Sequential Supernumerary Teeth Development in a Non-Syndromic Patient; Report of a Rare Case

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Abstract

Isolated impacted supernumerary teeth are quite rare, but they can be seen associated with several syndromes such as cleidocranial dysostosis or Gardner’s syndrome. This article aims to discuss a case of sequential formation of supernumerary teeth with no other associated disease or syndrome.

A 17-year-old Iranian male with 8 impacted supernumerary teeth was referred to the department of pediatric dental clinic at Shahid Beheshti Medical University in Tehran with a history of several impacted unerupted teeth. Repeated and periodical clinical and radiographic examinations revealed newly formed teeth buds in unusual dental ages. All extra teeth were associated with generalized enamel hypoplasia to some degree on their relative permanent adjacent teeth. The patient did not have any record of a systemic disease or any syndromic condition to relate his dental problem to.

This rare condition involved repeated and continued formation of extra teeth out of the normal numbers and dental age evident in serial radiographs.

Key Words: Tooth, Supernumerary; Serial; Syndrome; Child

INTRODUCTION

Supernumerary teeth (ST) are defined as an extra set of teeth that are counted more than normal count in the component of primary and/or permanent dentition [1]. To date several different theories have suggested varying etiologies; factors including: 1. abnormality or hyperactivity of the dental lamina, [2] reactivation of the remaining epithelium after break up in the tooth band (dichotomy theory) [3], and 3. The process of atavism, the return to or appearance of an ancestral condition or type [4]. In addition, familial tendency and racial influences should also be noted when looking at the cause [5].

The formation of multiple supernumerary teeth (MST) does not follow the normal path in Mandelian pattern [6].
The anterior maxillary region appears to be the most usual site of occurrence with mesiodense being the most common ST located at and around the palatal midline [7]. The prevalence of ST is reported between 0.3% and 3.8% in America with the male population being affected twice as frequently as the female group [1]. ST may occur in single or multiple patterns, unilateral or bilateral. Multiple ST is counted for 14% of all ST cases, [1] while it is rarely found in individuals with no other associated syndrome [8]. The most known syndrome associated with ST is cleidocranial dysplasia. There are several other syndromes with a lower frequency of association with ST including: Apert syndrome, Gardner syndrome, Down syndrome and Crouzon disease (Table 1 and 2).

The size and shape of ST is different, but in certain cases it is close to the normal adjacent teeth, the so-called supplemental teeth. They may be as variable as those with tuberculates (more than one cusp or tubercle) or even conical (peg in shape) [9]. Several methods have been introduced to classify these extra teeth: 1. Non-sequential in which all ST are developed in a short period of time with no further formation of any tooth bud after a certain age [2]. Sequential where tooth formation will continue for a longer period with newly formed tooth buds appearing in a later stage of life [10, 11-14]. MST has a direct influence on the treatment protocol and its outcome [15].

The purpose of this article was to report a rare case of sequential MST in a non-syndromic Iranian patient.

CASE REPORT
A 16-year-old male patient was referred to the pediatric dental clinic at Shahid Beheshti Medical University, Tehran in fall 2008 with failure of eruption of several upper and lower permanent teeth (Fig 1).

The patient was the second child in his family with no trace and history of any similar condition in other family members. Medical and laboratory test histories revealed no specific medical condition with an overall normal health. The level of intelligence and mental development was assessed as normal based on Wechsler criteria. Physical examination revealed a thin body, slight hyperlaxity in the finger joints raising the possibility of joint problem, later ruled out by a full orthopedic evaluation. Results of a general laboratory test including the thyroid gland function revealed no problem.

The present teeth were as follows in the first visit:

| 6 | E | D | C | B | A | 1 | B | C | D | E |
|---|---|---|---|---|---|---|---|---|---|---|
| 7 | 5 | 4 | 3 | 2 | 1 | 1 | C | 5 |

Enamel hypoplasia was noted in most of the erupted teeth. Severe caries was noted especially in retained primary teeth. The 1st upper left and 1st lower right permanent molar teeth were absent as extracted earlier due to severe caries (Fig 2).

Radiographic evaluation of the chest and wrist showed no sign of abnormality. Panoramic views (three year apart) were also available for evaluation of the patient’s development progress (Figure 3, 4 and 5).
The first radiograph was taken when he was 11 years old, at which three supernumerary teeth were located at the upper anterior and right premolar regions namely (Fig 3).

\[
\begin{array}{cccc}
4' & 1'' & 1' & 4' \\
4' & 4' & 5' \\
\end{array}
\]

The second panoramic radiograph was taken at 14 years of age showing four other supernumerary teeth bilaterally in the mandibular premolar and left maxillary premolar areas as indicated below:

\[
\begin{array}{cccc}
4' & 4' & 5' \\
4' & 5' \\
\end{array}
\]

Interestingly, no signal of any of these teeth could be traced in earlier radiographs. The patient’s history revealed that the right and left upper 6 was extracted due to severe caries (Fig 4).

