Case Report

Anesthetic management of laparoscopic pyeloplasty in a Klippel-Feil syndrome patient with aortic coarctation: A multidisciplinary approach

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ABSTRACT

Klippel-Feil syndrome (KFS) is a rare congenital condition characterized by fusion of cervical vertebrae, often associated with other systemic anomalies, such as cardiac malformations and renal abnormalities. These patients pose significant anesthetic challenges due to difficult airway anatomy, potential cervical spine instability, and associated comorbidities. Beyond airway difficulty, KFS patients often have systemic issues further compounded by physiologic changes from laparoscopy, requiring tailored anesthetic planning. We present here anesthetic management of laparoscopic pyeloplasty for a solitary functioning kidney with pelvic-ureteric junction obstruction in a patient with recently diagnosed KFS with aortic coarctation. This case highlights the critical importance of thorough pre-operative assessment, meticulous planning, and a multidisciplinary team approach. Early identification of hidden abnormalities, preparedness for airway challenges, and collaborative perioperative care are vital to ensure safe outcomes in patients with complex syndromic presentations, such as KFS.

Key words: Coarctation of aorta, Difficult airway, Klippel-Feil syndrome, Laparoscopic pyeloplasty, Solitary kidney

Klippel-Feil syndrome (KFS) was originally defined by Klippel and Feil in 1912, characterized by a triad of short neck, severe limitation of head movement, and low posterior hairline. Its incidence is 1:42,000 births and is one of the common congenital causes of a difficult airway [1]. This syndrome may also be detected as an incidental finding. The main anesthetic challenge is the potential spine abnormality, prone to an increased risk of neurological damage during airway management [2], and managing other congenital abnormalities associated with this syndrome.

As the literature is scarce regarding the perioperative management of laparoscopic surgeries in patients of KFS with Coarctation of the Aorta (CoA). To the best of our knowledge, fewer than 50 studies were published on this topic between 1974 and 2023 [3], with only <5 addressing anesthetic management during laparoscopic surgery in KFS patients and none presenting a combination of KFS with CoA and laparoscopy. We report and discuss here our experience of managing an initially undiagnosed case of KFS with other multiple associated congenital anomalies for laparoscopic pyeloplasty and emphasize the fact of a multidisciplinary team approach. No established anesthetic management guidelines currently exist for this patient population. The anesthetic plan for this particular patient was formulated through a meticulous assessment of the potential interplay between anesthesia, surgery, and his underlying syndrome.

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CASE REPORT

A 45 kg, 16-year-old male was scheduled for right laparoscopic pyeloplasty for a solitary functioning kidney with pelvic-ureteric junction obstruction. He complained of episodes of headache that got relieved on taking analgesic drugs. He had no significant past history with good functional activity.

His pulse was 98/min. Blood pressure (BP) was 160/96 mmHg in the left upper limb. Cardiac auscultation revealed a holosystolic murmur over the left parasternal area. On airway assessment, the mouth opening was 1.5 fingers, Mallampati grading was IV, thyromental distance of 1 cm, neck extension and flexion severely restricted <10º (Fig. 1). Spine examination revealed cervical and upper thoracic ankylosis and scoliosis. It was further observed that the patient had a short webbed neck and a low posterior hairline (Fig. 2).

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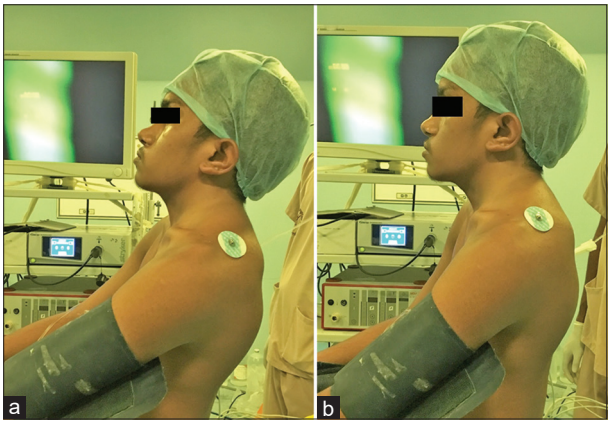

Figure 1: (a) To assess neck extension, when the patient was asked to look at the ceiling, he used his whole spine and raises his eyes (neck extension <10⁰); (b) severely restricted neck movements, both flexion and extension

