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Case Reports

ANESTHETIC MANAGEMENT IN A CASE OF KLIPPEL-FEIL SYNDROME AND LITERATURE REVIEW

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ABSTRACT

Klippel-Feil syndrome is known by the classic triad of shortness of the neck, limitation of neck movements, and a low posterior hairline. There are often accompanying cervical spinal abnormalities such as kyphoscoliosis as well as urogenital and cardiac abnormalities.

Presented here we have a 20 year old young man with hypoesthesia and decreased motor function in the right hand. The problem began one year back following a minor head trauma and had a progressive course involving the legs, especially the feet. Cervical magnetic resonance imaging was compatible with C3-C4 cord compression as well as blocked vertebrae. The patient was evaluated to be in Mallampati class II. Endotracheal intubation was performed employing gentle manual axial traction in both anterior and posterior operative approaches without any neurological sequela. It is recommended that in situations where fiberoptic or Bullard laryngoscopes are not available and Mallampati class is low, direct laryngoscopy associated with gentle axial traction may be a plausible substitute.

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Keywords: Klippel-Feil syndrome, Endotracheal intubation, Axial traction, Cervical spine instability.

INTRODUCTION

Klippel-Feil syndrome was described for the first time by Klippel and Feil in 1912. The syndrome is associated with short neck and limitation in cervical movements.[1] It is observed in 0.2 to 4.2 neonates out of 1000 live births. In some patients, cervical spine abnormalities associated with kyphoscoliosis, and urogenital abnormalities and cardiac anomalies are observed.[2]

These patients may show neck rigidity and limited neck extension due to vertebral fusion, making endotracheal intubation a difficult and potentially hazardous task.[1,3,4] The patients are susceptible to spinal cord injuries and neurological deficits after laryngeal intubation[5] with recommendations by some to avoid induction of general anesthesia by intravenous agents in these particular patients if possible.[6]

However in patients in whom general anesthesia is warranted, various methods such as awake intubation, fiberoptic laryngoscopy,[1,7] Bullard laryngoscopy,[8] cricothyroidotomy and transtracheal ventilation[9] have been suggested. Airway examination in our case revealed

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the soft palate and the base of the uvula, but not the tonsillar fauces (Mallampati class II).[10]

Our patient was intubated twice for two different surgical approaches under general anesthesia, employing axial in-line traction and direct laryngoscopy. The intubation had been relatively easy and uneventful.

CASE REPORT

A twenty year old man complaining of mild weakness and paresthesia in all the four limbs was admitted at this center in Sept. 2002 for operative treatment of the cervical spine. The illness started one year back and had a progressive course necessitating his admission. There was also a positive history of minor head trauma at the onset of the symptoms. Plain radiographs revealed fusions at C3 to C4 levels (Fig. 1), and magnetic resonance imaging showed C3-C4 disc herniation and canal stenosis at the same level causing severe cord edema (Fig. 2). A positive family history for such conditions was not forthcoming. The patient had a low hairline associated with neck deformity and an apparent cervical scoliosis (Fig. 3). There was limited neck extension (<50°) associated with a positive Lhermitte's sign,[11] Flexion and rotation were however not limited. Airway examination revealed Mallampati class II. Thyromental span was 8 cm and normal. No dental abnormality or occlusion was present. Cardiopulmonary and genitourinary evaluation proved to be normal.

General anesthesia for cervical laminectomy and posterior fusion was performed employing a balanced standard anesthesia under conventional monitoring devices. Prior to induction, diazepam 7.5 mg and fentanyl 100μg were administered. Induction was conducted using thiopental sodium 300mg and succinylcholine 80mg was used to achieve muscular relaxation, Lidocaine 90mg was injected soon after succinylcholine to ensure hemodynamic stability. The head traction was applied gently by an attending neurosurgeon and laryngoscopy conducted with a Macintosh blade 3 (Riester, Germany) employing a No.8 cuffed Roche(TM) orotracheal tube. At laryngoscopy the posterior extremity of the glottis was visible and intubation was successful during the first attempt. After securing the orotracheal tube, the patient was rotated to the prone position with necessary precaution and maintenance of anesthesia managed with halothane

