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Perioperative Challenges in Airway and Ventilatory Management of a Neurosurgical Patient with Klippel–Feil Syndrome

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Abstract

Klippel–Feil syndrome, a rare congenital anomaly, has a classical triad of low posterior hairline, short neck, and restriction of neck movements. Complex airway anatomy increases the risk of neurological damage while handling the airway during positioning and laryngoscopy. We address the possible anesthetic challenges that one might face in these patients, such as difficult airway, difficult weaning from the ventilator with increased chances of reintubation, prolonged intensive care and mechanical ventilation, and difficult tracheostomy. Every anesthesiologist must be aware of the available options in the management of a difficult airway in emergent circumstances. Despite the numerous encounters, we were able to effectively manage a case of Klippel–Feil syndrome perioperatively.

Keywords
- Klippel–Feil syndrome
- airway management
- tracheostomy
- intracranial hemorrhages

Introduction

Klippel–Feil syndrome (KFS) is a rare skeletal disorder characterized by abnormal fusion of two or more cervical vertebrae. Securing the airway in such patients is challenging and a thorough examination overcomes it. Although awake fiberoptic intubation is considered ideal for difficult airway (DA), the use of video laryngoscopes in patients with adequate mouth opening may offer a superior glottis exposure.[1] Long-term intensive care unit (ICU) stay, delayed weaning from the ventilator, increased incidence of reintubation, and difficult tracheostomies are the other challenges in patients with deviated anatomy. We report a case of hemorrhagic stroke in a patient with KFS scheduled for emergency decompressive craniotomy and challenges faced in the perioperative period.

Case Report

A short-statured (height 122 cm; weight 46 kg; body mass index 30.9 kg/m^2), 43-year-old female with no known medical comorbidities was presented with a history of sudden onset unresponsiveness followed by altered sensorium. She was a diagnosed case of KFS and had undergone neck contracture release at 1 year of age. Her family history was unremarkable. Her Glasgow Coma Scale (GCS) was $E_2V_3M_4$ and pupils were bilaterally equal and reactive to light. Cardiovascular and respiratory system examinations were unremarkable. Vitals were stable but there was airway obstruction with tongue fall, requiring emergency endotracheal intubation. Airway examination revealed a short-webbed neck, mouth opening of 2 finger-breadths, Mallampati grade 4, macroglossia, and complete restriction

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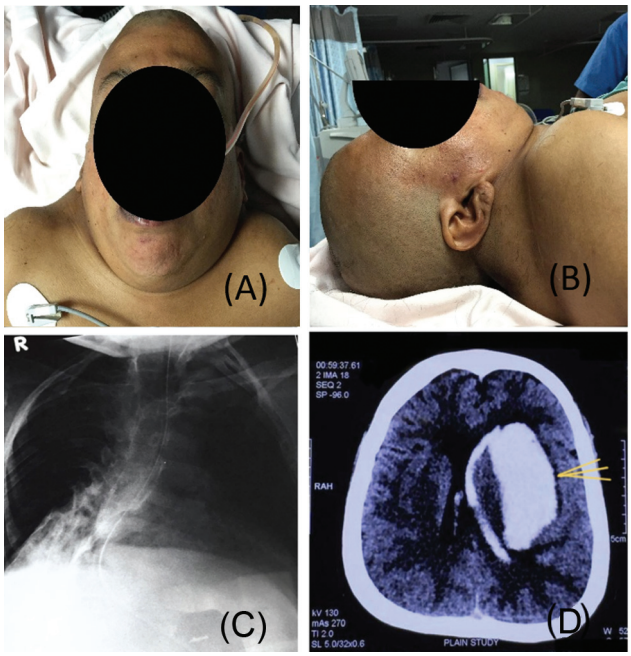

Fig. 1 (**A**, **B**) Anteroposterior (AP) and lateral profile. (**C**) Chest radiograph. (**D**) Computed tomography (CT) brain showing left external capsular bleed with mass effect.

of movement at the cervical spine. Trachea was deviated to the right and the tracheal rings were not well appreciated (►**Fig. 1A, B**).

Chest radiograph showed scoliosis of the thoracic spine and tracheal shift to the right (►**Fig. 1C**). Noncontrast computerized tomography (CT) of the brain revealed a left external capsular bleed with midline shift and mass effect (►**Fig. 1D**). The patient was planned for emergency decompressive craniotomy and hematoma evacuation. Four percent lignocaine 100 mg as nebulization was administered 30 minutes before surgery.

