Anaesthetic management of a patient with Klippel-Feil syndrome

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Citation

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Abstract

Klippel feil syndrome is a rare entity. After its first description in 1912 by Maurice Klippel, a French neurologist and psychiatrist and André Feil, a French neurologist the management of this syndrome still poses a big challenge to clinicians, orthopaedic surgeons , anaesthetists and intensivists. They described patients who have a short neck, decreased range of motion in the cervical spine and a low posterior hairline. It involves congenital failure of segmentation of cervical vertebrae which results from failure of normal segmentation of cervical somites at 3-8 weeks of gestation resulting in multiple fused segments. Spectrum of deformity ranges from fusion of 2 vertebrae to involvement of entire cervical spine. Fusion of C2 & C3 is most common. Here we report the anaesthetic management of a case of Klippel-Feil syndrome who presented with fracture both bones left forearm scheduled for open reduction and internal fixation.

INTRODUCTION

Klippel Feil Syndrome was described independently in 1912[1] by Maurice Klippel and Andre Feil. It is caused by a failure in the normal segmentation of the cervical vertebrae during the early weeks of fetal development. Features include a short neck, restricted mobility of the upper spine and a low hairline. Any of the cervical vertebrae can be involved but the commonest fusion is of C2,3. Fiel classified the syndrome into 3 types[2]; Type I is massive fusion of many cervical and upper thoracic vertebrae into bony blocks. Type II is fusion at only one or two interspaces although hemivertebrae, occipito-atlantal fusion and other anomalies might also occur. Type III is both cervical fusion and lower thoracic or lumbar fusion. They pose a challenge to the anaesthesiologist with regard to the management of the difficult airway. These patients have a potentially unstable cervical spine and abnormal atlanto-occipital junction and are prone to an increased risk of neurological damage not only during laryngoscopy and intubation but thereafter.

CASE REPORT

A 53 year old male was scheduled for open reduction and internal fixation of both bones fracture of left forearm which he sustained following a fall. He was a known case of Klippel-Feil syndrome with a short webbed neck, sprengel shoulder and low posterior hair line and no other known co-morbidities. On examination the patient was well built and nourished with a pulse rate of 82 beats per minute and a blood pressure of 140/82 mm of Hg. He had a short webbed neck (Figure 1) and a low posterior hairline with very much limited flexion (Figure 2A, 2B) and extension (Figure 3A, 3B). Examination of the airway showed that there was limited flexion or extension of the upper cervical spine, he had adequate mouth opening and his Malampatti grade was 3. The systemic examination was normal. His investigations were all within normal limits, except x-ray of cervical spine which showed fusion of the upper cervical spines (Figure 4).

