

Cleidocranial Dysplasia: A Case Report Under Spinal and General Anesthesia in Adult Female

Tanveer Baig*

Consultant Anesthetist, Aga Khan University Hospital, Karachi, Pakistan

Abstract

Cleidocranial dysplasia is a very rare autosomal dominant disorder, characterized by defective ossification of membranous bones. Difficulties in neuraxial placement and airway management are a major concern from anesthesia point of view.

Keywords: Cleidocranial dysplasia; CCD

Introduction

Cleidocranial dysplasia (CCD) is characterized by defective ossification of the membranous bones primarily the clavicles, cranium and pelvis. Syndrome was first described in 1765 by Martin [1] in a patient with absent clavicle. Major characteristics of CCD are delayed closure (ossification) of the fontanels, premature closing of the coronal suture, a protruding mandible, mid-face retrusion, abnormal dentition including delayed eruption of secondary dentition, failure of primary teeth eruption, supernumerary teeth with dental crowding, and clavicular hypoplasia [2]. Potential difficulties for airway management and neuraxial placement were described in the general anesthetic management of a patient with CCD who underwent a variety of obstetrical procedures [3].

Case Presentation

A 30 year-old female with CCD presented to our operating room for emergency cesarean section due to pre-eclampsia and cephalopelvic disproportion. Past medical history revealed repeated chest infections. During pre-operative evaluation patient had a limited mouth opening (Mallampati Class III), brachycephaly, frontal bossing, and short stature. No prior ultrasound was done.

So spinal anesthesia was planned, routine monitoring and pre-load with ringer solution started, Patient explained about procedure, back was cleaned with Iodine solution, spinous process palpated but spaces were uneven may be due to hemivertebrae after two efforts we succeeded, 26G spinal needle was used, 0.5% heavy bupivacaine 10 mg loaded in subarachnoid space after free flow of cerebrospinal fluid seen. Surgery finished uneventfully and the patient was shifted to Obstetrics ward but after three days patient developed sign of intestinal obstruction. A difficult intubation trolley was prepared and induction was done with 2 mg/kg propofol, rocuronium 1.2 mg/kg and fentanyl 1 microg/kg. It was not difficult to ventilate, but on laryngoscopy with external neck manipulation only posterior commissure was seen so fibro-optic flexible laryngoscope was used and 6.5 mm size endotracheal tube was passed and fixed after chest auscultation. Anesthesia was maintained at 30% oxygen, 70% nitrous oxide and isoflurane 1.2%. Postoperative analgesia was provided by intravenous paracetamol 15 mg/kg and ketoprofen 1 mg/kg. Duration of surgery was 60 minutes and emergence from anaesthesia was smooth. Patient was shifted to Intensive care unit for monitoring and observation. Patient was then shifted to surgical ward after two days.

Discussion

Cleidocranial dysplasia is characterized by delayed fontanel

closure, hypoplastic or aplastic clavicles, various dental abnormalities and shoulders are brought forward into close proximity to the chest due to partial or complete absence of clavicles [4].

The major findings in this case were absence of right clavicle, overcrowded thoracic vertebra and hemi vertebra as shown in Chest X-ray (Figure 1). The X-ray pelvis PA view showed big gap between pubis bone, small pelvis and gap between sacroiliac joint as shown in (Figure 2). She also had frontal bossing and slopping of shoulders. Due to hyper mobility of shoulder and absence of clavicle she was able to bring both shoulders in front of chest.

Pre-operative assessment of cardiac and respiratory function is necessary. Facial skeletal abnormality may lead to difficult intubation on one hand, vertebral crowding and hemi vertebra causes problem in spinal and epidural insertion on the other hand. Spinal and vertebral abnormalities should be evaluated before considering neuraxial blockade [5]. Positioning under anesthesia should be carefully done because bones are fragile and joint are hyper mobile. Fiberoptic intubation was not done as this facility was not available at that and its one of the limitation in this case report.



Figure 1: Chest X-ray of 30 years old female patient with Cleidocranial Dysplasia showing hemivertebra and absent right clavicle.

***Corresponding author:** Tanveer Baig, Consultant Anesthetist, Aga Khan University Hospital, E-194, Sector 31-E, Luchknow Housing Society, Korangi, Karachi, Pakistan, Tel: 00923243372395; E-mail: tanveerbaig65@yahoo.com

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Figure 2: X-ray Pelvis showing gap between two pubic bone.

Conclusion

Cleidocranial dysplasia is a rare syndrome involving facial bone and spine so should be evaluated carefully pre-operatively to reduce morbidity and mortality.

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