In the operating room, she was placed in a propped-up position, and an oropharyngeal airway of size 2 was inserted to prevent tongue fall. After administering injection fentanyl 50 µg and injection esmolol 25mg, laryngoscopy was performed with Karl Storz C-MAC video laryngoscope size 3 blade, and the trachea was intubated with styleted 7.0 mm ID flexometallic cuffed endotracheal tube and secured at 15.5 cm after confirming position. Anesthesia was induced with propofol titrated to response, vecuronium 6 mg, and fentanyl 80 µg. The intraoperative period was uneventful with the brain being lax postevacuation of hematoma. Postoperatively, as we decided to electively ventilate, the flexometallic tube was exchanged with a 7.5-mm ID Portex cuffed endotracheal tube. She was shifted to neurointensive care for postoperative ventilation and further management. Antiedema and antiepileptic measures were continued. Over 10 days, neurologically she improved with a GCS of $E_4V_tM_5$ and mild hemiparesis of the right upper and lower limbs. After spontaneous breathing trials, she was extubated but had to be reintubated within 3 days of extubation as she could not maintain oxygen saturation with noninvasive ventilation (NIV). Anticipating prolonged ventilation, the patient was planned for tracheostomy. Bedside ultrasonography (USG) of the neck uncovered a deep-seated trachea which was confirmed on CT thorax (8 cm from skin). It also revealed a fused atlantoaxial joint with a complex segmentation anomaly of the cervical spine (►**Fig. 2A**). The possibility of sternotomy during tracheostomy was suggested by the otorhinolaryngologist. Specialized extra-long tracheostomy tubes with adjustable flanges known as Extra TracheoFlex tubes by RUSCH (Georgia, United States), sizing from 7.5 to 9.5 mm ID with a length of 15.5 cm (►**Fig. 2B**) was arranged. As it was deemed high risk, the patient's relatives did not consent for tracheostomy.

We continued mechanical ventilation with graded intermittent spontaneous breathing trials for another 10 days along with adequate enteral nutrition, physiotherapy, and measures to prevent bedsore and deep vein thrombosis. With improved neurological status and pulmonary function, she was extubated onto preemptive NIV and finally to oxygen supplementation via facemask. She was discharged and advised to continue physiotherapy, antiepileptics, and neuroprotective medications.

Discussion

KFS is a congenital skeletal disorder occurring due to abnormal segmentation of the cervical somites during organogenesis. It was first described by Maurice Klippel and Andre Feil in 1912.[2] The clinical triad of short neck, decreased cervical mobility, and low posterior hairline is seen only in 40 to 50% of cases.[3] Cases are mostly sporadic, few due to autosomal dominant or autosomal recessive inheritance.[4]

Awake fiberoptic intubation is considered the gold standard for the management of the airway in patients with KFS. Shah et al reported awake fiberoptic intubation in a KFS parturient with type 1 Arnold-Chiari malformation undergoing elective caesarean delivery. Conscious sedation was achieved using dexmedetomidine infusion until successful tracheal intubation.[5] Stallmer et al described a series of successful airway management in 10 patients with KFS, four of them with laryngeal mask airway placement and six patients underwent endotracheal intubation of which five were accomplished with direct laryngoscopy while only one patient required fiberoptic intubation.[3] The dynamic nature of the airway has been described by Pavani and Krishna, where the airway was managed by videolaryngoscopy and direct laryngoscopy in the same KFS patient under different surgical contexts.[6] In our patient with a low GCS and compromised airway, sedation was avoided and awake intubation was achieved with the aid of a videolaryngoscope.

Anthropometric studies using chest radiograph, neck USG, and CT anticipates unconventional extended-length tracheostomy tubes with adjustable flanges when skin-trachea distance was > 4.4 cm.[7–9] Ahuja et al reported the use of endotracheal tube in a difficult tracheostomy scenario arising secondary to surgical emphysema.[10] It can be tried in such dire emergencies when specialized tracheostomy tubes are unavailable. But navigating and securing the tube would be a cumbersome process.

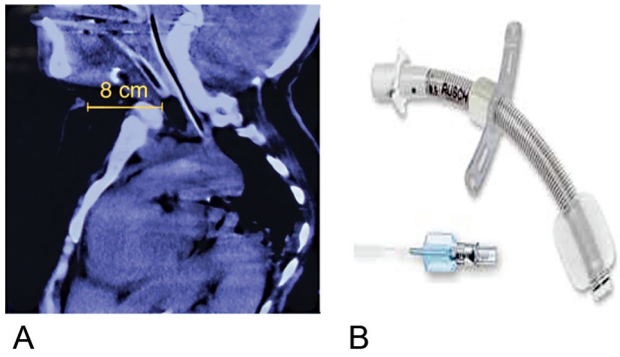

Fig. 2 (**A**) Computed tomography (CT) thorax—trachea at 8 cm skin depth. (**B**) Extra TracheoFlex tube with adjustable flange by RUSCH.

There is a lack of literature about the management of such patients postoperatively. Weaning and extubation attempts should be gradual and graded with spontaneous breathing trials and cuff leak tests onto preemptive NIV or high-flow nasal cannula. Attempting early tracheostomy and weaning off the ventilator decreases their ICU stay, prompting early mobilization and rehabilitation thereby reducing the incidence of hospital-acquired infections and other complications.

Conclusion

Even though awake fiberoptic intubation is considered the gold standard for the management of DA in cases of KFS, other options could also be explored. The use of imaging tools like plain radiographs, USG, CT, and magnetic resonance imaging of the neck and chest can help in the planning and execution of intubation. Depending on the neurological status of the patient, one can anticipate prolonged mechanical ventilation and ICU care. The use of unconventional extra-long tubes for tracheostomy can be of help in early weaning and discharge.

Conflict of Interest

None declared.

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