ORAL MEDICINE

Cardiac, Dental and Skeletal Anomalies: Proposal of a New Syndrome

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ABSTRACT

Background: **Dental anomalies are the formative defects that occur as a result of multifactorial interactions between genetic or environmental factors during the process of tooth development. Literature has revealed presence of multiple dental anomalies such as supernumerary tooth, tooth agenesis, ectopic tooth eruption, atypical tooth size or shape, taurodontism, pulp stones etc in various syndromic or non-syndromic conditions. Although asymptomatic, but early diagnosis is requisite for better patient management to prevent the occurrence of associated complications.**

Material and Methods: **Here, we report a rare case of 23-year-old male patient who presented with cardiovascular manifestations along with multiple dental and skeletal findings.**

Conclusion: **Such combination of dental and skeletal findings has not been reported earlier in association with any individual syndrome. Hence, we propose it to be a new syndrome of non-genetic type associated with multiple cardiac, skeletal and dental anomalies.**

KEY WORDS

cardiovascular manifestations, cone beam computed tomography, dental anomalies, pulp stones, supernumerary teeth

INTRODUCTION

Dental anomalies are the formative defects which are secondary to genetic, epigenetic or environmental factors that may occur during the process of tooth development. Presence of multiple dental anomalies such as supernumerary tooth, tooth agenesis, ectopic tooth eruption, atypical tooth size or shape, taurodontism, pulp stones etc have been reported in the clinical practice. However, compared to the more common oral diseases such as dental caries and periodontal diseases, incidence of dental anomalies is low and varies among different population groups[1)]. Although condition is asymptomatic, but can result in malocclusion, aesthetic and functional problems during treatment planning, therefore early diagnosis plays a key role towards optimal patient management and treatment outcome[2)].

A syndrome is a collection of signs and symptoms that are noted in, and characteristic of, a single condition. In medical genetics, a syndrome is referred specifically to any medical condition where the underlying genetic cause has been recognised, and the set of symptoms is pathogenetically related. Examples of syndromes in medical genetics includes, Down syndrome, Stickler syndrome, and Williams syndrome, whereas certain conditions like Toxic shock syndrome and Acquired Immune Deficiency Syndrome are examples of non-genetic syndromes[3)]. If the underlying genetic cause is not clearly established, the condition is referred to as an "association". By definition, an association represents the collection of signs and symptoms that occur in combination more frequently than would be likely by chance alone[3)]. Multiple dental anomalies in individuals or families, are commonly seen with other systemic manifestations or syndromes[4)].

Here, we report a rare case of 23-year-old male patient who presented with cardiovascular manifestations like ventricular septal defect, atrial septal defect, patent ductus arteriosus, right coronary cusp prolapse, systolic anterior motion, severe aortic regurgitation, severe pulmonary arterial hypertension and sinus rhythm, along with dental anomalies like supernumerary teeth, anterior open bite, large overjet, proclined upper and lower incisors, cleft tongue, protruded upper and lower lip, teeth with enlarged pulp space, pulp stones, and skeletal findings like hypodivergent skeletal and facial pattern, acute gonial angle and skeletal class II Div 1 malocclusion. Such combination of dental and skeletal findings has not been reported earlier in association with any individual syndrome. Hence, we propose a new syndrome associated with multiple cardiac, skeletal and dental anomalies.

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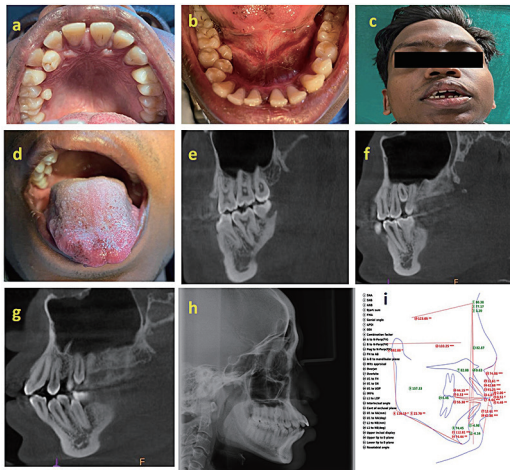

