Case Report

Anesthetic Management of a Case of Cleidocranial Dysplasia

Arun Sundaram, Shibinath Veluthamanil, Abisha Sahaya Mercyline, Aparna Gilbert, Godwin Jino, Sudha Kumara Dhas

Department of Anaesthesiology, Dr. Somervell Memorial CSI Medical College, Thiruvananthapuram, Kerala, India

Abstract

Hereby, we present the case of an adult with cleidocranial dysplasia who underwent multiple tooth extraction. In this article, characteristics of this genetic disorder and implications for an anesthesiologist are being discussed.

Keywords: Anesthesia, cleidocranial dysplasia, Marie-Sainton syndrome

Introduction

Cleidocranial dysplasia (CD) was first described in 1765. The responsible gene has been mapped to 6p21. The clinical spectrum of this disease can vary from mild dental and skeletal abnormalities to rare, more serious conditions such as syringomyelia.[1] Facial dysmorphisms, skeletal anomalies, and recurrent respiratory tract infections[2] present unique challenges for the anesthesiologist. It can be inherited as an autosomal dominant characteristic with high penetrance and variable expressivity.[3]

The associated skeletal anomalies and recurrent respiratory tract infections present unique challenges for an anesthesiologist. Bony abnormalities such as brachycephalic skull with mid-face retrusion, abnormal dentition, micrognathia, abnormal palatal shape, and fragile teeth together may produce a "cannot-ventilate, cannot-intubate" like situation. In this article, we are presenting our approach to airway assessment and anesthetic management of a patient with CD undergoing multiple tooth extraction under general anesthesia (GA).

Case Report

A 19-year-old male, who was diagnosed with CD at the age of ten, was posted for multiple teeth extraction under GA. His past medical history was unremarkable, and family history was insignificant for any genetic disorder.

Physical examination of the patient revealed short stature with brachycephaly, frontal bossing, large nasal bridge, hypertelorism, micrognathia, and presence of supernumerary teeth [Figure 1]. He also had brachymetatarsia bilaterally with broad thumbs and toes [Figure 2]. Further examination revealed drooping of shoulders, and he was able to approximate the two acromial regions below the chin [Figure 3]. His cardiovascular and respiratory system evaluation showed no abnormalities.

Patient's mental status was appropriate for age and muscle tone was found to be normal on neurological examination. Airway assessment showed adequate mouth opening, supernumerary and fragile teeth, and Mallampati Class-III, and he had adequate range of neck movements.

Radiographically, there was bilateral hypoplasia of clavicles [Figure 4] and impacted supernumeraries teeth [Figure 5] and hypoplasia of maxilla which confirmed the clinical diagnosis of CD. Orthopantomogram showed supernumerary deciduous and permanent teeth.

Anesthetic concerns of cleidocranial dysplasia

As spinal abnormalities may pose challenges, spinal or vertebral anomalies should be ruled out before considering neuraxial blockade. Preoperative evaluation for difficult airway, restrictive lung disease, and kyphoscoliosis should be done.

Anatomic abnormalities of the skull and facial structures (including dentition, such as delayed eruption of secondary

Address for correspondence: Dr. Arun Sundaram, Asst Prof., Somervell Memorial CSI Medical College, Parassala, Vellarada Road, Karakonam, Thiruvananthapuram - 695 504, Kerala, India.
E-mail: arunsam85@yahoo.com

Access this article online

Quick Response Code:

Website: www.karnatakaanaesthj.org

DOI: 10.4103/kaj.kaj_10_18

How to cite this article: Sundaram A, Veluthamanil S, Mercyline AS, Gilbert A, Jino G, Dhas SK. Anesthetic management of a case of cleidocranial dysplasia. Karnataka Anaesth J 2017;3:82-4.

