Scheuthauer-Marie-Ainton Syndrome-A Report of Rare Case and Multidisciplinary Management

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Abstract

Cleidocranial dysplasia is also known as Scheuthauer-Marie-Sainton Syndrome. It is a rare disease which can occur in either sex equally or by autosomal dominant trait. Problems associated with this condition are multiple supernumerary teeth, retained deciduous teeth, delayed eruption, change in shape, impacted teeth and absence of deciduous teeth resorption. By profession pediatric dentist is the person who identifies the retained deciduous teeth and delay in eruption or absence of permanent teeth. Here we report a rare case of cleidocranial dysplasia in a 25 year old female patient and interdisciplinary management.

Key Words: Supernumerary teeth, Retained teeth, Cleidocranial dysplasia, Dysostosis

Introduction

The Cleidocranial Dysplasia (CCD) is a rare entity which occurs in elderly individual in both the sexes and in any ethnic group [1]. They have persistent fontenelle of the cranium and late closure of the same. The sutures of the cranium remain opened. The parietal bones, frontal and occipital are prominent, the paranasal sinus are underdeveloped with skeletal dysplasia. The absence or hypoplastic clavicles is the characteristic feature with the individual and allow the movement of the shoulders up to the medial plan of the body without any discomfort. Cleidocranial dysplasia is also known as Marie and Sainton Disease, Scheuthauer Marie- and Mutational dysostosis [2].

Regarding the manifestations of odontological interest it is important to mention that these patients shows intra oral findings such as narrow and deep palate, supernumerary teeth, impacted and ectopic, crown and root anomalies, eruption is retarded and an absence of root resorption in the deciduous teeth, hypodontia and dentigerous cysts can also be observed.

Extra oral findings include underdeveloped maxilla and shorter than normal in relation to the mandible, resulting in a pseudo mandibular prognathism. The zygomatic and lacrimal bones are also underdeveloped. In the region of the simphysis of the mandible an incomplete fusion is common to be observed. We can also find the absence or less quantity of the cellular cementum in the roots of the permanent teeth and in some cases in both dentitions [3].

The management of cleidocranial dysplasia involves the restoration of the deciduous teeth when they are cavities, because their extraction does not necessarily induce the eruption of the permanent teeth. If maxillary hypoplasia, it involves the orthognathic surgery and the surgery of the impacted teeth along with the orthodontic and or prosthetic therapy. Here we report a rare case of cleidocranial dysplasia in a 25 year old female patient and interdisciplinary management.

Case Report

A female patient aged 25 years reported to private clinic with a chief complaint of delayed eruption of permanent teeth.Patient gave history of uterus cyst and was on medication for the same. Blood investigation was done to rule out abnormalities with thyroid, serum calcium and phosphatase level and found normal. There was no family history of hyperdontia or systemic disease. In the extra-oral physical examination we could notice short height, reduced inter acromial distance, prominent frontal and parietal bones, determining an increase in the cranial perimeter moderately nourished with a concave facial profile. When asked to move the shoulders, she was capable of bringing closer the humeral heads, which characterized the hyper-mobility of the shoulders. The intra oral examination revealed exfoliated lower primary incisors and erupting lower left permanent mandibular central incisor with all other primary teeth retained in both the arches, atresia of the maxilla and deep palate (*Figure 1*).

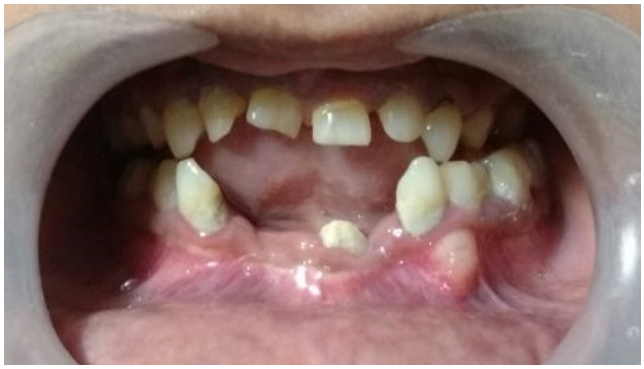

Figure 1. *Intra oral view of the patient with retained deciduous teeth in mouth.*

The panoramic radiographic revealed the existence of several teeth which were retained and impacted in maxilla and mandible. The presence of several impacted permanent teeth,

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along with multiple supernumerary teeth in the right and left canine and premolar areas and underdeveloped maxillary sinus were seen (*Figure 2*).

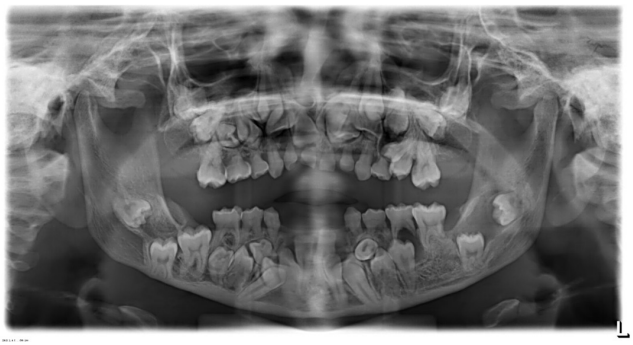

Figure 2. *Panoramic radiography showing the existence of several dental elements retained and impacted in maxilla and mandible.*

According to the clinical and radiographical findings, the diagnosis of cleidocranial dysplasia was made.

Discussion

Cleidocranial dysplasia was first described in 1765, whereas only in 1898 Marie & Sainton [4] had described the disease and associated patterns of inheritance. Later, Bauer and Kallialla [5] suggested the genetic mutation as an etiological factor of the disease. In 1962 Lasker and Forlan [6] had concluded that it was a genetic disease with an autosomal dominant inheritance.