Figure 1: 1a: Supernumerary teeth in maxillary arch; 1b: Supernumerary teeth in mandibular arch; 1c: Anterior open bite, proclined upper and lower incisors, protruded upper and lower lip; 1d: Bifid tongue; 1e: Saggital section of CBCT showing enlarged pulp space and pulp stones; 1f: CBCT showing supernumerary tooth in upper arch; 1g: CBCT showing supernumerary tooth in lower arch; 1h: Lateral cephalogram of patient; 1i: Cephalometric analysis

CASE REPORT

A 23-year-old male patient presented with the complaint of forwardly placed upper front teeth. Medical History revealed that patient had ventricular septal defect, atrial septal defect, patent ductus arteriosus, right coronary cusp prolapse, systolic anterior motion, severe aortic regurgitation, severe pulmonary arterial hypertension and sinus rhythm. Patient was treated with aortic valve replacement with St. Jude aortic valve prosthesis (size 25 mm), pericardial patch closure of ventricular septal defect, subaortic membrane excision, direct closure of atrial septal defect, and ligation of patent ductus arteriosus was done. Past dental history, family and personal history was non-contributory. On General physical examination no abnormality was detected and all the vital signs were within the normal limits. Extraoral examination revealed that he had a convex facial profile, symmetrical face, incompetent lips, and everted upper and lower lip. Intraoral examination revealed two supernumerary teeth palatal to 13 and 16 (Figure 1a), and two supernumerary teeth lingual to 45 and 46 (Figure 1b) Along with this, anterior open bite, proclined upper and lower incisors, increased overjet, (Figure 1c), and cleft tongue (Figure 1d) was noted.

Cone beam computed tomography (CBCT) revealed enlarged pulp space of molar teeth (Figure 1e), pulp stones (Figure 1e) and supernumerary teeth (Figures 1f and 1g). Linear and Angular measurements performed on Lateral Cephalogram revealed hypodivergent skeletal and facial pattern, acute gonial angle and skeletal class II div 1 malocclusion (Figures 1h and 1i). Based on the patients history, typical clinical features and radiographic examination we hypothesize it as a case of new syndrome dento-skeletal syndrome of non-genetic type. Patient was advised extraction of supernumerary teeth and orthodontic treatment and orthognathic surgery was planned to correct skeletal Class II Malocclusion.

DISCUSSION

Prenatal hypoxia has been known to cause intrauterine retardation of growth. It has been found to adversely affect the myocardial function, thereby resulting in decrease in cardiac performance. Study by Yang *et al* has shown that prenatal hypoxia might affect the fetal skeletal growth. Although underlying mechanism is still unclear, but prenatal hypoxia has been found to retard fetal skeletal growth in rats, it inhibited the synthesis of extracellular matrix and down-regulated insulin-like growth factor 1 signalling in fetal growth plate chondrocytes[5]. To the best of our knowledge only one report available in the literature by Kaner *et al*., described the oral manifestations of congenital heart disease. Marked enlargement of the pulp chamber of the maxillary primary and permanent incisors on periapical radiographs in patients with congenital heart disease was observed, the findings suggested that the enlargement of the root canals may be secondary to marked arterial hypertension[6]. Similarly in the present case, enlarged pulp space of molar teeth known as Taurodontism was observed on the CBCT sagittal sections which may be secondary to the severe arterial hypertension in the patient.

Taurodontism is morpho-anatomical alteration in the usual contour of teeth in which the body of a tooth is enlarged with the decrease in roots size[2]. The pulp size of the taurodont teeth is enlarged in size with an increased vertical dimension and lacking the normal constriction at the cementoenamel junction[3]. The aetiology of taurodontism is not precisely understood. It has been proposed that failure of Hertwig's epithelial sheath diaphragm to invaginate at the proper horizontal level, may lead to the decreased root size and increased dimension of the tooth body and pulp cavity[4,5]. This condition has been also affiliated with several developmental anomalies and syndromes like Ackerman syndrome, Amelogenesis imperfecta, type IA and IV, Cerebellofaciodental syndrome, Dentin dysplasia, type 1, Hyperphosphatemic familial tumoral calcinosis 1, Hypohidrotic X-linked ectodermal dysplasia, MOMO syndrome, Oculodentodigital dysplasia, Otodental syndrome, Taurodontia-absent teeth-sparse hair syndrome[3-5]. Another peculiar finding bifid or cleft tongue (glossoschissis) was noted in our patient which is a groove or split running lengthwise along the tip of the tongue as a result of incomplete fusion of the distal tongue buds. Syndrome most commonly associated with bifid tongue is Orofacial-digital syndrome I, III and IV. Various other syndromic and non-syndromic conditions to have bifid tongue as an unusual finding include Robinow syndrome, autosomal recessive and dominant types 1 and 3, and Short-rib thoracic dysplasia's 3 and 13 with or without polydactyly[7].

