

Perioperative Management Of The Case Of Klippel Feil Syndrome Operated For Cervical Spine

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Abstract

Klippel feil syndrome is a rare entity. After its first description in 1912 the management of this syndrome still poses a big challenge to clinicians, orthopaedic surgeons, anaesthetist and intensivists. The syndrome is characterised by patient with short neck, low hair line, decreased cervical motion. 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. We came across a patient in KEM hospital, 18 years old male, suffering from this syndrome. We will like to share our experience in his management which was carried out by us perioperatively.

CASE REPORT

An 18 years old male patient, with short neck and Sprengel shoulder, presented to us with complaints of spastic severe quadriplegia and inability to ambulate without support of 4 months duration, without any other systemic involvement. On preoperative assessment, difficult intubation was anticipated with MPC grading IV. All routine investigations were within normal limits. X-ray neck AP/Lateral showed fusion of second and third cervical vertebra. Cervical canal stenosis was detected on MRI spine. 2D Echo, baseline ABG were within normal limits. Right side ectopic kidney was an accidental finding on USG abdomen-pelvis.

With adequate premedication patient was given routine general anesthesia. Difficult intubation tray was kept ready for anticipation of difficult intubation. Laryngoscopy and intubation were uneventful. The anesthesia maintained on oxygen and N₂O 60:40 and vecuronium. All haemodynamic parameters were meticulously monitored and kept within normal limits. Decompression was done by enlarging foramen magnum. Occipito cervical wiring (occiput to C2 cervical spinous process) was carried out. Duration of surgery was 7 hours, with blood loss of 1 L which was replaced. After confirming an acceptable ABG patient was shifted to ICU for observation on T-piece.

Figure 1



COURSE IN THE INTENSIVE CARE UNIT

The patient maintained saturation for 10 hours, after which he was put on volume SIMV mode for desaturation.

Supportive treatment consisting of appropriate antibiotics, analgesics, sedatives, water bed, prophylaxis for DVT, chest physiotherapy and postural drainage were provided.

Cervical joint immobility was assured with cervical braces. In view of prolonged stay in ICU unit, tracheotomy was done.

Figure 2

Day in ICU	Course in ICU	Significant Ix reports	Measures undertaken
1 st day	Patient conscious, power in both limbs 1/5		
4 th day	Neurologically same anticipated prolong ventilation		tracheostomy
7 th day	Spiking fever, neurologically same		Culture sensitivity was done.
10 th day	Fever, systolic BP <90mmHg, neurologically same		Change of antibiotics, dopamine support
14 th day	Resolution of fever and hypotension, neurologically same.		Continuation of antibiotics
3 rd week	Spiking fever, neurologically same.		Culture sensitivity report, change of antibiotics
4 th week	Spiking fever, hypotension, generalized edema, neurologically same.	Albumin 1.2 gm% INR – 2.4	Support on dopamine, noradrenalin, albumin and FFP transfusion
5 th week	ARDS, altered mentation	X-ray chest- fluffy exudates on B/L lung fields	Shift on pressure SIMV
6 th week	ARDS, oliguria, patient comatose		
45 th day	Death		Postmortem report- multiorgan dysfunction

DISCUSSION

Patients with Klippel Feil syndrome usually present in childhood. The syndrome is characterized by short neck, low hair line, decreased cervical movement. 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.

- 3 grades :
- 1: a massive fusion of cervical spine
 - 2: fusion of half of vertebrae
 - 3: presence of thoracic or lumbar anomalies with 1 or 2.

The pattern may involve more than one level giving rise to wasp waist sign which is a valuable radiological sign of Klippel Feil syndrome(6), but was not seen in our case. Cervical spondylosis, disc herniation and secondary degenerative changes are seen more at levels adjacent to fused vertebra(7).

ASSOCIATED CONDITIONS

Affected patients manifest a wide variety of anomalies.

Cervical anomalies have a variable influence on the shape of the neck. The shortening of the neck may be variable giving rise to web neck appearance (“pterygium colli”). Sprengel anomaly (High lying shoulder) may be unilateral, or bilateral (present in up to 30% of individuals), or familial (Scott 1993; Larson et al 2001).

Sprengel and Klippel-Feil anomalies have been associated with a unique point mutation (Pro250Arg) in fibroblast growth factor receptor 3 in some, but not all, cases (Lowry et al 2001). 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). Cervical ribs are recognized clinically, but have also been verified in anatomical specimens (Tubbs et al 2006).

The cervical ribs may be a cause of nerve compression, with secondary numbness and pain in the arms and hands; the resultant thoracic outlet syndrome may actually lead to the diagnosis of Klippel-Feil syndrome and be relieved surgically (Konstantinou et al 2004). Pneumatocyst has been observed in the cervical rib of a patient (Hactanir et al 2005). A variety of upper limb defects or deficiencies involving the humerus, ulna, or radius are recognized (Thomsen et al 2000). Radial anomalies may manifest as Fanconi anemia with radial aplasia (Mc Gaughan 2003) or involve the first metacarpal and thumb (Narang and Goyal 2006).

Kyphoscoliosis with deformity of the thoracic cage may be a cause of pulmonary hypertension and respiratory failure (Suga et al). Lumbar fusion has been reported in one patient (Mahirogullari et al 2006). Patellar hypoplasia has been observed (Rahimi- Movagharet al 2004).

Hearing deficits occur in 20% to 50% of patients and may be conductive, sensorineural, or mixed (Scott 1993). Affected individuals may have microtia or malformations of ossicles or temporal bones.

Eye anomalies such as strabismus, nystagmus or chorioretinal atrophy occur in 20% of patients.

