Research Article

Non-syndromic supernumery maxillary molars- an unusual case report

E. Elavarasi, V. Sadesh Kannan*, T. Saravanan, SP. Indira Kumar, G. Suresh Kumar and E. Gayathri Priyadarshini

ABSTRACT

A supernumery tooth is one that is additional to the normal series and can be found in almost any region of the dental arch. They may be single or multiple, unilateral or bilateral, erupted or unerupted and is one or in either jaws. The cases of multiple supernumery molars which is unusual and rare in individuals without any syndrome. This case report presents multiple permanent molars in a 15 year old male patient.

INTRODUCTION

Supernumery teeth are described as the teeth formed in excess number found in a normal dentition .They are more common in permanent dentition with males affected twice more commonly than females. The most common site for multiple supernumery teeth is found be mandibular pre-molar region. Even though the etiology of supernumery tooth is not clear a few hypothesis are put forward - Dichotomy of tooth bud; Local, independent or conditional hyperactivity of dental lamina; Genetic causes; & Atavism.

Supernumery teeth can be in two ways first according to the location – Mesiodens (Present in the incisor region); Paramolar (Present besides a molar); Distomolar (Present distal to molar); & Para-premolar (Present beside a premolar), second according to shape - Conical (Peg shaped); Tuberculate (Made of more than one cusp or tubercle); Supplemental (Tubercle resembles normal teeth); & Odontome (Seen as mass of dental tissues).

Multiple supernumery teeth can be seen associated with syndrome like: Cleidocranial dysplasia; Gardener's Syndrome; Down's syndrome; Crouzon's Disease; & Orodigitofacial Dystosis. It is very rarely found in individuals with no other associated syndromes.

CASE REPORT

A 15 year old male patient reported to our dental hospital with complaint of many teeth in upper right back tooth region. On extra –oral examination the face appears asymmetrical with both the eyes not in line with each other and in medial canthus of left eye drooping downwards (Figure 1) .

Intra-oral examination reveals full set of permanent dentition except 3rd molars with supplemental molars in the first quadrant which are four in number. Out of these, three molars are seen palataly to second pre-molar , first molar and second molar .Fourth molar is seen palataly placed to mesial most para-molar. Right posterior-lateral aspect of hard palate is occupied by these para-molars (Figure 2). All the four supplementary teeth shows morphological Variation from normal with presence of numerous fissures and cusps on the occlusal surface.

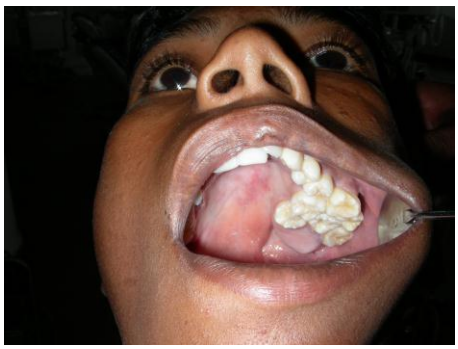

Fig. 1: Front view

Fig. 2: Intra-oral view of Maxilla

The condition couldn't be correlated with any syndrome , thus diagnosed as " MULTIPLE SUPERNUMERY MOLARS''. Such a case of multiple supernumery molar, not associated with any syndrome is of rare occurrence.

We have planned to go for extraction of supernumery molars out of the arch under local anaesthesia and sent for orthodontic opinion & management for Malocclusion (Figure 3).

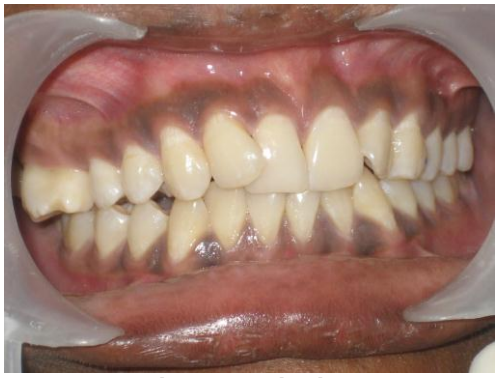

Fig. 3: Occlusion

DISCUSSION

Crouzon syndrome[1] is a genetic disorder known as a brachial arch syndrome. Specifically, this syndrome affects the first brachial (or pharyngeal) arch, which is the precursor of the maxilla and mandible. Since the brachial arches are important developmental features in a growing embryo, disturbances in their development create lasting and widespread effects.

This syndrome is named after Octave Crouzon[1], a French physician who first described this disorder. He noted the affected patients were a mother and her daughter, implying a genetic basis. First called "craniofacial dysostosis", the disorder was characterized by a number of clinical features. This syndrome is caused by a mutation in the fibroblast growth factor receptor II, located on chromosome 10.

Breaking down the name, "craniofacial"[2] refers to the skull and face, and "dysostosis" refers to malformation of bone.

