successfully applied after failed LMA application in patient with BS.

In fact, Ozkan et al. (13) presented successful difficult airway management provided by rigid endoscopes in field conditions. In our case, classic LMA was tried firstly, but i-gel was successfully applied when it could not succeed in settlement. Awake fiberoptic intubation is the first choice in patients with expected difficult airway, but because of the small age of the patient, it was not performed due to non-cooperation. Probably, LMA could not be placed completely because of the limited neck extension and high arched plate. This problem was overcome by i-gel.

CONCLUSION

Patients with BS must be evaluated preoperatively for difficult intubation due to craniofacial anomalies and preparations for management of difficult airway must be made preoperatively. We reported that airway management could be achieved successfully with i-gel in patient with BS.

Conflicts of interest: None

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