C- mac videolaryngoscope: an aide to new findings in Goldenhar Syndrome

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ABSTRACT

Goldenhar Syndrome (GS) is a congenital disorder with male predominance and sporadic occurrence. Various anomalies are seen in these patients involving almost all systems. The anesthesiologist usually finds difficulty in the airway of these patients as a spectrum of anomalies is seen. We report a new finding in the airway of Goldenhar syndrome with the help of a C-Mac videolaryngoscope, with the base of tongue and uvula being bifid and causing inability to intubate in an 11-year-old girl.

Keywords: *Airway, C-Mac videolaryngoscope Goldenhar syndrome.*

Goldenhar Syndrome (GS) named after the scientist of its discovery, is a congenital disorder with male predominance and prevalence from 1 in 3500 to 5600 [1]. It occurs as sporadic cases and its inheritance is still of the query [1,2]. A congenital anomaly in the formation of the first and second branchial arch is the reason for the development of this syndrome [3,4]. Various anomalies are seen in the patient including facial asymmetry and hypoplasia due to microtia, hemifacial microsomia, mandibular hypoplasia, external ear malformations, ocular anomalies with lid colobomas, microphthalmia, corneal hypoplasia, vertebral anomalies with possibilities of subluxation at the atlanto-occipital joint, cardiac malformations with Tetralogy of Fallot being the commonest followed by septal defects and situs inversus, nervous system and genitourinary anomalies [5,6].

Anesthesiologist usually find difficulty in airway of these patients as a spectrum of anomalies are seen ranging from normal airway to high arched or cleft palate to tongue abnormalities and the larynx being anterior in majority of them. [4,7,8,9]. We report a new finding in the airway of Goldenhar syndrome with the help of a C-Mac videolaryngoscope, with the base of tongue and uvula being bifid and causing the inability to intubate.

CASE REPORT

An11-year-old girl child,a diagnosed case of Goldenhar syndrome with the typical features like microcornea, uveal coloboma, corneal opacity, limited neck mobility, lower cervical spine showing segmentation on X-ray, receding mandible and bilateral anotia using hearing aide since 8years. She presented to us with a cyst in the posterior fossa and a communicating hydrocephalous for which a ventriculo-peritoneal shunt was to be done. She had a significant history of being taken up for the same surgery in a peripheral center where surgery was deferred in view of the inability to intubate after three attempts of laryngoscopy by a trained experienced anesthesiologist. On examination, the airway was an easy laryngoscopy by Wilson's scoring system.

We had prepared our operation theater with all the equipment for difficult airway along with C-Mac videolaryngoscope, flexible fiberoptic, percutaneous tracheostomy and retrograde intubation. Informed consent was taken from the parents and the patient was wheeled into the theater. Standard monitors were applied and intravenous access was obtained. Baseline vitals were as follows: heart rate 78 beats per minute, blood pressure102/68 mm Hg and pulse oximetry reading 100% on room air.

Our plan for general anesthesia was to get a view of the glottis before any muscle relaxant was given and maintaining manual in-line stabilization all throughout. The induction of anesthesia was done by 2mcg/kg fentanyl and slow titrated aliquots of propofol. Sevoflurane and oxygen were used to deepen the plane of anesthesia. Once the plane was achieved, C-blade no. 2 of the C-Mac video laryngoscope was introduced. We visualized the bifid uvula and base of the tongue, which was also bifid (Fig. 1). Epiglottis could not be seen even on external manipulation, Cormack and Lehane grade 4 was labelled.