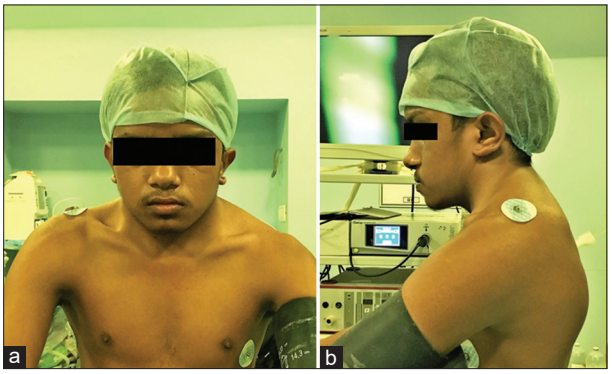

Figure 2: Short webbed neck (a), low posterior hairline (b)

We suspected our patient was an undiagnosed case of KFS. Routine blood and biochemical investigations were normal. Pulmonary function tests revealed mild restriction. The electrocardiogram (ECG) showed sinus tachycardia. Cardiology consultation evaluated differences in BP, in all limbs (Left upper limb-150/94 mmHg, right upper limb-148/90 mmHg, right lower limb-100/64 mmHg, and left lower limb-107/64 mmHg), bilateral feeble femoral pulses, and radio-femoral pulse delay pointing to CoA. Doppler echocardiography revealed CoA with a pressure gradient of 55 mmHg. Descending thoracic aortoplasty with bare metallic stent (BMS) was planned by a multidisciplinary team consisting of a cardiologist, a nephrologist, and an urologist ahead of pyeloplasty after considering the risk-benefit ratio (Fig. 3). Post-procedure pressure gradient reduced to 14 mmHg. The patient was then started on tablet ecosprin 75 mg, clopidogrel 75 mg, and amlodipine 5 mg.

As the patient had a solitary functioning kidney, organ saving, percutaneous nephrostomy was done under local anesthesia (LA), and the patient was discharged with advice to follow up in the genetics and pediatrics department for further examination, which later confirmed our suspicion and diagnosed the patient with KFS. After one month, the patient was readmitted, and clopidogrel was stopped before the definitive surgical procedure.

Awake fiberoptic intubation (AFOI) was planned. The patient was nebulized with 4% lignocaine in the pre-operative area. In the operating room, routine monitors

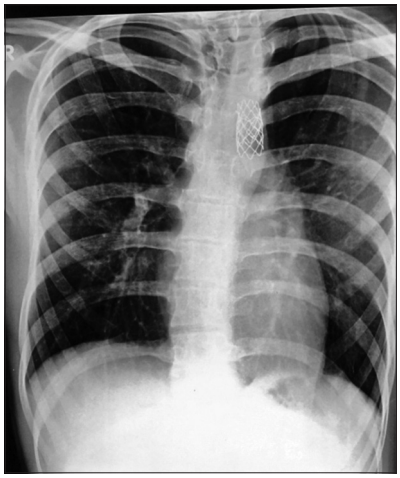

Figure 3: Thoracic scoliosis with bare metallic stent *in situ*

(ECG, pulse oximeter, and non-invasive BP in the lower limb) were attached. Intravenous midazolam 1mg and fentanyl 50 µg were administered. The right radial artery was cannulated under LA. A difficult airway cart was kept on standby. Oxygen at 5 L/min was given through the opposite nostril using a nasal cannula. Airway blocks were given, a local anesthetic spray was done in the oral cavity, and the trachea was intubated with the AFOI technique, followed by induction of anesthesia with fentanyl 100 µg, propofol 80 mg, and vecuronium 5 mg. Anesthesia was maintained with sevoflurane in an oxygen-air mixture. Pressure-controlled ventilation was used with the following settings: Peak inspiratory pressure of 20, positive end-expiratory pressure of 5, and respiratory rate of 16. Ondansetron 4 mg, dexamethasone 8 mg, and paracetamol 1 g were all administered intravenously as routine intraoperative medications. The surgery was performed in the left lateral position. Adequate padding and positioning of the patient was taken care of with special care to the scoliotic and ankylotic spine. The pneumoperitoneum was maintained at 8–10 mmHg. The surgery went uneventfully for 2 h. Intraoperative fluid administered was normal saline 1000 mL. At the end of the surgery, a right transversus abdominis plane block and left rectus sheath block were administered using 15 mL and 5 mL, 0.25% bupivacaine, respectively. For renal protection, nephrotoxic agents, such as non-steroidal anti-inflammatory drugs were avoided. Residual neuromuscular blockade was then reversed, and the patient was extubated. He was observed in the post-operative room and then shifted to the ward.

He was discharged home on the 3rd day, with advice to follow up in the cardiology and urology departments.

