Case Report

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Anaesthetic Management of Klipple Feil Syndrome- Real Challenge in Rare Case

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Abstract

Rare cases in gynecological surgery always pose challenge to anaesthetist. Klipple Feil Syndrome stands high in the list. It is a rare skeletal disorder primarily characterized by short neck with decreased movements and low posterior hairline. It is believed to be caused by faulty segmentation of mesodermal somites. It is associated with other anomalies like cardiac and renal anomalies, deafness, scoliosis etc which make the anaesthetic management difficult.The technique of anaesthesia decides the outcome of patient. Here we present two cases of Klipple Feil Syndrome with skeletal and renal anomalies posted for laparotomy for malignant ovarian mass done successfully under regional anaesthesia and emergency LSCS done under general anaesthesia.

Keywords: Klipple Feil Syndrome; Multisystem Involvement; Laparotomy; LSCS; General Anaesthesia; Regional Anaesthesia.

Introduction

Klipple Feil Syndrome comprises of short neck with restricted neck movements and low posterior hair line;the triad being hallmark of the disease.Various types of Klipple Feil Syndrome are seen as per inheritance and mutation. Proper technique of anaesthesia in patients with multiple anomalous systemic involvement makes it a real challenge.

Here we are presenting a case of Klipple Feil Syndrome posted for removal of huge ovarian mass done successfully under regional anaesthesia and another case of Emergency cesarean section done under general anaesthesia.

Case Report

A 60 years old female patient was posted for exploratory laparotomy for malignant ovarian tumour. Patient was nullipara with primary amenorrhea .She did not have any significant past history of medical, surgical or anaesthetic exposure.

On physical examination she was short stature (140cm) with bilateral webbed neck and limited neck movements.Mouth opening was Mallampatti grade II. She had low posterior hair line. Her other vitals were normal. Her examination of neck and back revealed hypoplasia of left scapula.Both scapulae were placed high in thorax with medial border everted (Sprengel's deformity),scoliosis with concavity to left in the thoracic region. No other obvious skeletal anomaly was found. No facial anomaly was seen. On auscultation her respiratory and cardiovascular systems were unremarkable.

Her Laboratory reports such as Hemogram, Random Blood Sugar,LFT,KFT,Sr.electrolytes were within normal limits. Electro-cardiogram did not reveal any abnormality. Her chest X ray PA view revealed scoliotic deformity with concavity to left, bony rib deformity bilaterally with cervical rib to left. There was evidence of old healed fracture of rib with generalized osteoporosis. X-ray cervical spine was suggestive of short neck, multiple congenital block vertebrae at C5-C6 &C6-C7 level. X-ray lumbar spine was suggestive of osteoporosis. Anticipating a difficult airway an indirect Laryngoscopy was advised which was found to be WNL. Among the special investigation CA 125 was 528 U/ml, suggestive of overian malignancy .

Intravenous urography revealed ectopic malrotated pelvic right kidney with grade 2 hydronephrosis secondary to calculi, one in superior calyx & another in pelvis with slightly delayed excretion. Grade 2 to 3 hydronephrosis on left side with displaced renal pelvis and ureter laterally secondary to soft tissue opacity. CT Scan of abdomen and pelvis was suggestive of neoplastic

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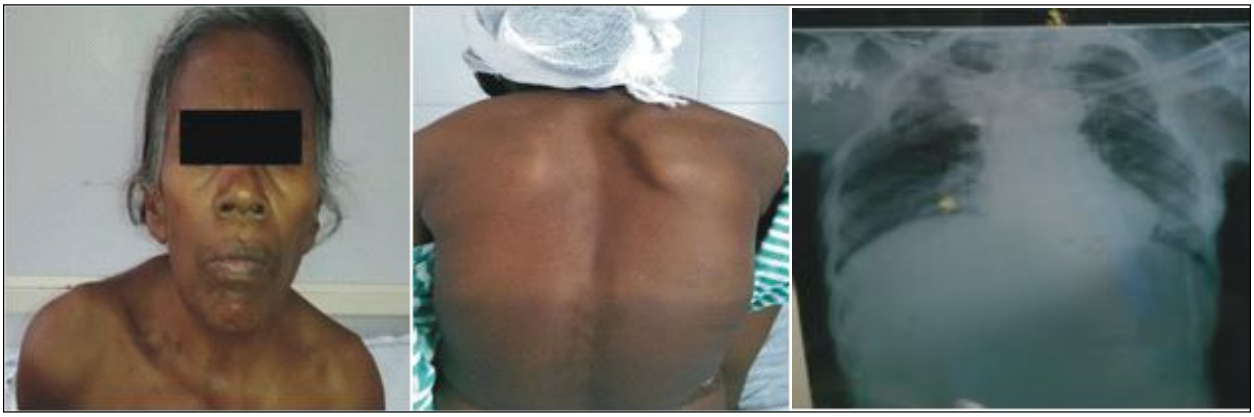
mass in left adnexa mostly ovarian in origin. Left ureter was compressed due to mass causing hydronephrosis. Ectopic right kidney was seen with moderate hydronephrosis due to renal calculi.CT findings were confirmed on MRI of abdomen and pelvis.