New supernumerary teeth were observed in the third radiograph taken at 17 years of age that did not have any trace at earlier radiographs.

The locations of these extra teeth were defined between 1 and 1'. This newly appeared tooth was named as 1''.

The upper left maxillary central incisor was present at the time of oral examination while the right central and both upper lateral insisors remained unerupted.

In addition, the lower left lateral incisor was also present in the oral cavity (Fig 5). There was a total number of 8 extra teeth in the jaw. The collective figure of these extra teeth are shown as:

\[
\begin{array}{cccc}
4' & 1'' & 1' & 4' \\
4' & 1' & 4' & 5' \\
\end{array}
\]

Consultations were sought from oral surgery and orthodontic departments in order to plan the best treatment option for the patient. An overall agreement was achieved to remove all the malformed and malposed teeth including: several unsavable permanent teeth as listed below:

\[
\begin{array}{ccccccc}
4 & 1 & B & 6 & 7 \\
2 & 4' & 5' & 6 & 7 \\
\end{array}
\]

The treatment process included a phase of multiple extractions under general anesthesia as several surgical sites needed intervention. This was later followed by an initial step of orthodontic forced tooth movement in order to encourage the eruption of impacted teeth including:

\[
\begin{array}{cc}
32 & 34 \\
4 & \\
\end{array}
\]

The patient was discharged the day after the operation with his orthodontic visits set along with a full mouth follow up at 6 and 12 months.
The patient’s current status is as close to normal with an appropriately aligned set of teeth in order as shown in Fig 6.

DISCUSSION
Supernumerary teeth are usually seen along with and at the same time as the normal teeth. They can also be seen associated with certain syndromes.
The rate of multiple supernumerary teeth has been reported as 23% among all supernumerary cases [4,16].
An important point is that the development of 5 supernumerary teeth of this report was noticeable over a 5-year period (sequential supernumerary teeth formation).
Multiple nonsequential, non-syndromic supernumerary teeth tend to occur more in the mandibular premolar area, as recently reported [17-21]. (Table 3).

Both premaxillary and mandibular premolar regions were equally affected in the current case, followed by maxillary premolar region with a lower incidence (less affected teeth). Gomes et al. (2008) reported three cases of sequential supernumerary teeth in mandibular premolar and premaxillary regions considered as the most frequent areas for these teeth to be detected respectively. The maxillary premolar region was, however, the site least affected. Supernumerary teeth of the current case were more frequent in the maxilla (five in the maxilla and 3 in the mandible) as seen in other reports [9].
Non-syndromic multiple supernumerary teeth are more likely to occur in patients whose other relatives are also involved [2]. However, the patient in the current case did not have a positive family history similar to the case reported by Gomes et al. (2008) [9].

| Syndrome’s Name                      | Clinical Manifestation                                                                 |
|-------------------------------------|----------------------------------------------------------------------------------------|
| Apert syndrome                      | Scaphocephaly, craniosynostosis, bilateral syndactyl, midface hypoplasia                |
| Cleidocranial dysplasia             | Aplastic clavicles, frontal bossing, hypoplastic midface                                |
| Gardner syndrome                    | Osteomas, epidermoid cysts, odontomas, intestinal polyps                                |
| Down syndrome                       | Brachycephaly, mental retardation, epicanthal folds                                    |
| Crouzon disease                     | Craniosynostosis, exophthalmos, hypoplastic midface                                    |
| Sturge-weber syndrome               | Angiomatosis and calcification of leptomeninges, seizures, port-wine nevi of face       |
| Oral-facial-digital syndrome        | Hypoplastic alar cartilage, cleft tongue, clinodactyly                                 |
| Hallermann-streiff syndrome          | Dysephaly, mandibular hypoplasia, hypotrichosis                                         |

Table 1. Classic syndrome with multiple supernumerary teeth

Fig 3. First panoramic radiograph with teeth present
Continuous supernumerary teeth formations are thought to be due in part to reactivation of the tooth follicle with no clear explanation. Another possible mechanism is unresorbed remained dental lamina. This could lead to reactivation of the normal permanent teeth at the time of crown completion. Lastly it is thought that the crypts of the supernumerary teeth may have been present but were not detectable on earlier radiographs [2]. The Wnt gene has been considered another likely etiology for supernumerary teeth [22]. Jarvinen et al. (2006) stated that Wnt signaling has been associated with tooth renewal in the human. Tooth bud expressing stabilizes beta-catenin (intracellular mediator of Wnt signaling) in the epithelium, leading to the formation of dozens of teeth. Wang et al. (2009) stated that even adult dental tissues could lead to the formation of new teeth in response to either loss of function of epithelial APC (adenomatous polyposis coli gene protein) or Wnt/beta-catenin activation [22].