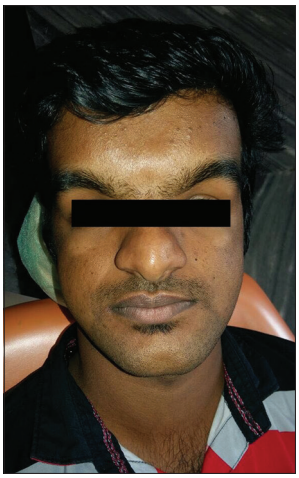

Figure 1: Hypertelorism

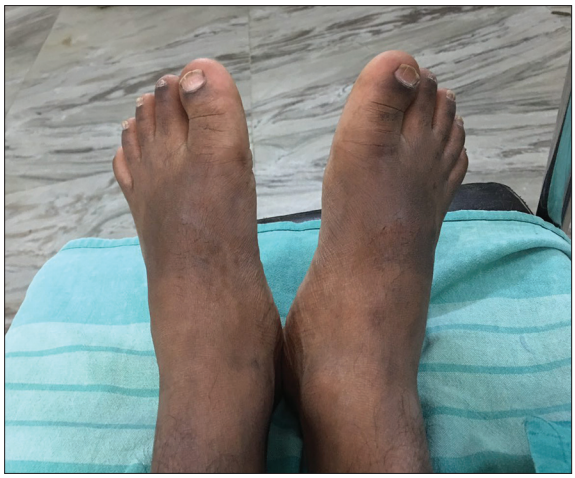

Figure 2: Brachymetatarsia

Figure 3: Anterior approximation of both acromion processes

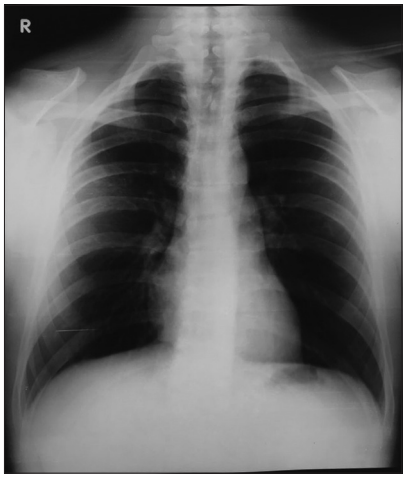

Figure 4: Hypoplasia of clavicles

dentition, failure of primary teeth eruption, and supernumerary teeth with dental crowding) may impede with mask ventilation and endotracheal intubation. Hence, difficult intubation cart should be kept ready.

As the patients usually have joint laxity and tendency for joint dislocations, they should be carefully positioned. The use of ultrasound is for brachial plexus blocks and central venous line insertions were advised.

Before the surgery, the patient was kept nil per oral for 6 h; an 18-gauge intravenous (IV) cannula was positioned in the dorsum of left hand under local anesthesia. Both nostrils were prepared with oxymetazoline nasal drops (0.05%) to facilitate nasal intubation and minimize nasal bleeding. Routine American Society of Anesthesiologists monitors were placed, and the patient was premedicated with IV midazolam. He was denitrogenated followed by IV induction with propofol (2 mg/kg) and fentanyl (2 mics/kg).

After confirming mask ventilation vecuronium 0.1 mg/kg was given, with a Macintosh 3 blade and some external laryngeal manipulation, 6.5 sized (difficulty in negotiating >7 sized Ring Adair Elwin (RAE) tube through both nostrils) cuffed nasal RAE tube was passed through the left nostril and was negotiated into the patient's trachea under direct vision with the aid of Magill forceps. Throat pack was kept. Anesthesia was maintained with 50% oxygen, 50% nitrous oxide, 1%–2% sevoflurane, and vecuronium and fentanyl 20 µg IV bolus hourly. Toward the end of the procedure, inhalational agent and vecuronium were tapered and stopped. Prophylaxis for postoperative nausea and vomiting with ondansetron 0.1 mg/kg and dexamethasone 0.1 mg/kg was given. After nasal and oral suction, throat pack was removed. He was reversed with neostigmine and glycopyrrolate and was extubated after he was fully awake. Postoperative analgesia was provided with IV paracetamol 15 mg/kg 6th hourly. The postoperative course was uneventful, and the patient was discharged after 24 h.