After intermittent bag and mask ventilation and propofol boluses, the next attempt of laryngoscopy was done with D-blade no.3 of C-Mac video laryngoscope. Now the epiglottis came in view, which was liftable and with external manipulation, a tube

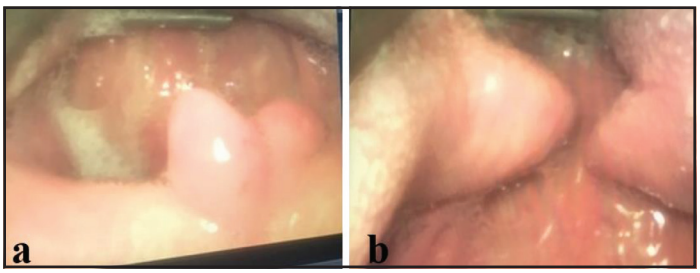

Figure 1: C-Mac view showing bifid (a) uvula and (b) base of the tongue.

of size 6.5mm was introduced into the trachea. After confirmation of the tube position, rocuronium 1mg/kg was given. After settling the vital parameters, a flexible fiberoptic bronchoscope was introduced through the tube to check for any tracheal anomaly, but nothing was found. Surgery and extubation were uneventful. The patient was observed in post-anesthesia care unit for 2 hours and then shifted to the ward. She was discharged from the hospital on 3rd postoperative day with advice for a follow-up.

DISCUSSION

In 1963, Gorlin et al. devised the term occulo-auriculo-vertebral dysplasia to distinguish the patients with microtia, macrostomia, mandibular hypoplasia, vertebral anomalies and epibulbar dermoids [5]. Feingold and Baum did a retrospective study on 17 patients and made criteria for the diagnosis of these patients which included atleast two of the following: eye abnormalities (lipomas, lipodermoids, epibulbardermoid) along with ear, mandible or vertebral anomalies [10].

Airway stands apart amongst all the anomalies of these patients as an inability to manage the airway and unavailability of alternative plans can lead to catastrophe. Assessment of a pediatric airway is very difficult to let alone it being a syndromic child. The anesthesiologist usually finds difficulty in the airway of these patients as a spectrum of anomalies are seen ranging from normal airway to high arched or cleft palate to tongue abnormalities and the larynx being anterior in the majority of them [4,8-10]. We were being kept on the edge by the fact that our patient was deferred from surgery elsewhere because of the inability to secure the airway.

C-Mac video laryngoscope is an advent in the airway management. The D blade of this laryngoscope was developed by Prof Volker Dorges from the University of Kiel. It is a hemi-moon shaped blade with a 40-degree curvature and 80-degree field of vision, making visualization of glottis easier without any undue pressure or lifting force [11]. We used this D blade to visualize and lift the epiglottis.The vision of a bifid uvula and base of the tongue was a finding which has not been reported earlier. Sun et al used McGrath videolaryngoscope to displace the tongue and enlarge the oral cavity so that nasal fiberoptic intubation could be done [12]. Others have reported fiberoptic intubation through an I-gel supraglottic airway [13]. Another report states how the tongue was held outside the oral cavity with surgical gauze in order to make room for fiberoptic in a severely receding mandible [14].

We planned on not giving any muscle relaxation before the glottis view as we did not want to lose the patient's respiratory efforts in an untoward event of not being able to intubate, whereas, on the other hand, it can also be considered that since the bag-mask ventilation was effective and easy, short-acting muscle relaxant, succinylcholine could have been used for a better glottis view.

Other associated anomalies were all to be taken care of. We maintained manual in-line stabilization all throughout our efforts of intubating and extubating the patient for fear of any impending subluxation that may occur with our neck manipulations. Our patient had scoliosis and so cotton padding was done appropriately in order to prevent any soft tissue soreness at the end of surgery. Adequate eye cover was placed in order to prevent any damage to the already affected eyes.In the postoperative period, strict vigilance has to be maintained for any unexplained periods of apnoea, which may happen due to undiagnosed vertebral anomaly.

CONCLUSION

The patient with Goldenhar syndrome requires a thorough preoperative examination but still, the anesthesiologist should be prepared for any new findings in the airway that may make it difficult to secure the airway. A backup alternative plan should be prepared and the last resort of a percutaneous tracheostomy should be readily available.

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