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A Triad of Dental Anomalies: A Rare Case Report Involving the Mandibular Canines

Abstract: The aetiological factors related to many dental anomalies are still uncertain. Clinical descriptive terms may be useful but the distinction between different anomalies is often unclear. This paper describes a case of dental anomaly affecting the mandibular canines. These teeth exhibit a combination of hypoplasia and dilaceration. In addition, the left hand canine is fused with the adjacent lateral incisor. The possible aetiologies will be explored and we will discuss the management options in this unusual case.

Clinical Relevance: This case highlights the diverse nature of dental anomalies. They are often difficult to classify when severe malformation has occurred. It is not always possible to determine the aetiology of dental anomalies. It is important to recognize that the management of dental anomalies may require a multidisciplinary approach.

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Dental anomalies are caused by complex interactions between genetic, epigenetic and environmental factors that regulate the long and multi-faceted process of dental development. Over 300 genes have been identified as being involved.[1]

A disturbance to the process may result in abnormalities in:

- Tooth size (microdontia and macrodontia);
- Shape (fusion, gemination and concrescence);
- Number (anodontia, hypodontia and hyperdontia);
- Structure (amelogenesis imperfecta, dentinogenesis imperfecta and hypoplasia).

The distribution of anomalies is a good starting point when considering the aetiology. It can suggest whether the aetiology is genetic, systemic or local in nature.

The severity of the resulting malformation can result from a genetic defect with incomplete penetrance and variable expression or, in cases of local or systemic aetology, can be dependent on the severity of the insult itself. In addition, certain stages of tooth development are critical, leaving the tooth germ more vulnerable at specific times.

Hypoplasia

Enamel hypoplasia is defined as any reduction in the quantity of dental enamel and is the result of ameloblastic disruption during the secretory phase of enamel development. It can appear as a single pit or several pits, focal loss of enamel, or as horizontal grooves, also known as linear enamel hypoplasia. These may appear on the lingual/palatal or buccal tooth surface or the entire circumference. In an extreme case, it may lead to arrest of development of the permanent tooth germ.[2]

Enamel defects can be caused by the following:

- Trauma to the permanent tooth germ;
- Longstanding periapical infection of a primary tooth;
- Genetic disorders; or
- Systemic metabolic stresses.

In cases of genetic aetiology, all teeth in both primary and permanent dentition may be involved. In cases of systemic aetiology, all of the teeth developing during exposure to the aetiological factor may be affected. In cases of local aetiology, mainly the permanent incisors and premolars are involved.[3]

Approximately 100 aetiological

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agents have been reported to cause developmental defects of enamel and 70 genetic disorders are associated with enamel defects. Some of the most common genetic disorders that are associated with enamel defects are:

- Epidermolysis bullosa;
- Orofaciodigital syndrome;
- Ectodermal dysplasia;
- Prader-Willi syndrome; and
- Trichdento-osseous syndrome.

Systemic metabolic stresses include: ingestion of chemicals (fluorides, tetracyclines, dioxins, thalidomide); prematurity/low birth weight; severe malnutrition, neonatal hypocalcemia; vitamin D deficiency; bilirubinemia, thyroid and parathyroid disturbances; maternal diabetes; neonatal asphyxia; severe infections; and metabolic disorders.[4,5]

Data

- Prevalence ranges from 24% to 49% in the primary dentition and 9% to 63% in the permanent dentition.[6-12]
- Most commonly affected teeth are primary canines and second deciduous molars,[10] permanent incisors and mandibular canines.[11]
- There is no gender predilection.[13,14]

Dilaceration

Dilaceration is an abnormal angulation or bend in the root or, less frequently, the crown of a tooth. Any tooth may be affected.[15,16]

The exact aetiology of dilacerations is still controversial but the most accepted cause is mechanical trauma to the primary predecessor tooth.[17-20] However, this pathogenesis has been questioned.[21,22] Other possible contributing factors that have been proposed include:

- The ectopic development of the tooth germ;
- Presence of scar tissue;
- Infection;
- Cyst;
- Tumour;
- Developmental anomaly of tooth germ;
- Lack of space;
- Syndromes and hereditary factors.[23]

Data

- The prevalence ranges from 0.32%[24] to 98%.[25]
- The maxillary arch is affected more than the mandibular arch.[25]
- Permanent teeth > primary teeth.[26-28]
- Posterior teeth > anterior teeth.[27]
- Mandibular third molars are affected most often.[27]
- This may occur bilaterally in some patients.[29]
- There is no gender predilection.[24,25,30,31]

Fusion

Fusion is a tooth-shaped anomaly caused by the union of two or more dental germs, and can involve the permanent, primary or supernumerary dentition. Depending on the developmental stage at the time of union, fusion may be incomplete, involving only the tooth crowns, or complete, involving both the crowns and roots. Clinically, the fused tooth usually has a wide crown and two independent root canals or, less often, a single root and one or two pulp chambers.[32]

The aetiology and pathogenesis of fusion remains unclear, however, pressure or physical force producing close contact between two developing tooth buds has been reported as a possible cause. Trauma, genetic and environmental factors, such as foetal alcohol exposure, thalidomide embryopathy and hypervitaminosis A of the pregnant mother, have also been implicated as possible contributing factors.[16,33,34] Fused teeth may also form part of syndromes such as achondrodysplasia, chondroectodermal dysplasia, focal dermal hypoplasia and osteopetrosis.[35-39]

Data

- The prevalence ranges from 0.05% to 5% in permanent dentition and 0.7% in deciduous dentition.[40]
- The mandibular arch is affected more than the maxillary arch.[41]
- Anterior teeth > posterior teeth.[28]
- Incisors and canines are most frequently affected.[42,43]
- May be unilateral or bilateral.[42,44]
- There is no gender predilection.

