Nonsyndromic Familial Hyperdontia: Two Case Reports and Review of Literature

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Abstract
Nonsyndromic familial hyperdontia/supernumerary teeth is a rare disorder of odontogenesis characterized by an excess number of teeth with familial background. They can be supplemental (resembling natural teeth), conical, tuberculate or molariform.

Early clinical and radiographic examination of the supernumerary teeth allow for optimal yet minimal treatment. This article reports two rare cases of nonsyndromic hereditary hyperdontia both in the maxillary anterior region. First case report with bilateral mesiodentes palatal to the central incisors in sister and brother and second case report with presence of fused mesiodentes in father and conical mesiodens in son.

Though the exact etiology of this dental anomaly remains unclear, genetics as key factor in development of supernumerary teeth is highlighted.

Keywords: Supernumerary teeth, tuberculate tooth, mesiodens.

INTRODUCTION
Hyperdontia is the condition of having supernumerary tooth/teeth, which appear in addition to the regular number of teeth.1 Around 90 to 98% of supernumerary teeth occur in maxilla and 90% of these are restricted to premaxilla.2 The most common supernumerary tooth is mesiodens, which is a malformed, peg-like tooth that occurs between the maxillary central incisors. The prevalence of hyperdontia is between 0.1 and 3.8% in various population and mesiodens accounts for one third of cases.3 Brook4 found that supernumerary teeth were present in 0.8% of primary dentition and in 2.1% of permanent dentition.

According to the morphology supernumerary can be conical, tuberculate, supplemental or molariform.5 Conical mesiodens is a small peg shaped tooth most commonly found in permanent dentition. It can cause rotation or displacement of permanent incisor, but rarely delays eruption. Tuberculate type of supernumerary possesses more than one cusp or tubercle. They are often paired and are commonly located on the palatal aspect of the central incisors. Root formation is delayed compared to that of permanent incisors. They rarely erupt and are frequently associated with delayed eruption of the incisors.6

Exact etiology of supernumerary teeth is not well-understood. Though many theories have been put forward to explain the anomaly based on developmental interference and hereditary. Autosomal dominant inheritance with incomplete penetration has been proposed genetic theory.7 The importance of early diagnosis of supernumerary teeth and the importance of family history and genetic factors as the possible cause of hyperdontia is presented in these case reports.

CASE REPORT 1
A 14-year-old female patient along with her 11-year-old brother and mother reported to the Department of Oral Medicine and Radiology, Desh Bhagat Dental College and Hospital, Muktsar, Punjab with the chief complaint of extra-teeth erupting behind upper front teeth. History of presenting illness revealed recurrent injury to the tongue while making tongue movements. Also there is lodgment of food between upper front teeth and erupting extra teeth leading to difficulty in chewing food. Patient visited the local dentist and was advised to wait for sometime. Family history revealed that child was the first of the two siblings born of a nonconsanguineous marriage, at full term by normal delivery. No significant postnatal illness was reported. The developmental milestones were normal. Child was of normal intelligence, doing well at school. Detailed investigation suggested missing upper front teeth in her brother’s mouth. There was no history of similar anomalies among her parents and forebearers.

Her medical history was noncontributory. General examination showed that patient was of normal gait, average built and moderately nourished. All the vitals were in normal limits. Extraoral examination did not show any abnormality. Intraoral examination revealed two palatally erupting diverging tuberculate mesiodentes behind 11 and 21 (Fig. 1) and dental caries of 27. Intraoral periapical radiograph of 11 and 21 region along with topographic occlusal radiograph of maxilla (Fig. 2) showed well-defined tuberculate mesiodentes with fully formed roots.

Intraoral examination of her brother showed mixed dentition with missing 11 21 and 25 (Fig. 3). IOPA of 11 21 region and
topographic occlusal view of maxilla showed four impacted teeth, i.e. 11 21 and two tuberculate mesiodens with open apical foramen in premaxilla region, erupting 25 (Fig. 4).

Based on the history clinical examination and radiographic investigation, final diagnosis of nonsyndromic familial hyperdontia was made and patients were referred to pedodontia department for extraction for supernumerary teeth.