Now known as Crouzon syndrome, the disease can be described by the rudimentary meanings of its former name. What occurs in the disease is that an infant's skull and facial bones, while in development, fuse early or are unable to expand. Thus, normal bone growth cannot occur. Fusion of different sutures leads to different patterns of growth of the skull. Examples include: trigonocephaly[3] (fusion of the metopic suture), brachycephaly[3,5] (fusion of the coronal suture), dolichocephaly[4] (fusion of the sagittal suture), plagiocephaly[5] (unilateral premature closure of lambdoid and coronal sutures), oxycephaly[3] (fusion of coronal and lambdoidal sutures), Kleeblattschaedel[6] (premature closure of all sutures)

Associations with mutations in the genes of FGFR2[7] and FGFR3[7] have been identified.[5] [6]

Crouzon syndrome is autosomal dominant; children of a patient have a 50% chance of being affected.

As a result of the changes to the developing embryo, the symptoms are very pronounced features, especially in the face. Low-set ears are a typical characteristic, as in all of the disorders which are called brachial arch syndromes. The reason for this abnormality is that ears on a fetus are much lower than those on an adult. During normal development, the ears "travel" upward on the head; however, in Crouzon patients, this pattern of development is disrupted. Ear canal malformations are extremely common, generally resulting in some hearing loss. In particularly severe cases, Ménière's disease may occur.

The most notable characteristic of Crouzon syndrome is cranial synostosis, as described above, but it usually presents as brachycephaly, which results in the appearance of a short and broad head. Exophthalmos (bulging eyes due to shallow eye sockets after early fusion of surrounding bones), hypertelorism[8] (greater than normal distance between the eyes), and psittichorhina (beak-like nose) are also symptoms. Additionally, a common occurrence is external strabismus, which can be thought of as opposite from the eye position found in Down syndrome. Lastly, hypoplastic maxilla (insufficient growth of the midface) results in relative mandibular prognathism (chin appears to protrude despite normal growth of mandible) and gives the effect of the patient having a concave face. Crouzon syndrome is also associated with PDA (patent ductus arteriosus) and aortic coarctation.

For dentists, this disorder is important to understand since many of the physical abnormalities are present in the head, and particularly the oral cavity. Common features are a narrow/high-arched palate, posterior bilateral crossbite, hypodontia (missing some teeth), and crowding of teeth. Due to maxillary hypoplasia, Crouzon patients generally have a considerable permanent underbite and subsequently cannot chew using their incisors. For this reason, Crouzon patients sometimes eat in an unusual way--eating fried chicken

with a fork, for example, or breaking off pieces of a sandwich rather than taking a bite in it.

CONCLUSION

For reasons that are not entirely clear, most Crouzon patients also have noticeably shorter humerus and femur bones relative to the rest of their bodies than members of the general population. A small percentage of Crouzon patients also have what is called "Type II" Crouzon syndrome, distinguished by partial syndactyly[3].

This case is also not clear crouzon syndrome except the supernumery molars[9], high arch palate with posterior cross bite, midline shift of upper & lower incisors (Figure # 3), asymmetrical nose and lower lip, hypertelerorism and drooping of left medial canthal ligament. Diagnosis of Crouzon syndrome usually can occur at birth by assessing the signs and symptoms of the patient. Further analysis, including radiographs, Magnetic Resonance Imaging[3] (MRI) scans, genetic testing, X-rays and CT scans can be used to confirm the diagnosis of Crouzon syndrome.

ACKNOWLEDGEMENT

1) Dr.A.S.Ganesan, Honourable Chancellor, Vinayaga mission University, Kilpauk, Chennai, for his kind support, &
2) Prof.Dr.P.Anusuya, Dean, Arupadaivedu Medical College & Hospital, Puducherry, for his eminent guidance,

REFERENCES

1. Tanwar R[1], Iyengar AR. Crouzons syndrome: a case report with review of literature. J Indian Soc Pedod Prev Dent. 2013 Apr-Jun; 31(2):118-20.
2. Buchanan EP[1], Xue AS. Craniofacial syndromes. Plast Reconstr Surg. 2014 Jul; 134(1):128e-153e.
3. Paritosh C Khanna[1], Mahesh M Thapa[2], Ramesh S Iyer[2], Shashank S Prasad[3] .Pictorial essay: The many faces of craniosynostosis.Indian Journal of Radiology and Imaging, Vol. 21, No. 1, January-March, 2011, pp. 49-56
4. Beckett JS, Pfaff MJ. Dolichocephaly without sagittal craniosynostosis. J Craniofac Surg. 2013 Sep; 24(5):1713-5.
5. Schulz M, Spors B. Results of posterior cranial vault remodelling for plagiocephaly and brachycephaly by the meander technique. Childs Nerv Syst. 2014 Sep; 30(9):1517-26.
6. Tubbs, RS Sharma. Kleeblattschädel skull: a review of its history, diagnosis, associations, and treatment. . Childs Nerv Syst. 2013 May; 29(5):745-8.
7. Fenwick AL, Goos JA. Apparently synonymous substitutions in FGFR2 affect splicing and result in mild Crouzon syndrome. BMC Med Genet. 2014 Aug 31;15:95.
8. Miamoto CB[1], Soares RL. Ocular hypertelorism in an orthodontic patient. Am J Orthod Dentofacial Orthop. 2011 Apr; 139(4):544-50.
9. Menardia-Pejuan V, Berini-Aytes L. Supernumerary molars. A review of 53 cases. Int Res Sci Stomatol. 2000;402(2-3):101-5.