Case report

An 18-year-old male presented to the Department of Orthodontics, University Dental Hospital of Manchester regarding malformed mandibular canines; the aesthetic impact was of some concern to the patient's mother but the patient expressed no concerns. Neither parent could recall any significant childhood illnesses.

Clinical examination (Figure 1) revealed an unrestored dentition, severely malformed mandibular canines, and what appeared to be a missing lower incisor. Plaque control was fair, but significantly worse around the malformed mandibular canines.

The patient exhibited bi-dentoalveolar proclination on a Class I skeletal base with a decreased overbite and lower centreline discrepancy.

The three lower incisors were well aligned with insufficient space available to accommodate a fourth incisor. There was 9 mm space for the mandibular canines bilaterally.

Radiographic investigation

OPG and periapical radiographs (Figure 2) of the lower labial segment were requested. Radiographs confirmed the bizarre crown morphology of the mandibular canines; a convoluted enamel layer and external surface, with a more normal root morphology and dilaceration of the apical thirds. It also revealed a probable full complement of teeth present as radiographic reporting advised that the lower left lateral incisor and canine were in fact fused.

On the basis of clinical examination and radiographic findings, a diagnosis of hypoplasia of the lower right canine, hypoplasia and fusion of the lower left

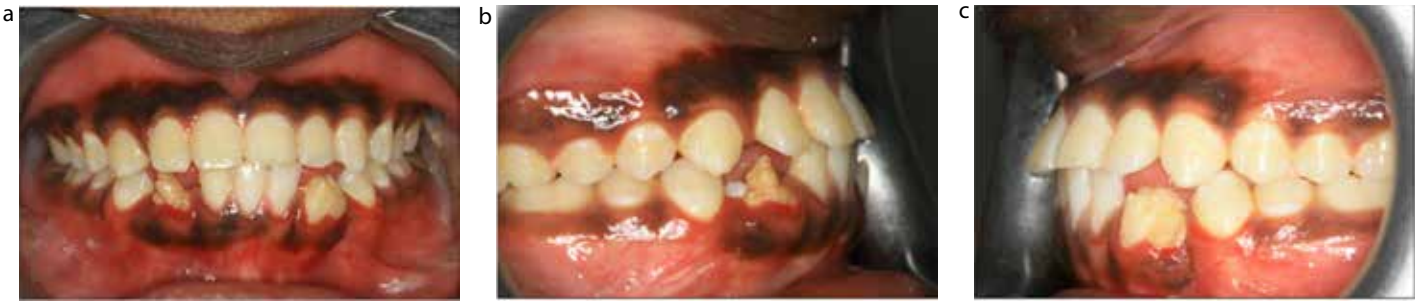

Figure 1. Clinical examination: **(a)** frontal view; **(b)** right and **(c)** left view.