CASE REPORT 2

An 11-year-old male patient along with his mother reported to the Department of Oral Medicine and Radiology with the chief complaint of a tooth erupting behind upper front teeth leading to malalignment of teeth in the upper front teeth region. Patient’s parents noticed the erupting extra-tooth 5 to 6 months back resulting in unesthetic appearance of child. Patient never visited the dentist before. His medical history was non-contributory but detailed investigation into family history suggested the father to be having an odd looking extra-tooth in the upper midline (as narrated by the mother). There was no history of similar anomalies among their forebearers or the other sibling.

The father was requested to report on the next visit along his 11-year-old son.

Extraoral examination of son did not show any abnormality. Intraoral examination revealed mixed dentition stage. A palatally erupted tuberculate mesiodens was seen causing labial displacement of 21 (Fig. 5). IOPA of 11 and 21 region (Fig. 6) was noted to confirm the finding.

Intraoral examination of the father showed well-aligned mesiodens with abnormal morphology (Fig. 7) and missing 13 26, dental caries 16 47. Both soft and hard tissue deposits were present.

IOPA radiograph of 11 12 region and topographic occlusal radiograph of maxilla showed two fused mesiodens and congenitally missing 13 (Fig. 8). However, being quite satisfied with the appearance, he refused to undergo any treatment option explained to him.

Based on history, clinical examination and radiographic investigations, the final diagnosis of nonsyndromic familial hyperdontia was made. Patient was referred to pedodontia department for extraction of mesiodens and orthodontic treatment.
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tooth bud. Another theory, well supported in the literature, is the hyperactivity theory, which suggests that supernumeraries are formed as a result of local, independent, conditioned hyperactivity of the dental lamina. Heredity may also play a role in the occurrence of this anomaly, as supernumeraries are more common in the relatives of affected children than in the general population. However, the anomaly does not follow a simple Mendelian pattern, probably as an autosomal dominant gene with lack of penetrance in some generations; sex-linked inheritance with increased incidence in males to females (2:1), and phylogenetic process of atavism. Multiple supernumerary teeth are rare in individuals with no other associated diseases or syndromes. Further support of a genetic component in hyperdontia is evidenced by the simultaneous occurrence in identical twins.

CLASSIFICATION

Conical
This small peg shaped conical tooth is the supernumerary most common found in the permanent dentition. It develops with root formation ahead of or at an equivalent stage to that of permanent incisors and usually presents as a mesiodens. It may occasionally be found high and inverted into the palate or in a horizontal position. In most cases, however the long axis of the tooth is normally inclined, but rarely delays eruption.

Tuberculate/Molariform
The tubeculate type of supernumerary possesses more than one cusp or tubercle. It is frequently described as barrel shaped and may be invaginated. Root formation is delayed compared to that of the permanent incisors. Tuberculate supernumeraries are often paired and are commonly located on the palatal aspect of the central incisors. They rarely erupt and are frequently associated with delayed eruption of the incisors.

Supplemental
The supplemental supernumerary refers to a duplication of teeth in normal series and is found at the end of a tooth series. The majority of supernumeraries found in the permanent dentition are of the supplemental type and seldom remains impacted.

Problems Associated with Supernumerary Teeth
1. Failure of eruption of a maxillary central incisor. It may also cause retention of the primary incisor.
2. It may cause displacement of the permanent tooth. The degree of displacement may vary from a mild rotation to complete displacement.
3. Erupted supplemental teeth most often cause crowding in upper anterior teeth.
4. Dentigerous cyst formation may be associated with supernumerary teeth. Primosch reported an enlarged follicular sac in 30% of cases, but histological evidence of cyst formation was found in only 4 to 9% of cases.

DISCUSSION
A large percentage of anterior supernumerary teeth remain unerupted. It has been stated that only 25 percent of maxillary anterior supernumeraries erupt. Various theories exist for the different types of supernumerary. One theory suggests that the supernumerary tooth is created as a result of a dichotomy of the

Fig. 6: Showing IOPA of palatally erupted mesiodens

Fig. 7: Showing mesiodens with abnormal morphology

Fig. 8: Showing occlusal radiograph of maxilla with two fused mesiodens

Primosch reported an enlarged follicular sac in 30% of cases, but histological evidence of cyst formation was found in only 4 to 9% of cases.
A radiographic examination is indicated if abnormal clinical signs are found. An anterior occlusal or periapical radiograph is useful to show the incisor region in detail. The buccolingual position of unerupted supernumeraries can be determined using the parallaxes radiographic principle.

REFERENCES