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SURGICAL EXPOSURE OF TEETH IN CLEIDOCRANICAL DYSPLASIA UNDER PROCEDURAL SEDATION:- CASE REPORT

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ABSTRACT

Cleidocranial Dysplasia (CCD) is a rare autosomal dominant condition which is mainly characterised by skeletal abnormalties like hypoplastic or aplastic clavicles, increase in transverse diameter of the cranium, delayed closure of fontanelles with presence of open sutures and skeletal changes in maxilla and mandible. 2,4 These patients usually presents with delayed exfoliation of deciduous teeth, delayed or failed eruption of the permanent dentition with multiple supernumerary teeth, protruding mandible and mid- face retrusion.2 We present a case report of a 16 year old girl undergoing multiple surgical exposure of impacted permanent teeth under conscious sedation so as to move them in normal occlusion with orthodontic traction.

INTRODUCTION:-

Cleidocranial Dysplasia (CCD), also known as Scheuthauer-Marie-Sainton syndrome, Marie-Sainton Disease, is a rare disorder which is an autosomal dominant inherited condition caused by mutation in RUNX2, product of this gene is associated with osteoblastic differentiation and chondrocyte mutation during the process of endochondral ossification. This disorder is mainly characterised by atleast 1 or both aplastic or hypoplastic clavicles, delayed skull ossification, with severe dental abnormalities. [1,2,4] Since, most of the craniofacial abnormalities become obvious only during adolescence, diagnosis is quite difficult in early stage of the disorder. The effects of health by this disorder are not severe and don't cause any cognitive or intellectual improvement. Patients present with brachycephalic skull appearance and delayed closures of sutures, fontanelles as compared to normal.

These patients tends to be short statured with narrow chest and drooping of shoulders.[1] The prevelance of Cleidocranial Dysplasia is one in a million, with equal frequency among males and females.[2]

Occlusal abnormalities are mainly caused by prolonged exfoliation of primary dentition, delayed eruption of permanent teeth and multiple impacted supernumerary and permanent teeth.[1,2,3,4]

Extraction of impacted supernumerary teeth and retained primary dentition is usually carried out as primary step in oral surgery.[4] This is usually followed by exposure of permanent dentition by thinning of cortical bone.[4]

The present case was a typical presentation of cleidocranial dysplasia with characteristic features. This report describes the features of this rare disorder and explains the surgical procedure done to expose the unerupted teeth under procedural sedation which helps in reducing the patient's anxiety as well as increasing patient's and surgeon's comfort.

Case Report:-

A 16 year old female presented with the chief complaint of multiple missing teeth in upper jaw. Orthopantomogram was obtained for examination of multiple retained deciduous and retained permanent teeth.

Total retained teeth were five in number i.e 72, 73, 74, 75, 85 and impacted were fifteen in number i.e. 15, 13, 12, 11, 22, 23, 24, 25, 32, 33, 34, 35, 43, 44, 45.

4 teeth were missing which included 17, 27, 11, 47.

After orthodontic consultation, surgical exposure of permanent teeth was decided so as to move them in normal occlusion with orthodontic movements.

Extraction of 72, 73, 74, 75, 85 followed by 32, 33, 34, 35, 43, 44, 45 teeth were planned for surgical exposure under local anesthesia with procedural sedation followed by orthodontic buttons placement for orthodontic traction.

Apart from the bilateral hypoplastic clavicle, short stature, brachycephalic skull and supernumerary teeth and dental abnormalities, the patient's Past medical history was unremarkable. History from mother revealed normal development milestones, normal intelligence and no history of decreased exercise tolerance. Family history for any genetic disorder from maternal and paternal side was also negative. Assessment of respiratory function by auscultation and chest x-ray was normal, saturation was 98% in room air. Clinical evaluation of cardiac function by ECG and auscultation showed no abnormalities. Patient had normal muscular tone and mental status appropriate for age. Full blood count and coagulation profile was within normal

limits.Airway examination revealed normal mouth opening (two and half finger breadths), multiple missing teeth, mallampati grade II airway evaluation, normal thyromental distance and normal neck movements.