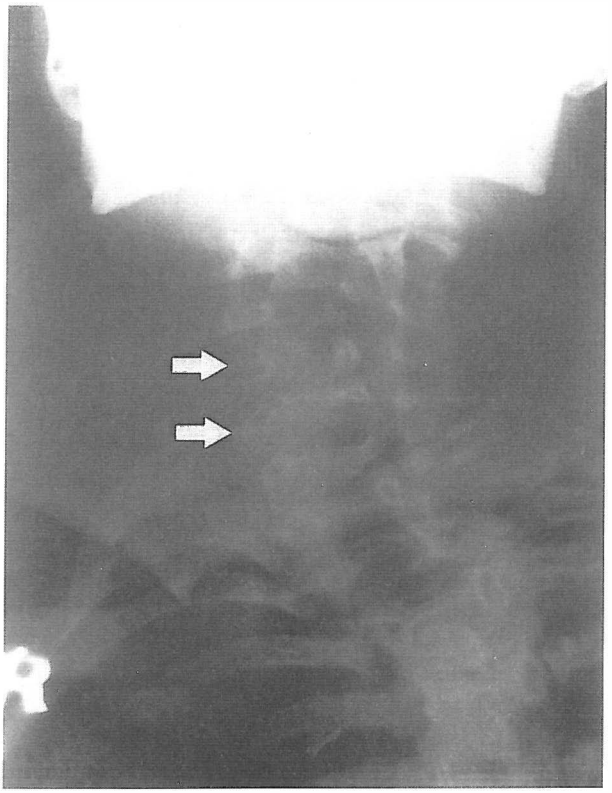

Fig. 1. A plane radiogram revealing C3-C4 instability associated with blocked vertebrae at superior and inferior levels.

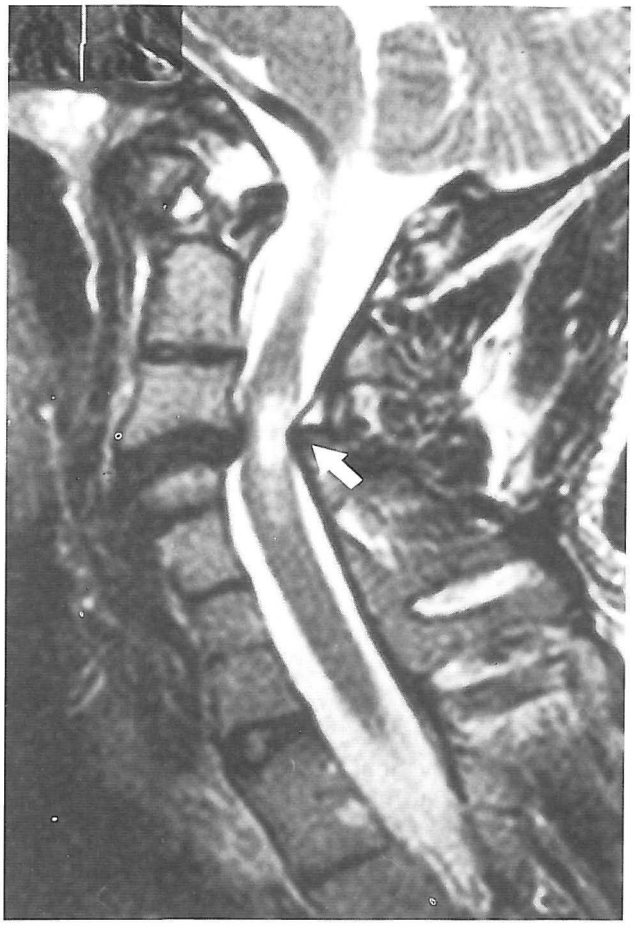

Fig. 2. Magnetic resonance weighted image of cervical spine revealing C3-C4 disc herniation associated with narrow canal at the same level. Note increased signal intensity in the spinal cord in favor of cord edema.

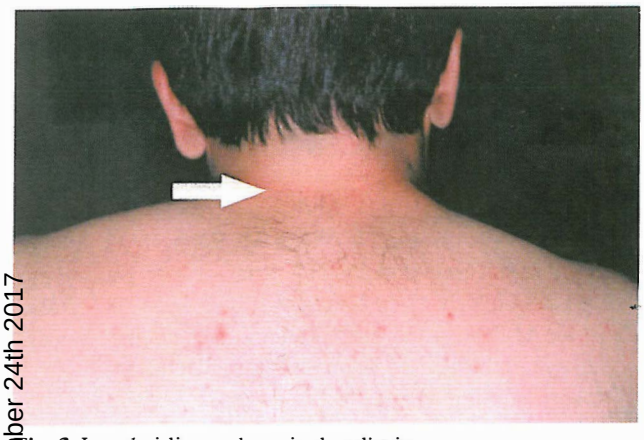

Fig. 3. Low hairline and cervical scoliosis.

0.6%, a 50% mixture of N_2O and O_2 along with incremental doses of pancuronium bromide and fentanyl. After completion of the surgery, which lasted 120 min, the patient was returned back to the supine position and residual muscle paralysis reversed with neostigmine 2.5mg and atropine 1.2mg. Extubation of the trachea was accomplished after ensuring that the patient was awake and had adequate tidal respiration. The same anesthetic protocol was again repeated 14 days later for anterior discectomy and fusion with no sequela except mild sore throat.