Figure 1

Figure 1: Short webbed neck

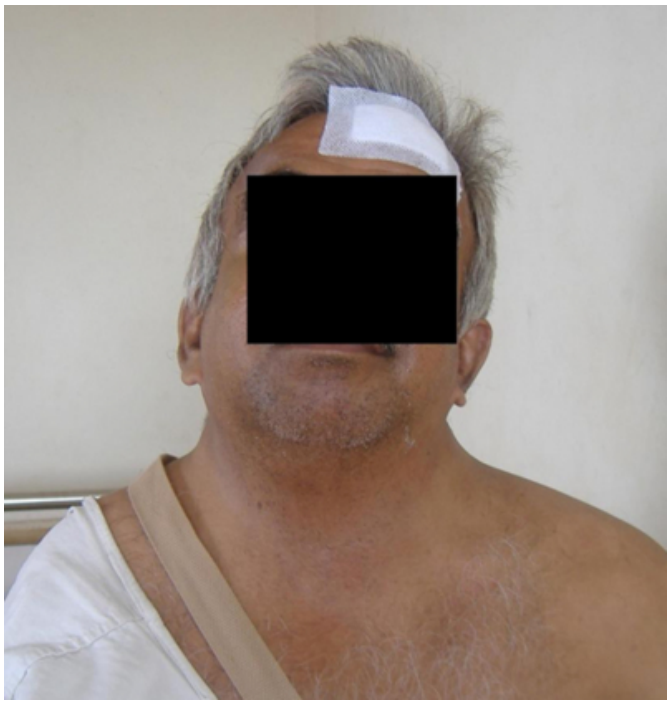


Figure 2

Figure 2A: Limited extension

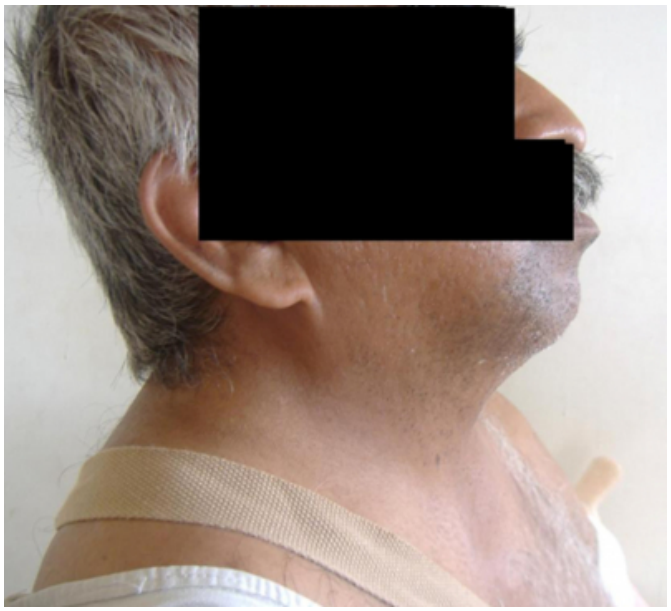


Figure 3

Figure 2B: Limited extension

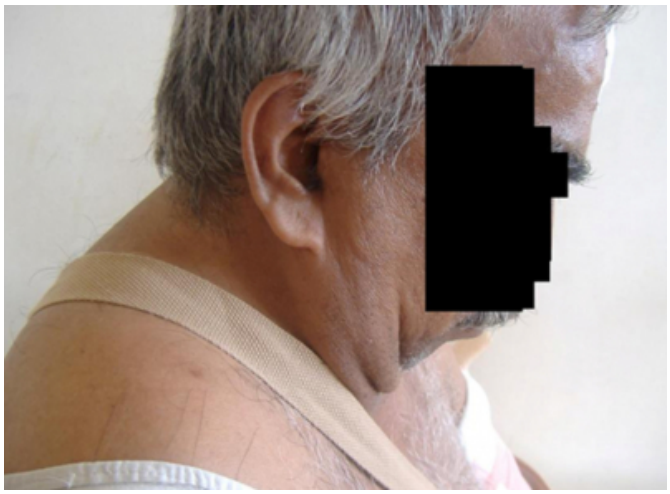


Figure 4

Figure 3: Limited flexion (lateral view)

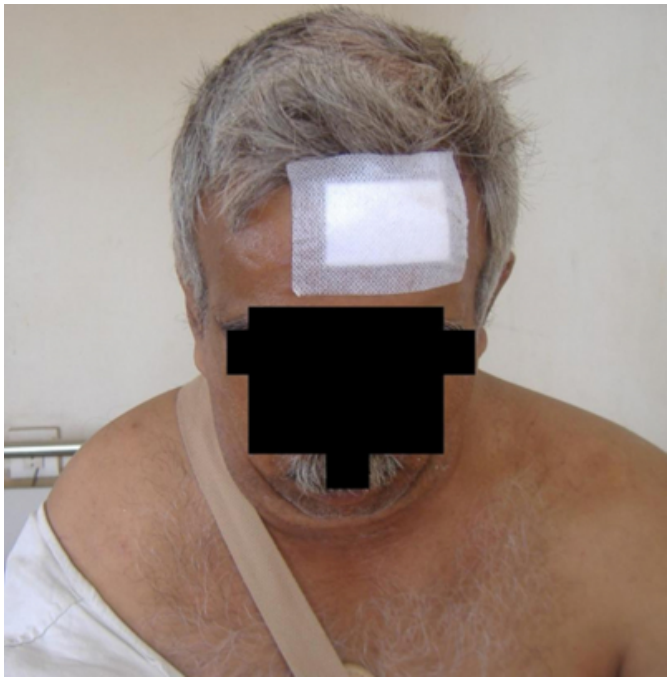
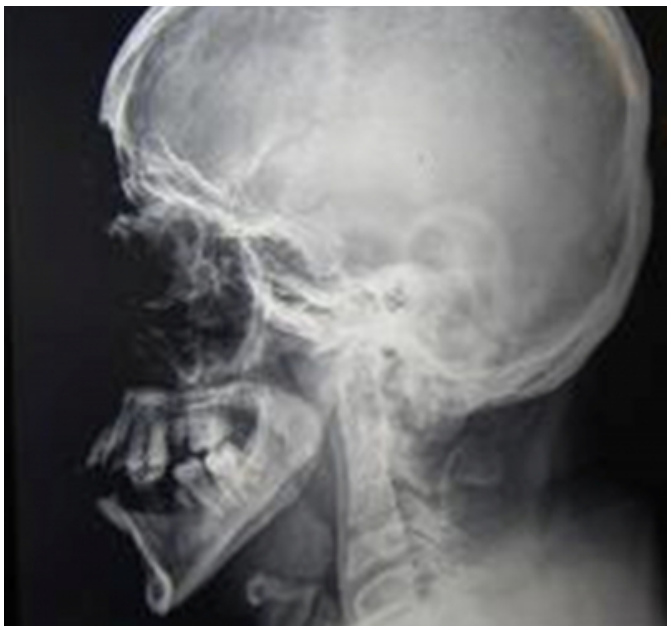