Patient was posted for excision of mass with hysterectomy. After taking written informed concent, antiemetic prophylaxis was done with IV 50 mg Ranitidine and IV 4mg Ondensetron. Multipara monitor with Pulse Ox, ECG ,NIBP was attached. Preloading was done with IV Ringers Lactate. Difficult intubation trolley was kept ready. Under all aseptic precautions, epidural catheter was inserted at L1-L2 space. 2CC of 2% lignocaine was given as test dose. Spinal puncture was done in L3-L4 space and 2.4cc of 0.5%Bupivacaine with 0.4ml(20µg)Fentanyl (total volume 2.8ml)was administered. The sensory block was achieved up to T6 level after 4 min. O_2 supplementation on mask was started. Surgical removal of 3Kg ovarian mass was undertaken but hysterectomy was not done due to agenesis of uterus. Both fallopian tubes were abnormally placed in pelvic wall. Total surgical time was one hour 10 min. No epidural top up was required. Epidural Catheter was removed postoperatively after giving Tramadol 50 mg through it for postoperative analgesia. Postoperative period was uneventful. Patient was haemodynamically stable throughout. Histopathological report was suggestive of adenocarcinoma ovary.

Second patient was 21 years primigravida with 126 cm height posted for emergency LSCS in view of severe pre-eclampsia and cephalopelvic disproportion and fetal distress. She had Sprengel's shoulders, short neck with restricted movements, low posterior hairline and congenital hypoplasia of right hand.There was no other obvious anomaly. Her past and family history was not significant. In her lab reports she had urine albumin 2+. Her heart rate was 90/min regular rhythm and BP was 140/112mmofHg.She had pedal edema but no epigastric pain or headache. She was given General anaesthesia for severe pre-eclampsia and drop in fetal heart rate.She was induced and intubated on IV 80 mg Propofol. Intubation was possible in single attempt. She was maintained on O2 ,Isoflurane and IV Atracurium as muscle relaxant given after delivery of baby.2.5 Kg baby was delivered with apgar score 9. Her perioperative period was uneventful.

Fig. 1: showing Bilateral webbed neck,Sprengel's anomaly.Cervical rib andscoliosis.



Discussion

Klipple Feil Syndrome (KFS) is an autosomal dominant disorder inherited by GDF6 gene on chromosome 8. The inheritance and mutation decides the type of Klipple Feil Syndrome. Three different types of KFS are suggested by Maurice Klipple and Andre Feil in1912 as follows:

Type I-Massive fusion of cervical spine.

Type II-Fusion of one or two vertebrae.

Type III-Presence of thoracic or lumbar spine anomalies along with Type I or Type II of KFS which was seen in our first patient [1].

Few more variations have also been suggested. Nagib et al [2] described three types and related the incidence of neurologic symptoms to each type as follows:

Type I - Two sets of block vertebrae with open intervening spaces that can sublux gradually or with acute trauma.

Type II - Craniocervical anomalies with occipitalization of the axis and basilar invagination; which causes increased mobility at the craniocervical level and can lead to foramen magnum encroachment. It can be associated with Arnold-Chiari malformation and syringomyelia.

Type III - Fusion of one or more levels with associated spinal stenosis. The syndrome is commonly associated with other congenital anomalies like scoliosis(incidence 60%) [3], Sprengel anomaly (raised and everted medial surface of scapula) [4] seen in 30% of patients, renal anomalies (incidence 35%), absent ribs [5] or extra rib which was seen in our patient. Along with this, patient may have deafness (incidence 25-50%), synkinesis (20%), congenital heart disease (14%.) commonest being ventricular septal defect, genitourinary, gastrointestinal and neurologic anomalies[6].

Though the true incidence of KFS is unknown, prevalence has been estimated at 1 in 50000 whereas female to male ratio is 1.3:1.

Klipple Feil Syndrome pose challenge to anaesthetist with regard to management of difficult airway due to restricted movement of neck, spinal deformity and multisystem involvement. Fibreoptic intubation becomes the most prudent and effective way of securing airway with high success rate and low complications [7]. Though majority of cases have been reported where general anaesthesia was given.Other methods like nasal or blind nasal intubation carry their own risk due to unstable spine. Manipulation of head and neck can be avoided by using intubating LMA but the cervical pressure generated by the laryngeal mask device can cause posterior displacement of normal cervical spine [8]. However general anaesthesia is not without risk due to restricted neck movements, scoliosis and rib anomalies which can alter the lung dynamics. We gave general anaethesia for emergency LSCS without any difficulty in intubation or postop outcome.

Another suggested and preferred alternative technique is regional anaesthesia.Sprengel deformity, kyphoscoliosis and lack of vertebral segmentation also make neuraxial block challenging. It also carries potential need for emergency tracheal intubation in the event of high block, failed block or local anaesthetic toxicity.

Limited literature is available regarding the use of regional anaesthesia in KFS[9]. Our first patient had restricted neck movements, scoliosis and rib anomalies, Sprengel deformity, renal anomaly without cardiovascular anomaly. So taking into account patient's safety and nature of surgery we decided to give combined spinal epidural anaesthesia. Fentanyl was added to prolong the duration of spinal block and epidural was planned for extension of block.

The other concerns regarding neuraxial block is the suboptimal position due to fusion of cervical vertebrae and scoliosis. It transfers the altered mechanical force making the adjacent non-fused segments excessively mobile. The associated spine anomalies like scoliosis and exaggerated lumbar lordosis make the intrathecal spread of drug unpredictable. Hence careful positioning is mandatory in such patients. In our case we successfully managed the patient in combined spinal epidural anaesthesia in single attempt.

Conclusion

A comprehensive and meticulous preoperative evaluation with good work up, availability of alternative technique and good coordination between surgeon and anaesthetist go a long way in ensuring favourable outcome of the patients with Klipple Feil Syndrome.

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