DISCUSSION

Our patient was an undiagnosed case of KFS, an autosomal dominant or recessive disease, due to abnormal fusion of two or more cervical vertebrae, leading to a classical triad of short neck, low posterior hairline, and severe restriction of neck movements. These anomalies can lead to chronic headaches and cervical spinal stenosis,

deformity, and instability. Other associated anomalies include scoliosis (60%), Sprengel's deformity (35%), cardiac abnormalities (4.2–14%), such as ventricular septal defect, CoA, and Genitourinary abnormalities, such as a single kidney [4]. All these anomalies were coincidentally present in our patient along with the classic triad, which led us to suspect the syndrome. Pediatrician evaluation confirmed our suspicion and diagnosed the patient with KFS. Cervical vertebrae and the genitourinary system differentiate during the same embryological period; therefore, urinary system abnormalities may be encountered in 30–35% of KFS cases, the most common being unilateral renal agenesis [5]. Our patient had a solitary functioning kidney along with CoA.

Our patient presented with a history of chronic headache, an incidental holosystolic murmur with differences in BP in all limbs, which on evaluation was diagnosed as CoA. Diagnosis of CoA is often delayed until the patient develops congestive heart failure, which is common in infants, or hypertension or an incidental murmur, which is common in older children [6,7]. As our patient was of the younger age group with high-pressure gradient, it was decided by the multidisciplinary team to proceed with BMS ahead of pyeloplasty. BMS achieves superior immediate relief of pressure gradient, with lower risk, and is cost-effective [8].

In CoA, there is limited flow across a stenotic outflow. The main concern during laparoscopy in such patients is increased intra-abdominal pressures, resulting in a fall in pre-load, an increase in systemic vascular resistance and afterload, a decrease in stroke volume and blood flow distal to the coarctation [9]. We maintained pneumoperitoneum at 8–10 mmHg. Low-pressure laparoscopy reduces early pain scores, post-operative nausea and vomiting, and the length of stay [10]. Creation of pneumoperitoneum decreases the blood flow distal to the coarctation. It is prudent to measure the perfusion of the body by measuring arterial blood pressure both proximal and distal to the coarctation [11]. We measured invasive BP in the upper limb and non-invasive BP from the lower limb.

The anesthetic goal aims to manage the difficult airway due to anatomical and pathological changes associated with the congenital anomalies and maintain higher arterial BP proximal to coarctation, increasing pre-load, avoiding tachycardia, and maintaining contractility to optimize the cardiac output and mean BP distal to coarctation above 60 mmHg. Although our patient had a BMS implanted, we took all precautions and maintained hemodynamics as per anesthetic goals.

Ankylosis and scoliosis involving the cervical and upper thoracic spine pose significant challenges in airway management. Daum and Jones [12] suggested that the most prudent and effective way is an awake fibreoptic intubation in KFS patients. AFOI, regarded as the gold standard in such cases, was employed to secure the airway in this patient with cervical spine pathology. In the event of AFOI failure, contingency plans included Glidescope video laryngoscope-guided intubation with manual in-line stabilization under sedation and emergency surgical airway access as last-line salvage intervention.

As surgery was performed in the left lateral position for pyeloplasty, positioning of the patient with an ankylosed and scoliotic spine is another challenge and requires utmost care. Adequate padding and positioning of the patient were done to prevent any neurological injury. Regional anesthesia was avoided as the surgery was planned laparoscopically, and the distorted vertebral anatomy with a likely compressed epidural or intrathecal space could render neuraxial techniques unreliable [13]. General anesthesia with endotracheal intubation was preferred for airway control, aspiration risk reduction, and better ventilation. Chua and Cruz similarly opted for AFOI with general anesthesia in a KFS patient undergoing laparoscopic pelvic surgery [3]. Similarly, in a case report where a KFS patient was posted for the left forearm fracture surgery, distorted neck anatomy and anticipated difficult airway precluded regional anesthesia; AFOI was chosen by Alex and Rao [14].

CONCLUSION

Suspected syndromic patients require thorough pre-operative evaluation and screening for associated anomalies, as hidden abnormalities may be present. Vigilance is essential not only for airway management but also for patient positioning and anticipating reduced respiratory reserve. Understanding the altered physiology in patients of CoA needs to be understood while caring for these patients perioperatively. During laparoscopy, intra-abdominal pressure should be maintained below 12 mmHg to minimize hemodynamic effects while ensuring adequate surgical exposure. Effective perioperative management relies on close coordination among cardiologists, pediatricians, surgeons, and anesthesiologists, underscoring the importance of a multidisciplinary team approach.

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