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CASE REPORT

ANAESTHETIC MANAGEMENT OF EMERGENCY CAESAREAN SECTION IN A PARTURIENT WITH KLIPPEL FEIL SYNDROME

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ABSTRACT

A 20 year old pregnant female with Klippel Feil Syndrome was scheduled for emergency caesarian section during her first pregnancy. We present the successful anaesthetic management of this patient highlighting the various anomalies associated with Klippel Feil Syndrome and the presence of a difficult airway.

Key words:

Klippel Feil Syndrome,
Difficult airway,
Subarachnoid block.

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INTRODUCTION

In 1912, Klippel and Feil first reported on a patient with a short neck, a low posterior hairline, and severe restriction of neck movements due to complete fusion of the cervical spine, the classic clinical triad which is the hallmark of Klippel - Feil syndrome (KFS).

CASE REPORT

A healthy 20 yr old women was admitted at term in labour during her first pregnancy (G1P0A0). Investigation done at the time of admission was: Blood group: 0 (negative), Hb: 9.6 gm/dl, Ran-dom blood sugar: 86mg/dl. On examination the patient was a short (height 132 cm), moderately built and nourished with Weight-48kg, b.p.-122/78 mmHg, pulse-84/min, respiratory rate 18/min, no jaundice, mild pedal edema. She had a short webbed neck and a low posterior hairline with no flexion or extension of the neck (Figure-1). Examination of the airway showed that there was no flexion or extension of the neck, she had adequate mouth opening and reduced thyromental distance =3cm and sternomental distance=4.4cm and Mallampatti grade was 3. Electrocardiogram was within normal limits. Radiograph (x-ray taken after delivery of baby) revealed fusion of atlanto axial joint and associated thoracic kyphoscoliosis (Figure-3 and 4).

On auscultation bilateral air entry in lungs were equal, no added sounds S1 and S2 were normal and no murmur was present. On per vaginal examination: 2-3 cm os dilated, semieffaced cervix and Cephalo-pelvic disproportion also revealed. Patient was shifted to emergency OT for emergency LSCS. In O T, pre anaesthetic assessment was done. Patient was fasting for 6 hrs. There was no personal and family history of same problem. She was not taking any medication, and not having history of any drug allergy. The anaesthetic management option in this case included either a general anaesthesia with an anticipated difficult endotracheal intubation or a regional anaesthesia. After a through discussion, we planned to give subarachnoid block as we considered it safe. Emergency tray with direct laryngoscope, fiberoptic laryngoscope, different sizes ETT, ETT stylet, LMA of ap-proprieate sizes, Intubating LMA, Laryngial tube, boogie etc kept ready for intubation because of chances of failure of regional block or high spinal. Surgeon was standby for tracheostomy in condition of failed in-tubation. Intravenous line was secured with 18G cannula. Inj. Ringer lactate was started. In ranitidine 50 mg and Inj metoclopramide 10 mg were given by slow i.v. Injection. Monitors were attached to the patient. Preop vitals: NIBP: 130/86 mmHg, pulse: 88/min, SPO2: 98% on room air. Under all aseptic conditions, subarachnoid block was given in left lateral position at L3 -L4 space using 25G quincke needle with 1.4 ml inj bupivacaine 5% heavy. Just after block patient was given supine position. Effect of subarachnoid block was checked and surgery for LSCS started. Inj-oxytocine 2.5 units stat and 10 units in

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infusion given after extraction of baby. Baby was crying spontaneously with APGAR score of 8. A total of 1500 ml ringer lactat given. Post operative vital was- BP: 122/86 mmHg, PR: 78/ min, RR: 14/min, patient was fully conscious and had no other complain.