Table 2. Rare syndrome with multiple supernumerary teeth and their characteristics

| Syndrome’s Name | Clinical Manifestation |
|-----------------|------------------------|
| Robinow⁹        | Short stature, hypertelorism, high nasal bridge, midface hypoplasia, microopenis |
| Ehler-Danlos¹⁰   | Skin hyperelasticity, Articular hypermobility, Vascular fragility |
| Nance-Horan¹¹   | Congenital cataract, Malformed teeth |
| G/BBB¹²         | Eye anomalies, Laryngotracheosophageal cleft, Congenital heart disease, Genitourinary anomalies, Gastrointestinal disorders |
| Zimmermann-Laband¹³ | Gingival fibromatosis, Absent or dysplastic distal phalanges, Vertebral defects, Hepatosplenomegaly, Ear, Nose, Bone, and Nail defects |
| Leopard¹⁴       | Multiple lentigines, Ocular hypertelorism, Delayed secondary sexual characteristics, Mild cardiac abnormalities |
| Enamel Dysplasia with Hamartomatous atypical Follicular Hyperplasia(EDHFH)¹⁵ | (only In black south Africans) Hamartomatous follicular hyperplasia, Central odontogenic fibroma(WHO type), Enamel Dysplasia |
| Tricho-Rhino-Phalangeal¹⁶ | Slowly growing sparse hair, Medially thick and laterally thin eyebrows, Bulbous tip of the nose, Long flat philtrum, Thin upper lip with vermilion border, Prominent ears, Cone-shaped epiphyses |
| Kabuki make up¹⁷ | Cleft lip/palate high-arched palate, Bifid tongue and uvula, anterior open bite, overretention of primary teeth, Long palpebral fissure, hypertelorism, palpebral ptosis, broad, depressed nasal root with a flat nasal tip, mild neuropsychomotor developmental deficits |
It is concluded that a small number of APC deficient cells is enough to induce the surrounding wild-type epithelial and mesenchymal cells to participate in the formation of new teeth [23].

The presence of general enamel hypoplasia of supernumerary and permanent teeth was another important finding in the current case with no report of such findings by other investigators. The presence of multiple supernumerary teeth would routinely suggest an underlying genetic disorder [22, 23]. It is important to mention that some eruption disturbances may be seen with the presence of supernumerary teeth. Any orthodontic force effect needs to be fully investigated in such individuals.

When supernumerary teeth are found a prompt decision should be made in order to decide their removal. Uncontrolled removal of supernumerary teeth could lead to further complications affecting adjacent structures. It is therefore important to determine if the risks of surgery outweigh the benefits of removal. Due to the high number of supernumeraries and impacted teeth, a stepwise therapy was planned in this case with extraction of the selected number of supernumerary teeth being the starting point followed by forced eruption as the next phase.

Table 3. Recently Reported non Syndromeical Multiple Supernumerary teeth

| Author                  | year | Age / Gender | Number | location                                                                 |
|-------------------------|------|--------------|--------|--------------------------------------------------------------------------|
| Batra et al8             | 2004 | 17/ girl     | 11     | One mesiodens,two upper premolar,two lower anteriors,six lower premolars |
| Arathi & Ashwini17       | 2005 | 14/ boy      | 12     | One lower anterior, five lower premolars, three upper anteriors,three upper premolars |
| Sivapathasundharam & einstein18 | 2007 | 20/ boy     | 14     | Nine lower premolars,three upper premolars,one upper anterior, one lower anterior |
| Aravindha & Srivatsa19   | 2007 | 19/ girl     | 10     | One mesiodens, one upper anterior,two lower canines,,four lower premolars, two lower distomolars(one in each side) |
| Rasi Beyer G20           | 2008 | 21/ boy      | 5      | One upper anterior, Four upper premolar                                  |
| Diaz et al21            | 2009 | 20/ girl     | 17     | twelve upper(premolars and distomolar), five lower premolars             |

Fig 5. Third panoramic radiograph showing further teeth formation at this stage.
CONCLUSION
A case of repeated development of several supernumerary teeth was noted with no history of any syndrome or medical condition. Periodic routine radiographic images confirmed the case as a rare but non-syndromic individual with sequential supernumerary teeth formation. Treatment includes elimination of these supernumeraries in order to help retain the teeth and the surrounding structures.

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