Figure 5

Figure 4: Neck X-ray lateral view showing fusion of upper cervical spines



Since anatomical landmarks in neck were distorted and a difficult airway was anticipated, possibility of a regional anesthetic technique (brachial plexus block) was ruled out. An awake fibreoptic intubation was planned. On the evening before surgery, the procedure was explained to the patient, giving stress to the importance of his co-operation in the success of the procedure. He was kept nil by mouth for 8 hours. The patient was brought to the pre-anaesthetic holding room and an intravenous access secured with a 18G cannula on his right hand. After calculating the maximum dose of local anaesthetic that could be used, patient was given 2% xylocaine viscous gargles which he was allowed to swallow so as to coat the posterior surface of the epiglottis. After 15mts patient was shifted to the operating room where his ECG, SpO_2 and BP were monitored. This was followed by aliquots of a 10% xylocaine spray which was instilled into the posterior oropharynx, with the patient taking deep breaths. Minimal sedation with 1mg midazolam and 50µg fentanyl was given intravenously to keep the patient calm and glycopyrolate 0.2 mg was given to reduce secretions. An oral airway well lubricated with 2% lidocaine jelly, which was slit longitudinally was inserted into the mouth. A 7.0mm ID cuffed endotracheal tube was threaded over the bronchoscope. The scope was then carefully manoeuvred through the slit oral airway until the vocal cords were visualized. 2 ml of 2% lidocaine was injected through the side port of the fibreoptic bronchoscope, using the 'saygo' (spray as you go) technique to anaesthetize the airway and vocal cords. The fibreoptic bronchoscope with the endotracheal tube threaded over it was then introduced into the trachea, the tracheal rings and the carina identified. Propofol 40 mg was given intravenously, the endotracheal tube inserted and its position confirmed after removal of the fibreoptic bronchoscope, by the chest movements as seen on inspection and bilateral equal air entry on auscultation. The $ETCO_2$ further confirmed correct placement of the endotracheal tube which was then fixed properly (Figure 5). The patient was then paralyzed with a non-depolarizing neuromuscular blocker viz: Atracurium and anaesthesia was maintained with isoflurane, 50% nitrous oxide and 50% oxygen, hourly fentanyl 50µg.

Figure 6

Figure 5: After securing the endotracheal tube

Surgery proceeded uneventfully and residual neuromuscular blockade was reversed with atropine $20\mu gkg^{-1}$ and neostigmine $50\mu gkg^{-1}$. The tracheal tube was removed after resumption of spontaneous breathing and return of good muscle tone. Post-operatively the patient was monitored in the recovery room and shifted to post anaesthesia care unit.

DISCUSSION

Klippel–Feil syndrome occurs due to a failure of normal segmentation of the cervical somites in the 3rd to 8th week of gestation. Its etiology is unknown and clinical presentation is varied because of the many associated syndromes and anomalies that can occur[3]. Other axial anomalies include cervical or fused ribs, cleft or hemi vertebrae, kyphosis, scoliosis, spina bifida occulta and sacral agenesis (Gjorup and Gjorup 1964: Winter 1996). Patients also present with facial asymmetry and torticollis[4] which occurs in 21-50% of patients. Decreased range of movement of the neck is the most consistent finding with loss of

rotation being more pronounced than is the loss of flexion and extension. High cervical abnormalities can cause acute spinal cord compression following comparatively minor trauma. A Sprengel's anomaly occurs in 20-30% of patients.. Renal anomalies[5] are common in individuals with Klippel Feil syndrome. Laryngeal cartilages may be malformed, causing aphonia or other voice impairment (Clarke et al 1994). Cardiovascular anomalies[67]occur in 14-29% of patients, the most common being Ventricular Septal Defect. Other less common anomalies are congenital limb deficiencies, craniosynostosis[8], ear abnormalities and craniofacial abnormalities.

Our patient had Feiel classification type1. There were no other associated anomalies.

Syncopal attacks may be precipitated by sudden rotatory movements of the neck in patients with Klippel Feil syndrome. O'Conner and Moysa[9] reported that airway control can be temporarily lost after induction and an LMA may be required to attain control of the airway.

These patients present to us for correction of associated anomalies like scoliosis,spinal canal stenosis, renal anomalies or other unrelated problems(as was the case with our patient). They pose a challenge to the anaesthesiologist with regard to the management of the difficult airway. These patients have a potentially unstable cervical spine and abnormal atlanto-occipital junction and are prone to an increased risk of neurological damage not only during laryngoscopy and intubation but thereafter.

In 1988, Daum and Jones[10] suggested that the most prudent and effective way is an awake fibreoptic intubation. The advantages are:

(1) an awake spontaneously breathing patient can maintain his own airway

(2) a tool for intubation that allows confirmation of tracheal tube placement

(3) no spinal movement is needed during intubation

(4) it has a high rate of success

(5) low rate of complications

(6) good patient acceptance

(7) patient can assist in clearing their own secretions, phonating or panting.

The intubating LMA has also been used to facilitate intubation without manipulation of the head and neck. Keller and Brimacombe[11] suggest that cervical pressures generated by the laryngeal mask devices can produce posterior displacement of the normal cervical-spine. Therefore caution must be taken when dealing with an unstable cervical-spine.

A difficult airway must be approached with caution. A comprehensive preoperative examination and workup, a very co-operative patient, availability of several alternate techniques, willingness to call for expert help, surgeons standing by to provide a surgical airway and/or moral support and a good deal of common sense go a long way in ensuring a favourable outcome.

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