Discussion

CD is a general skeletal dysplasia. Mutation in the gene on 6p21 encoding transcription factor, core-binding factor subunit alpha-1, or Runt-related transcription factor 2 is responsible for CD.[4]

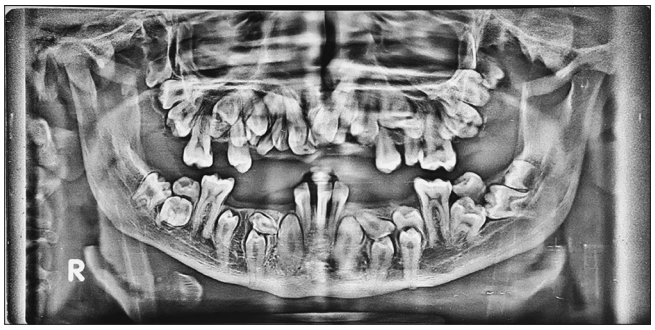

Figure 5: Supernumerary teeth

Patients with CD usually have short stature and marked drooping of shoulders. CD primarily affects the skull, clavicles, and dentition. The face appears small as a result of hypoplasia of the maxilla, they may have brachycephalic skull, with frontal and parietal bossing. Hypertelorism and depressed nasal bridges are present.[5,6]

Hypermobility of the shoulders may be present due to complete or partial absence of clavicular calcification, with associated muscle defects. Delayed closure of symphysis pubis and a wide symphyseal space will give a "chef's hat" appearance to the pelvis. Hemivertebrae and posterior wedging of the thoracic vertebrae may contribute to the development of kyphoscoliosis and pulmonary complications.[7,8]

Mandible appears to be relatively prognathic, due to maxillary hypoplasia. The palate is narrow and highly arched, and there is an increased incidence of submucosal clefts and complete or partial clefts of the palate. They have supernumerary teeth due to incomplete or severely delayed resorption of the dental lamina.

Features similar to CD are also seen in various other disorders like congenital pseudarthrosis of clavicle, which is usually unilateral; pyknocytosis, which does not have supernumerary teeth and Yunus Varon syndrome which is associated with low intelligence and failure to thrive.

Thereby, careful preoperative assessment and preparation with appropriate equipment will help in successful management of CD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Acknowledgment

First of all, I would like to thank God, the Almighty, for having made everything possible by giving me strength and courage to do this work. I take this opportunity to express my gratitude and deep regards to my head of the department, professors, and associate professors for their exempting guidance, concern, and support for this case report. I am obliged to my junior residents for the valuable information provided by them in their respective fields. I wish to express a sincere thanks to my patient who graciously agreed to participate in my study. I would like to thank my colleagues for their wonderful collaboration and support. I am grateful for the love and encouragement of my wife Aparna and daughter Ananaya who has made all the difference in my life. A special word of thanks also goes to my family for their continuous support and encouragement.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Datta R, Agarwal J, Sharma D. Anaesthetic considerations in an orphandisease with skeletal anomalies. Indian J Anaesth 2016;60:780-2.
2. Cooper SC, Flaitz CM, Johnston DA, Lee B, Hecht JT. A natural historyof cleidocranial dysplasia. Am J Med Genet 2001;104:1-6.
3. Fitchet SM. Cleidocranial dysostosis: Hereditary and familial. J Bone Joint Surg 1929;11:833-66.
4. Patil PP, Barpande SR, Bhavthankar JD, Humbe JG. Cleidocranial dysplasia: A Clinico-radiographic spectrum with differential diagnosis. J Orthop Case Rep 2015;5:21-4.
5. Mundlos S. Defects of human skeletogenesis – Models and mechanisms. Novartis Found Symp 2001;232:81-91.
6. Schuch P, Fleischer-Peters A. To the clinic of dysostosis cleidocranialis. Kinderheilkd 1967;98:107-32.
7. Jarvis JL, Keats TE. Cleidocranial dysostosis. A review of 40 new cases. Am J Roentgenol Radium Ther Nucl Med 1974;121:5-16.
8. Cole WR, Levin S. Cleidocranial dysostosis. Br J Radiol 1951;24:549-55.