Association between pulp stones and cardiovascular disorders has been well documented in the literature[8]. Pulp stones are calcified structures seen in coronal or radicular pulp of healthy, diseased and even unerupted teeth and may be found both in deciduous and permanent dentition[1]. Studies have reported that prevalence of pulp stones increases with age with higher incidence of pulp stones is in molars[1,8]. Presence of pulp stones in our patient further supports the association of cardiovascular disorder with an increased incidence of pulp stones. In addition, presence of pulp stones has been noted in certain syndromes like Down's syndrome, Klinefelter's syndrome, Saethre-Chotzen syndrome, Elfin facies syndrome, Ehlers Danlos syndrome type I, Otodental syndrome[1].

Radiographs play an important role in early detection of pulp stones in systemic diseases which appear as discrete single or multiple structures within the coronal or radicular pulp on periapical or bite wing radiographs[8,9]. Recently, CBCT is becoming popular in identification of pulp stones due to its potential advantages of providing three dimensional images of high resolution and without superimposition of structures. To this consideration we evaluated the pulp stones by use of CBCT, which were seen as discrete radiopaque calcifications in the pulp chamber of maxillary and mandibular molar on sagittal CBCT sections[9].

Supernumerary teeth palatal to 13 and 16 and lingual to 45 and 46 was another clinical problem reported by our patient. Supernumerary tooth is an odontostomatologic anomaly characterized by an excessive number of teeth in relation to the normal dental formula[10]. Congenital genetic disorders that may show an association with multiple supernumerary teeth include Gardner's syndrome, cleidocranial dysostosis and cleft lip and palate. Whereas less commonly syndromes associated with this condition include Fabry Disease, Ellis-van Creveld syndrome, Nance-Horan syndrome, Rubinstein-Taybi Syndrome and Trico–Rhino–Phalangeal syndrome[10]. Early diagnosis and treatment of supernumerary teeth is essential to prevent the associated complications of delayed eruption, displacement, rotation, crowding and root resorption of adjacent permanent teeth. In the present case, extraction of the supernumerary teeth was planned followed by orthodontic referral for correction of skeletal Class II malocclusion.

Anterior open bite characterized by lack of upper and lower incisor contact, no vertical overlap of the lower incisors by the upper incisors, and contact of occlusal surfaces of the posterior teeth was noted in our case. Anterior open bite has multifactorial aetiology, but can be broadly described as being dental or skeletal in origin. Various syndromes such as Crouzon syndrome, Beckwith Wiedman Syndrome, Treacher Collin Syndrome, Down Syndrome, Turner Syndrome, Gorlins Syndrome, Nooman Syndrome, Maroteaux Lamy Syndrome, Lennox Gastant SyndromeMoebus Syndrome are known to be associated with an anterior open bite. The severity of anterior open bite should be taken into

account during orthodontic treatment planning which varies from an almost edge-to-edge relationship to a severe handicapping open bite. Anterior open bite with no skeletal anomaly has been reported to be self-correcting, however cases with hypodivergent growth pattern and skeletal class II malocclusion require orthodontic correction[11,12].

Considering all above facts and after reviewing the literature, to our best knowledge such combination of cardiac, dental and skeletal findings has not been reported, and characteristic dental findings seen in this case are rare and differs from the previously reported cases hence we propose it to be a new syndrome, "Dento-Skeletal Syndrome" of non-genetic type.

CONCLUSION

The combination of dental anomalies, as seen in this case, probably indicates a common unknown genetic factor in the aetiology giving rise to different phenotypic manifestations. The constellation of dental findings seen in this case is certainly rare and differs from previously reported cases. The case is also sporadic, with no positive family history. Mutations in developmental regulatory genes are known to cause a variety of dental defects. The wide variation in clinical manifestations in cases of non-syndromic occurrence of multiple dental anomalies remains intriguing and is an area for further research. For the clinician, the difficulty lies in diagnosis, as a number of syndromes and pathologies need to be ruled out, due to large constellation of dental findings. Such a case needs to be diagnosed early and requires multidisciplinary treatment approach.

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