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ANAESTHETIC MANAGEMENT OF A PATIENT WITH CLEIDOCRANIAL DYSPLASIA: A CHALLENGE FOR ANAESTHETIST

Anaesthesiology

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ABSTRACT

Introduction: Cleidocranial Dysplasia, a rare autosomal dominant genetic syndrome, possess abnormal anatomical features of the head, mouth, neck and spinal column. These features may result in perioperative problems such as difficulty in airway management and complicated regional anaesthesia. **Case Report:** We report the anaesthetic management of a young male with cleidocranial dysplasia undergoing open reduction and internal fixation for comminuted fracture of distal humerus. **Conclusion:** Difficult airway management presents a major concern and early initiation of neuraxial analgesia is recommended.

KEYWORDS

cleidocranial dysplasia, brachial plexus block, fracture distal humerus

INTRODUCTION:

Cleidocranial dysplasia is a rare congenital defect of autosomal dominant inheritance, primarily affecting bones that undergo intramembranous ossification, i.e. generally the calvarian but also the clavicular bones. It is also known as Marie and Sainton disease, Mutational dysostosis and Cleidocranial dysostosis. It was first described by Pierre Marie and Paul Sainton in 1898 [1].

Case Report:

A 30 year old, 150 cm, 60 kg male presented to orthopaedic OPD with history of fall while playing kabbadi. He presented with pain and swelling in and around the elbow and a diagnosis of fracture distal end of humerus was made. He was planned for open reduction and internal fixation. During pre-anaesthetic checkup, patient was short statured, had frontal bossing, large nasal bridge, high arched palate, dental malalignment and hypoplastic clavicles. A diagnosis of Cleidocranial Dysplasia was made. There were no respiratory, cardiac or renal problems. Laboratory investigations and biochemistry values were within normal limits. His Mallampati grade was IV. There was no family history of similar condition. As this was a case of difficult airway, surgery was planned under regional anaesthesia. But due to non availability of ultrasound, in our setup we planned peripheral nerve stimulator guided supraclavivular brachial plexus block.

Figure 1: Patient having frontal bossing

A difficult airway cart was kept ready. After performing PAC, patient was shifted to the operative area, ASA monitoring- NIBP monitor, ECG and pulse oximeter were attached to the opposite arm. Patient was not given any premedication. The patient was placed in the supine position with the head turned to the opposite side of the operating arm and arm to be operated was supported on abdomen. Due to hypoplastic clavicle, interscalene groove was not palpable, but subclavian artery pulsation was palpated. The thumb of the non-dominant hand was placed on the arterial pulsation. The skin wheal was raised with 2% lignocaine. A 22-G, 50 mm short- beveled needle (Stimuplex, B.Braun) was inserted in the parasagittal plane parallel to the midline and posterolaterally to the tip of the thumb placed on the pulsation of subclavian artery. After achieving the target flexion response of the fingers at 0.4 mA, a total of 35 ml of 0.75% local anaesthetic (0.75%Ropivacaine and 8 mg Dexamethasone) was instilled slowly for single shot block. Level of sensory and motor block was assessed every 2 minutes after drug administration. Desired level of anaesthesia was achieved. Surgery lasted for 2 hours and was performed uneventfully. Care was taken during positioning to avoid excessive range of movements at all joints.

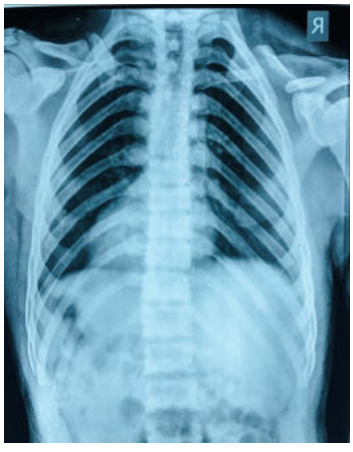

Figure 2 : Chest-X ray showing hypoplastic clavicle

DISCUSSION:

Cleidocranial dysplasia is an inherited disease caused by mutation in RUNX2 gene on chromosome 6p21. This mutation impairs the intramembranous ossification process, most notably at the skull and clavicles [2].The causative gene encodes runt-related transcription factor 2 [3]. CCD presents with skeletal defects of several bones, the most striking of which are partial or complete absence of clavicles, late closure of the fontanells, presence of open skull sutures and multiple Wormian bones. The skull base is dysplastic and reduced in growth resulting in increased skull width leading to brachycephaly and hypertelorism. Delayed closure of anterior fontanel and metopic sutures result in frontal bossing [4]. Thoracic cage is small and bell shaped with short ribs. Typically, clavicles are underdeveloped to varying degrees and in approximately 10 percent of cases, are completely absent. Other bones may also be affected including long bones, vertebral column, pelvis and bones of hands and feet [5]. Diagnosis is usually made shortly after birth, but in our case it was delayed. Facial dysmorphisms, skeletal anomalies and recurrent respiratory tract infections in a background of near-normal general

health, cognition and intellect present unique challenges for the anaesthesiologist. The clinical spectrum of this orphan disease can vary from mild dental and skeletal abnormalities to rare, serious afflictions such as syringomyelia [6]. A variety of dental problems may occur in CCD, in particular, supernumerary teeth (hyperdontia) in the primary and secondary dentition may lead to dental crowding and malocclusion [7]. Facial abnormalities may lead to difficult mask ventilation and intubation.The facial dysmorphism, micrognathia, abnormal palatal shape and fragile teeth together may produce a 'cannot-ventilate, cannot-intubate' like situation and difficult airway cart should be ready [6].These various abnormalities can potentially pose challenges to anesthetic management; however, there is limited literature describing anesthetic implications of CD patients undergoing surgery. The potential anaesthetic considerations are due to anatomical abnormalities [1].The narrow thorax may cause restrictive lung disease and lead to respiratory failure and development of right heart failure [8].Preoperative assessment of cardiac and respiratory function is necessary. Hypoplastic or aplastic clavicles, cone shaped thorax, hypoplastic scapulae may lead to respiratory insufficiency, especially in the immediate post operative period. Positioning under anaesthesia should be done carefully as bones are very fragile and joints are hypermobile [9]. Due to non availability of ultrasound, we planned single shot peripheral nerve stimulator guided block and surgery was performed uneventfully.

CONCLUSION:

Patients with cleidocranial dysplasia have many potential problems related to anatomical abnormalities and very little has been published about anaesthetic management in patients with CCD. Difficult airway management presents a major concern and early initiation of neuraxial analgesia is recommended.

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