Patient met the pre-operative fasting requirement (>6 hours) and was medically cleared and all the pre-operative tests were withing normal limits. Monitors included a pulse oximeter, 5-lead electrocardiograph, noninvasive blood pressure (BP) and heart rate were attached. Her initial vital signs were as follows: BP 110/74 mm Hg, heart rate (HR) 87 bpm, and peripheral capillary oxygen saturation (SpO2) 100%. 22 gauge i.v. cannula was placed on the dorsum of the left hand. Patient appeared anxious but was reassured. Patient was given 0.5 mg I.V. Midazolam + (0.5 ug/hr) 20 ug Fentanyl until the adequate sedation was achieved. (RSS 2-3)

Once RSS>2 was achieved, Local anesthetic was given by aspirating by the surgeon (2 % lidocaine with 1:200000 adrenaline) by negative aspirating technique. After the onset of anesthesia, crevicular incision was given followed by flap raise and extraction of the deciduous teeth i.e 72, 73, 74, 75, 85. Exposure was done by thinning of the cortical bone using surgical handpiece with thorough irrigation. Patient was provided with a mouth prop to help keep the mouth open during the operation.

After adequate exposure, orthodontic buttons were bonded to the teeth to allow proper traction for the tooth movement. Primary Closure was done with the help of interrupted suture using 3-0 Black braided silk suture. Procedure end time was approximately 65-70 min after the start of drug administration.

Hemodynamic and respiratory variables:- NIBP, Heart rate, ECG, oxygen saturation, and respiratory rate were recorded every 5 min. from the onset of procedure until discharge. Inj. Dexamethasone 4 mg iv, and Inj. Paracetamol 450 mg i.v was also given.

At the end of surgery, patient was kept in recovery area for 2 hrs. and non-invasive, BP, heart rate, and pulse oximetry were monitered. Post-operative antibiotic and analgesics prescription was provided.

Patient remained calm until discharge and was fully awake and ambulatory at the time of discharge accompanied by a family member. No complications were observed in immediate post-operative period. The patient came back after 24 hrs for follow up and complained of pain on the surgical site but stated that she was very satisfied with the sedation.

DISCUSSION:-

General dysplasia with various skeletal and dental deformities are characteristics of cleidocranial dysplasia. Skeletal feature is clavicular aplasia or hypoplasia with consequent hypermobility of the shoulders because of defective ossification of medial and lateral clavicular centers. Hypertelorism, recession of the nasal bridge, wide alar base, and maxillary hypoplasia with relative mandibular prognathism are the most common facial features [5,6]. Multiple supernumerary teeth with retention cysts that usually compromise the esthetics of dentition and function are significant dental anomalies.[4]

Patient age, craniofacial characteristics, dental anomalies, and social and economic circumstances decides the therapeutic plans for such patients.

Treatment is usually initiated during childhood; dental treatment options include generally surgical exposure of permanent retained teeth with orthodontic-guided eruption. Children usually exhibit behavioral problems and inability to cooperate especially during dental surgical procedures so they cannot be treated with locoregional anesthesia alone. Such cases must be done with the patient under conscious sedation.

In this case after extraction of retained deciduous tooth, surgical exposure of permanent tooth was done under conscious sedation with midazolam and fentanyl. These both provided best results in rapports of onset of action, depth of sedation, and anxio-analgesia.[7]

CONCLUSION:-

CCD individuals need extensive diagnostic work-up and a long-term treatment plan. It includes appropriate timing of removal of supernumerary and retained primary teeth, surgical exposure of impacted permanent teeth, orthodontic extrusion and alignment. For the surgical part patient's cooperation is must and mild sedation seems to provide promising results.

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