CASE REPORT

Anaesthetic Management of Upper Limb Fracture in a Patient With Cleidocranial Dysplasia

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ABSTRACT

Background: Cleidocranial dysplasia (CCD) is a rare autosomal dominant skeletal disorder which is characterised by defective skeletal ossification, mainly affecting the clavicles and skull, along with dental abnormalities.

Case Presentation: This study reports the anaesthetic management of a 12 year old boy with cleodocranial dysplasia with traumatic fracture of both bone right forearm without any neurovascular deficits. Ultrasound guided Supraclavicular brachial plexus block was administered under intravenous sedation. Vitals were monitored throughout the case which was stable and surgery was completed.

Conclusion: Considering the ease, safety and potency of ultrasound guided supraclavicular block, it may be used for patients with CCD with absent clavicle. Peripheral nerve block under intravenous sedation avoids airway manipulation in patients with CCD characterized by generalised skeletal and orofacial defects.

Keywords: Cleodocranial dysplasia, Orphan disease, absent clavicle, bothbone fracture, upperlimb fracture, ultrasound guided supraclavicular brachial plexus block, dexmedetomidine, ketamine

Introduction

Cleidocranial dysplasia (CCD) is a rare autosomal dominant skeletal disorder which is characterised by defective skeletal ossification, mainly affecting the clavicles and skull, along with dental abnormalities.

Cleidocranial dysplasia (CCD) is caused by mutations in the human osteoblast-specific runt-related transcription factor 2 (RUNX2) gene, also known as CBFA1 located on chromosome 6p21, which plays a critical role in osteoblast differentiation and bone formation.

CCD is characterized by delayed closure of the fontanelles and persistence of open skull sutures, hypoplastic or aplastic clavicles, supernumerary teeth, delayed eruption of permanent dentition, short stature, a wide pubic symphysis, and various other skeletal abnormalities. Delayed closure of the

anterior fontanelle and metopic sutures leads to frontal bossing.

A hallmark feature is clavicular hypoplasia or aplasia, which enables affected individuals to approximate their shoulders in the midline.The phenotypic spectrum is broad, ranging from mildly affected individuals with only dental anomalies to severely affected patients with complications such as syringomyelia. CCD is also referred to as Marie–Sainton disease, mutational dysostosis, and cleidocranial dysostosis.

Case description

12 year old boy of 20 kg, lower socioeconomic status, presented with history of fall while playing and sustained fracture of both bone right forearm.
The patient had brachycephaly, frontal bossing, hypertelorism, barrel shaped chest, open fontanelles,a low nasal bridge, reduced nasal length with increased nasal width, supernumerary teeth and low height for age. Mouth opening was adequate, no intellectual disability, co-operative. Clinically clavicles were not palpable and radiologically absent clavicles were observed. Other systems were within normal limits.

Anaesthetic Management

A detailed history of the child including past history, present complaints, and family history was obtained. Rapport was established with the child before accompanying him to the operation theatre. The risks, benefits, and available alternatives to the procedure were explained in detail to the guardians, and written informed consent was obtained.

In the operating theatre, intravenous access was confirmed to be patent, and Ringer's solution was started for maintenance. Supplemental oxygen at 5 L/min via face mask was administered, followed by premedication with 0.2mg glycopyrrolate and, sedation with 0.5 mg midazolam and 10 mg Ketamine. Dexmedetomidine infusion was started at 14mcg/hour and titrated according to haemodynamics.

The patient was positioned supine with the ipsilateral arm adducted and the head slightly turned to the opposite side. The skin over the neck was disinfected and prepared using povidone-iodine solution. Ultrasound guidance was used to identify the brachial plexus. The transducer was first placed in the transverse plane to identify the carotid artery and internal jugular vein. After identifying the artery, the probe was moved slightly laterally across the neck to visualize the anterior and middle scalene muscles and the brachial plexus located between them at the interscalene level. The transducer was then traced caudally until the brachial plexus appeared as a honeycomb-like cluster posterior and superficial to the subclavian artery. To obtain the "podium view," the ultrasound probe was angled caudad toward the thoracic cavity, allowing visualization of the brachial plexus near the subclavian artery. The visceral and parietal pleurae interface was identified as "lung sliding" on ultrasound. The first rib was also visualized as a hyperechoic line with posterior acoustic shadowing, with the plexus and subclavian artery seen overlying the rib. This positioning allowed the first rib to act as a bony backstop, thereby reducing the risk of pneumothorax. Slight posterior rotation of the lateral edge of the probe helped optimize this view. Colour Doppler was used prior to needle insertion to exclude the presence of major vascular structures along the needle path. A mixture of 3 mL of 2% lidocaine hydrochloride and 7 mL of 0.5% bupivacaine hydrochloride was injected after frequent negative aspiration. During injection, the spread of the local anesthetic around the plexus was visualized on ultrasound as a hypoechoic area. Throughout the procedure, heart rate, blood pressure, oxygen saturation, and end-tidal CO were continuously monitored.

Discussion

Supraclavicular brachial plexus block carries potentially serious complications such as pneumothorax and vascular puncture. The risk of pneumothorax is relatively higher in children due to the close proximity of the cervical pleura. The use of real-time ultrasound guidance significantly reduces these risks by allowing visualization of surrounding structures and enabling precise deposition of local anesthetic around the brachial plexus, and help to lower the local anesthetic volume needed to perform the blocks.

In our case, the block was performed in a child with absent clavicle, which altered the usual anatomical landmarks. Ultrasound guidance was therefore particularly valuable for identifying the brachial plexus and adjacent structures safely. The block was successfully performed with adequate spread of local anesthetic, resulting in effective anesthesia and satisfactory postoperative analgesia for the upper extremity surgical procedure.

Data Availability Statement

The original contributions presented in the study are included in the article/supplementary material, further inquiries can be directed to the corresponding authors.

Funding

Nil

Conflict of Interest

There are no conflicts of interest.

Discussion

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