Anesthetic management in Cornelia de Lange Syndrome

Cornelia de Lange sendromunda anestezi uygulaması

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Sir,

Cornelia de Lange syndrome (CdLS), is a rare (1/10000-60000 birth) multiple congenital disorder in which half of the cases are linked to a defect of the Nipped-B-like gene on chromosome 5 (1, 2). Patients are characterized by distinctive facial appearance including eyebrow fusion, elongated philtrum, micrognathia, short neck, psychomotor delay, hirsutism and upper limb malformations and gastrointestinal (GI) malformations (3). Difficult airway and aspiration risk due to gastroesophageal reflux and poor esophageal motility are the main challenges in anesthesia management (4, 5).

We present a case of CdLS subjected to eye surgery for nasolacrimal duct obstruction under general anesthesia with laryngeal mask airway (LMA) in a 3 year-old girl patient, 69 cm tall, 6 kg weight (ASA physical status class III). Recurrent respiratory infections and frequent food regurtation were reported on her medical history. Physical examination showed a child of short stature, with a small mandible, pronounced incisors, low frontal line of hair implantation, hirsutism, micrognathia and class IV modified Mallampati score (Figure 1). Blood pressure, oxygen saturation, and ECG were monitored in the operating room. Various large blades of Miller and Macintosh type, laryngeal mask airways, tracheal tubes with stilettes, fiberoptic bronchoscope and a tracheostomy set were kept ready. Anesthesia was induced with iv. lidocaine (1mg kg^{-1}), propofol (2 mg kg^{-1}) and remifentanil (0.5 µg kg^{-1}). Muscle relaxants were not used. A nasogastric tube was inserted and confirmed that the stomach was empty. Size-2 LMA (LMA-Classic™) was replaced and correct placement confirmed by auscultation and capnography. Anesthesia management was continued with iv infusion of 100-200 µg kg^{-1} dk^{-1} propofol and 0.15 µg kg^{-1} dk^{-1} remifentanil. Hemodynamic and other vital parameters were stable during intraoperative period. The duration of the surgery was 35 minutes. No respiratory or hemodynamic problems were occurred and she was discharged at same day.

CdLS is a rare syndrome with an early mortality form serious aspiration-related problems and consequent frequent infections. The craniofacial features related with difficult airway include macroglossia, cleft lip/palate, midface hypoplasia, high arched palate, and mandibular hypoplasia. Preanesthetic careful examination for cardiorespiratory system is essential (3, 4). Postoperative complications include unstable cardiac function and marked susceptibility to

infections therefore vigilant clinical observation is necessary. In conclusion, management of children with CdLS is a challenge for anesthesiologist and LMA is a good alternative to mask ventilation or tracheal intubation with excellent results in proper operations and patients.

Written permission was sought and received from the parents of patient to report this case.

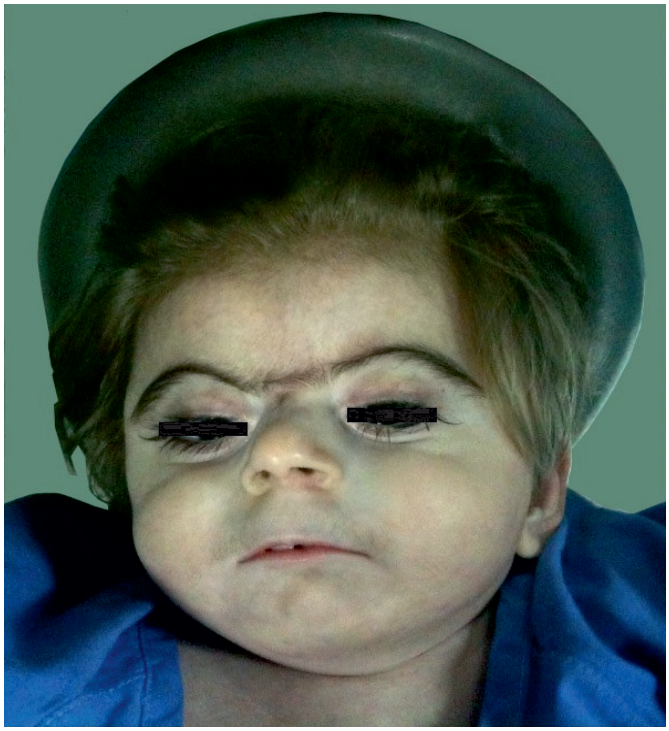

Figure 1: Distinctive facial appearance of patient with Cornelia de Lange syndrome (eyebrow fusion, elongated philtrum, micrognathia and short neck)

Yazarlarla ilgili bildirilmesi gereken konular (Conflict of interest statement) : Yok (None)

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