Figure 1. Short webbed neck and a low posterior hairline with no flexion or extension of the neck



Figure 2. Mallampatti grading

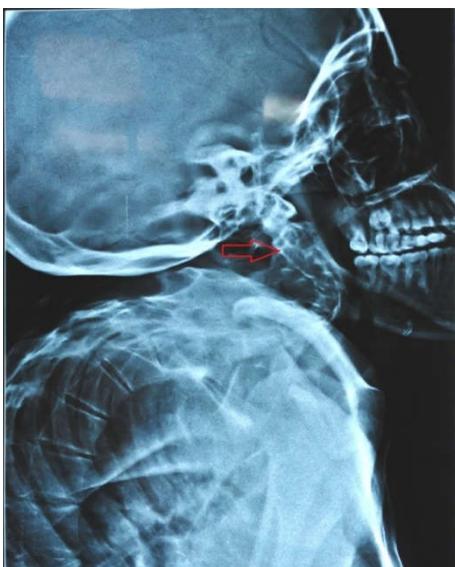


Figure 3. Radiograph of lateral view of cervical spine deviation showing fusion of atlanto-occipital joint and cervical vertebrae



Figure 4. AP view of thorax showing of trachea and marked kyphoscoliosis of thoracic spine

DISCUSSION

Some valuable lessons may be learnt from this case. Klippel feil syndrome is estimated to occur in 1 in 40,000 to 42,000 newborns worldwide. Mutations in the GDF6 and GDF3 genes can cause KFS (<http://ghr.nlm.nih.gov/condition/klippel-feil-syndrome>). But in some people there are no identified mutations in the GDF6 or GDF3 genes and the etiology remains unknown. Mutations in MEOX 1 have been found to occur in association with Klippel-Feil syndrome (Mohamed *et al.*, 2013). Most cases of Klippel Feil are sporadic. Some cases are due to autosomal dominant or autosomal recessive inheritance. It is a rare skeletal disorder primarily characterized by abnormal union or fusion of two or more cervical vertebrae. Other commonly associated anomalies include scoliosis, renal abnormalities, Sprengel deformity, deafness, synkinesia and congenital heart disease. The most common heart disease variant was ventricular septal defect. Less commonly associated are ptosis, lateral rectus palsy, facial nerve palsy and upper extremity anomalies. There are 3 variants of klippel feil syndrome (William *et al.*, 2003).

Type 1 is an extensive abnormality where elements of several cervical & upper thoracic vertebrae are incorporated into a single block. In type 2, failure of complete segmentation occurs at one or two cervical interspaces. Type 3 variant includes type 1 or 2 deformities with co existing segmentation errors in the lower thoracic or lumbar spine. Our patient had type 1 Klippel Feil syndrome with massive fusion of the cervical spines. These patients have a potentially unstable cervical spine and abnormal atlanto-occipital junction and are prone to an increased risk of neurological damage. Syncopal attacks may be precipitated by sudden rotatory movements of the neck in patients with Klippel Feil syndrome. Falk and Mackinnon⁹ reported that airway control can be temporarily lost after induction and an LMA may be required to attain control of the airway. In 1988, Daum and Jones (Daum and Jones, 1988) suggested that the most prudent and effective

way is an awake fiberoptic intubation with good conditions found in awake patients since they can assist in clearing their own secretions, phonating or panting. The nasal route is preferred since the tongue is out of the way and the patient cannot bite down on the tube or scope. However oral intubation may be attempted using an appropriate airway cum bite block like the ovassapian airway. Blind nasotracheal intubation is an alternate mode of securing the airway in difficult intubations (Crosby and Lui, 1990), this technique however is associated with flexion and extension, two maneuvers which are avoided in patients with an unstable spine. There was also a failure in fiberoptic intubation on two separate occasions (Asai and Shingu, 2004). The intubating LMA has also been used to facilitate intubation without manipulation of the head and neck. Keller and Brimacombe (Keller *et al.* 1999) suggest that cervical pressures generated by the Laryngeal mask devices can produce posterior displacement of the normal cervical-spine. Therefore caution must be used when extrapolating these findings to the unstable cervical-spine. It would entail difficulty for regional anaesthesia keeping in view the spinal fusion and scoliosis. The dose of a single bolus of spinal anaesthetic would be difficult to judge in this patient and epidural anaesthesia might prove technically difficult and is associated with an increased risk of inadvertent dural puncture and poor spread within the dural space (Dresner and Maclean, 1995). This patient's abnormality posed problems for all the commonly used techniques for caesarian section. GA could be complicated by difficult intubation while greater use of regional anaesthesia may have reduced the number of deaths due to failed intubation in obstetrical anaesthesia practice. A subarachnoid block for this emergency LSCS was considered as the best first option (with emergency intubation tray and standby surgeon

ready in failed spinal block or failed intubation condition) for delivery of the baby by our team and the case was managed successfully with favourable outcome to both mother & baby.

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