In our case, the patient family members did not report the existence of any clinical characteristic of cleidocranial dysplasia. The radiographic findings showed presence of the deciduous teeth still in the arch, impacted permanent teeth with delayed eruption, although the patient was in her third decade of life. The failure in the eruption can be related with the absence or less amount of cellular cementum in the permanent teeth roots [7]. This was also claimed by Smith, who had observed the absence of cellular cementum in the deciduous and permanent dentition [8]. There are also other hypothesis that explains this fact, as the lack of absorption of the deciduous teeth and subjacent bone and also the presence of a physical barrier, such as supernumerary teeth impacted or by a fibrous connective tissue interposed between the dental follicles and the mucosa. Although several bones in the body showed dysplasia.That is why the Pedodontist role is so important in the diagnosis of this condition, as well as in the referral and implementation of a therapeutic multidisciplinary planning, aiming the improvement in the life quality of the patients with this condition.

Interdisciplinary management of cleidocranial dysplasia, involves several approaches and have been reported over the years. The option of no treatment with wait and watch was common in the past, followed by provision of dentures [9]. Some regard this approach as too invasive, especially considering the extensive bone loss experienced after removal of teeth in a patient already deficient in alveolar bone. Pusey and Durie suggested removal of only the erupted teeth and use of a removable prosthesis to minimize alveolar bone loss. However, subsequent eruption of retained teeth can require further surgery and modification of the prosthesis [10].

Early identification of the syndrome permits the planning of dental treatment by selecting the teeth that should be removed. Osny [11] suggests the use of a three-dimensional method of locating the position of impacted supernumerary teeth, insisting upon the importance of the removal of the supernumerary teeth and the planning of an orthodontic treatment that will allow for occlusion of the retained teeth. Ismet recommends a method where orthodontic forces may be applied to un-erupted permanent teeth moving them into a satisfactory, functional and aesthetic position [12]. Occasionally, when the teeth fail to erupt after the removal of supernumerary teeth and orthodontic traction, a combination of orthodontic-prosthodontic treatments is necessary.

Preventive programme for osteoporosis should be initiated at a young age since peak bone mineral density is achieved in the second and third decade of life so treatment with calcium and vitamin D supplementation should be considered when bone density is low. So, early diagnosis of CCD is beneficial for prompt intervention and better restoration of craniofacial aesthetics and function [13].

Affected individuals with cleidocranial dysplasia should be managed interdisciplinary by a team of specialist.Infants should receive their first dental evaluation by one year of age, preferably by a pediatric dentist who is experienced to treat children with complex problems. In young children with CCD, the fontanels may be so large that they should ware helmet to protect the brain. Hearing tests should be performed by Ear Nose Throat (ENT) specialist at birth and regularly every year thereafter, because they may have recurrent middle ear infection related to abnormal formation of palate or eustachian tube dysfunction which may necessitate the placement of tympanostomy tubes [14]. Individuals with CCD are more prone to have upper airway obstruction, and sleep habits must be carefully monitored by pediatric surgeon.

Management of this case

Since management varies from individual to individual a stage wise management is considered in this case. In first stage it involves bonding and banding of orthodontic attachments for both upper and lower arch including existing primary and permanent teeth which is the main rigid arch wire. Surgical exposure of the lower right first permanent molar, followed by bonding molar tube for the same. Then uprighting and extrusion of lower right first permanent molar is carried out through orthodontic mechanics by placement of continuous arch wire. Sequence of arch wire used are owen 6 Niti, 17 x 25 Niti, 19 x 25 Niti, 19 x 25 Stainless steel wire (SS Wire) for levelling and alignment. This approach reduces the physcological trauma to the patient by disipmacting the individual teeth so it does not impair the function and esthetics of the patient.

Second stage is followed by placement of 19 x 25 SS wire after completion of alignment and levelling for both arches. As the patient is in her third decade of life, removal and surgical exposure of retained deciduous teeth, impacted supernumerary and permanent teeth needs to be carried out

according to the sequence of eruption and not according to age.

After surgical exposure of teeth according to sequence of eruption, each tooth is brought to occlusal level with the help of elastics tied to the main rigid arch wire. As lower anteriors have no preceding primary teeth, they are surgically exposed and brought into occlusion with orthodontic attachments, elastics and were included into the main arch wire and the anchorage is reinforced. The same procedure for upper anteriors needs to be done with the removal of retained deciduous teeth, followed by the removal of lower primary first and second deciduous molar and impacted supernumerary premolar. Surgically exposing the impacted lower premolar unilaterally or bilaterally depending on the compliance of the patient needs to be carried out followed by bonding of orthodontic attachment to the impacted teeth. Then teeth are pulled to the occlusal level by using elastics tied to the main arch wire. Once lower premolar comes into occlusion they are included in to the main arch wire and anchorage is reinforced. The same procedure is followed for upper premolars. Second molars are up righted with implant supported uprighting spring made up of 17 x 25 TMA wire. Third molars which are impacted are advised for extraction because they are not favourable. After completing orthodontic treatment, contouring of malformed teeth will be carried out with composites and crowns for esthetic purpose as prosthodontic management. Permanent lingual retention will be placed in both upper and lower arches. The new modified treatment approach is an ideal approach in the present case as it provides better anchorage system to disimpact the impacted teeth.

Conclusion

It is important to note that people with CCD are expected to be shorter than their typical peers and family members. With proper clinical, radiological diagnosis and anticipatory guidance, people with CCD usually lead healthy and productive lives.

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