DISCUSSION

Patients with Klippel-Feil syndrome are prone to major neurological sequela following minor trauma due to cervical spine instability.[3,12] C_2-C_3 and C_5-C_6 are the most commonly involved interspaces. However multisegment involvement is not uncommon.[13] Our patient had his C_2-C_3 disc space involved.

General anesthesia may become a necessity for these patients and a thorough physical examination is essential to unveil any potential risk. The patients pose the anesthetist to a high risk of difficult intubation as well as spinal cord damage due to instability.[14] Manipulation of the neck or an attempt to extend the neck during laryngoscopy and thereafter must be carefully controlled if neurological damage is to be avoided.[6,15]

Despite the afore mentioned problems encountered during intubation, there still does not exist a formal consensus on an intubation technique in these cases. Several options such as awake laryngoscopy under local anesthesia,[16] inhalation induction with halothane,[6] insertion of a laryngeal mask airway (LMA) and oral fiberoptic intubation (FIB),[17] have been advocated as possible alternatives in patients with Klippel-Feil syndrome. The most commonly advocated technique in these cases has been flexible fiberoptic laryngoscopy. The technique is safe but difficult at times and entails some time. At times Bullard laryngoscopy has been proposed to be safe, however it is not always available and remains ubiquitous in operative room settings.[8] Awake intubation may be a safe method but it may increase intracranial pressure[17] and furthermore there is limited view for the anesthetist.[9] Cricothyroidotomy is considered to be a good choice[9] but extubation remains a dilemma and moreover there may be an intrusion in to the operative field.[18] Some authors prefer direct laryngoscopy coupled with cervical stabilization procedures.[9] Direct laryngoscopy remains the cornerstone for safe and fast tracheal intubations.[8,9,18,19] In this method minimal surgical intervention is necessary and could be employed in difficult cases. Conspicuous atlanto-occipital joint extension can occur during intubation that may be hazardous for the spinal cord.[9] Therefore patients with C_1-C_2 instability are the most susceptible cases and axial-in-line traction has been recommended for these cases,[20] however there will remain a small but negligible probability for spinal cord damage.[18] Cadaver studies confirm this hypothesis.[9] On the other hand, traction maneuver may limit the oroglottic alignment and make intubation more difficult, but there is no evidence showing neurological worsening following intubation procedures under external head and neck stabilization.[9]

In our patient, laryngoscopy was performed under axial-in-line manual traction. During the first operation, intubation could be performed on the first attempt, but during the second operation, the intubation was successful after a second attempt and light pressure on the larynx was needed to accomplish the task. The patient on laryngoscopy had a Cormack and Lehane grade 2[21] which did not cause much of a problem during the first attempt, but caused some difficulty during the second time. The difficulty during the second time could be attributed either due to the cervical laminectomy and posterior fusion performed during the first stage which had made laryngoscopy difficult or perhaps an extra caution on our part to avoid cervical spinal cord damage.

Because of predicted difficulty with tracheal intubation in patients with Klippel-Feil syndrome, it is indispensable to secure the airway before induction of general anesthesia especially in emergent cases because emergency endotracheal intubation is normally considered to be difficult because of the altered anatomy and reduced neck mobility.[1,14] In our patient the intubation was successful because earlier prediction of the airway was found to be easy, the lesion level was at C_3-C_4 interspace away from the atlanto-occipital region and above all the procedure was cautiously accomplished under axial traction. Although FIB has been recommended in such cases, this modality might fail,[10] cause massive subcutaneous emphysema[22] or cause airway injury as

reported by Maktabi et al.[23] in three of their cases.

It could however be argued that if the scenario had progressed to a "can't intubate - can't ventilate" situation in this particular patient after having been paralyzed, and intubation had proved impossible, what would have been the plan B in our minds. Under such circumstances, we would have opted for an intubating laryngeal mask airway (ILMA) since an overall success rate of 100% has been reported with it.[24] A similar success rate has also been reported with FIB, but oxygen desaturation occurred more frequently with it.[24]

Our patient was to undergo an elective surgical procedure which provided us with ample time to assess the air-way and thus enabled us to opt for a method that would not only prove to be safe for the patient but at the same time would be easy to accomplish.

Finally, it is recommended that where fiberoptic or Bullard laryngoscopes are not available and airway class seems promising and easy, direct laryngoscopy associated with cautious manual axial traction under general anesthesia might be a plausible alternative in such cases.

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