Neurologic impairments manifest in approximately one-half of patients and include synkinesia, facial nerve palsy, spasticity, hemiplegia, paraplegia, triplegia, or quadriplegia (Mosberg 1953; Tubbs et al 2003). Quadriplegia may be transient in affected patients and mimic cervical cord neurapraxia (Gupta et al 2007). Morphologic abnormalities include frontonasal malformation, hydrocephalus, chiari malformation, meningocele, encephalocele, syringomyelia, diastematomyelia, intramedullary inflammatory mass, and posterior fossa dermoid cyst (Helmi and Pruzansky 1980;

OMIM 1997; Aksoy et al 2001; Andronikou and Fieggeru 2001; Hinojosa et al 2001; Dickerman et al 2004). It has been suggested that squamous cell carcinoma may develop secondary to dedifferentiation of a dermoid cyst (Oertel et al 2002). Intelligence is normal in about 90% of patients.

One case of Rothmund-Thomson syndrome (poikiloderma congenitale) and (remote) osteosarcoma has been reported (Gelaw et al 2004), as has been Poland anomaly in a 7-year-old girl (Erol et al 2004). Cleft palate or submucous cleft palate is recognized in about 17% of affected patients. An osseous choristoma of oral mucosa has been identified in one adult woman (Gaitan –Cepeda et al 2003). Additional dental problems include oligodontia of both primary and permanent teeth, maxillary constriction, and velopharyngeal insufficiency (Barbosa et al 2005). Laryngeal cartilages may be malformed, causing aphonia or other voice impairment (Clarke et al 1994).

Congenital heart disease (usually ventricular septal defect) may occur in up to 10% of patients. Aortic arch anomalies have been recognized, including coarctation in one adult (Franzen et al 2003) and absence of one internal carotid artery in another, the latter associated with headaches, paresthesia, and vertigo (Abbas et al). Aneurysm of a noncoronary sinus of Valsalva has also been reported (Kawano et al 2006). Embryological vascular disorder like subclavian artery supply disruption (SASD) sequence have been hypothesized to result in Klippel Feil syndrome (5). A case of thoracic bifurcation of carotid artery suggestive of SASD is observed (10).

Genital anomalies may include cryptorchidism, hypospadias, or absence of vagina, uterus, or Fallopian tubes. Unilateral renal agenesis is the most common renal anomaly (GOLDENBERG 1987; Mahirogullari et al 2006).

Rarely sleep disordered breathing like fatal obstructive sleep apnea or stridor or bradypnoea may be seen and all children diagnosed with Klippel Feil syndrome should be regularly followed up for these (9).

TREATMENT

If asymptomatic and no evidence of instability:

- periodic flexion extension radiographs
- avoidance of contact sports
- Avoidance of occupational and recreational activities which are associated with the risk of head

trauma.

Surgical intervention is considered for:

- progressive symptomatic instability
- Neurological compromise.

Persons with Klippel-Feil syndrome and cervical stenosis may be at increased risk for spinal cord injury after minor trauma as a result of hyper mobility of the various cervical segments (8). A 51 year old male is reported of quadriplegia following minor fall (1) which emphasizes the need to create awareness of the problem among patients suffering from this syndrome.

FEW DOCUMENTED OPERATED CASES OF KLIPPEL-FEIL SYNDROME

Journal of Chugoku- Shigoku orthopaedic Association reports a 8 yr girl with 2 yr history of tetraparesis who underwent transoral odontoidectomy. She had atlanto axial dislocation and basilar impression. The atlantoaxial dislocation could not be reduced, hence decompression of foramen magnum and C1 laminectomy and occipitoaxial fusion was carried out. Consequently tetraparesis improved but eleven months after the procedure patient gradually worsened. The reason for it was attributed to progression of anterior compression of the medulla oblongata by the odontoid. After transoral odontoidectomy the patient gradually improved (2). Craniocervical decompression is reported in 31 yrs, male who had developed tetraparesis following minor trauma (5).

A 36-year-old man presented with posterior neck pain and myelopathic symptoms was reported for cervical arthroplasty. His radiograph demonstrated congenital fusion of the vertebral bodies at C2–3, C4–5 and C5–6. On MRI, the spinal cord was compressed by a protruding cervical disc and bony spurs at C6–7. After anterior discectomy and decompression of the spinal cord at the C6–7 level, the disc was replaced with the Bryan cervical disc system to restore normal motion. The absence of adjacent segment degeneration and the preservation of cervical motion were noted 2 years after surgery. Arthroplasty may be performed in selected patients with Klippel-Feil syndrome in order to restore motion and to prevent degeneration of the adjacent segment by reducing hypermobility (3).

A case with Klippel-Feil (KFS) syndrome associated with hypertelorism, microtia, Sprengel deformity, hand and feet

deformities, scoliosis, clinical manifestations of Klippel-Trenaunay syndrome and severe arterial hypertension secondary to renal artery stenosis has been reported. This patient underwent for surgical revascularization unsuccessfully, they hypothesized that for patients with KFS and unilateral renal artery stenosis medical treatment with ACE inhibitors can provide more benefits than surgery or percutaneous transluminal angioplasty(4).

CONCLUSION

- for better results in the management, all underlying problems should be identified before sending patient for surgery.
- Anesthesiologist's should be careful for manipulation of the neck and head during induction of anesthesia.
- The final outcome will be related to underlying cardio-respiratory, renal, and nervous system problem.
- For operations involving decompression of cervical segments, postoperative ventilatory support and ICU care should be ensured.
- Operations at higher cervical spine carry bad prognosis.
- The decision to treat such cases conservatively or

surgically should also involve consideration of prolonged ICU stay and complications associated with it, the cost involved in it.

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