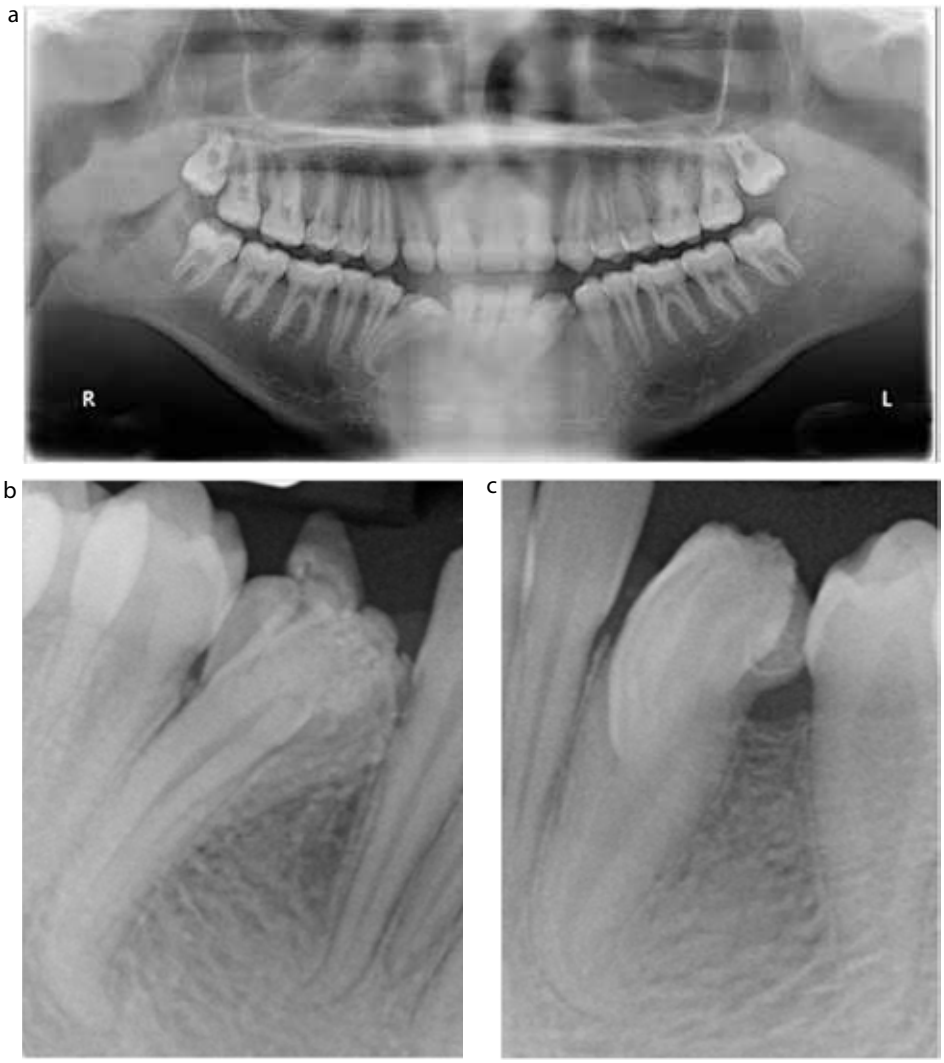

Figure 2. **(a)** Orthopantomogram and **(b, c)** periapical radiographs.

lateral incisor and canine was made, along with dilaceration of the apical third of the mandibular canine roots.

Discussion

Although the specific aetiology cannot be identified in this case, the malformed mandibular canines appear to have resulted from a systemic metabolic stress, rather than from genetic or traumatic causes. This is because of their localized and symmetrical distribution, in the absence of any local trauma or infection at the time of development.

Such anomalies are referred to as 'developmental' defects and indicate non-specific physiological stress during childhood. These influences on amelogenesis are related to the timing of development, the severity and duration of the insult and the host's susceptibility and response. Therefore, the same insult can result in a range of different responses in different individuals.

In addition to this, different teeth may favour different developmental reactions to stress, depending upon the degree of genetic control over their development. More developmentally stable teeth, such as lower canines and upper central incisors, exhibit more hypoplasia than surrounding teeth.[10] These teeth may be more susceptible to hypoplasia because their development is less easily disrupted. This may help explain why the certain tooth types that are rarely absent are also found to be the most frequently hypoplastic.[45,46,11] The insult to the mandibular canines in this case appears to be so severe that, in a less developmentally stable tooth, the same insult may have led to complete agenesis of the tooth germ.

In this particular case, a number of dental anomalies are seen together. There is known association between missing maxillary permanent canines and peg-shaped maxillary lateral incisors.[47,48] This is mirrored in an association between hypodontia and tooth size. The smaller teeth seen in an individual with hypodontia often also show morphological differences such as reduced form, tapering of the crown of microdont lateral incisors, as well as reduced cusp number and a more rounded occlusal perimeter in molars. In a controlled study of dental anomalies, significant reciprocal associations existed between aplasia of second premolars, small size of maxillary lateral incisors, infraocclusion of primary molars, enamel hypoplasia, and palatal displacement of maxillary canines.[49]

These associations could suggest a common genetic origin for these conditions or, in the case of adjacent dental anomalies, such as peg laterals and an absence of maxillary permanent canines, it might have resulted from an alteration of the local environment. Development of an adjacent unstable tooth germ may alter the local environment sufficiently to result in a reduction in size of the lateral incisors.[50] A similar hypothesis can be proposed in the present case.

Management

The following treatment options were discussed with the patient:

1. Accept the malocclusion and with no dental intervention.
2. Accept the malocclusion and attempt restorative camouflage of the mandibular canines. A restorative opinion was sought and it was deemed that reliable bonding with direct composite additions would prove difficult. Thus, extensive tooth preparation would be required to achieve a cosmetic result with the concomitant risks of eventual need for endodontic treatment in teeth with complicated root canal morphology.
3. Accept the malocclusion; extraction of both mandibular canines and subsequent restorative management of the residual spaces by means of bridges or implants. This option would be dependent on the patient improving and maintaining his dental health.
4. Extraction of both mandibular canines, along with a unit in both the upper right and left quadrants, followed by fixed orthodontic appliances, aimed at (partial) space closure to treat the bi-dentoalveolar proclination and correct the lower centreline (leaving a LR2 space to restore). This option would be dependent on the patient improving and maintaining his dental health.

The patient chose to accept the malocclusion and appearance of the mandibular canines, with the understanding that, should he change his mind in the future, both the restorative and orthodontic avenues

could be revisited.

Conclusion

The process of tooth development is complex, as is determining the aetiology of any disturbance affecting the process. These disturbances can lead to a vast array of dental anomalies ranging from small areas of hypoplasia to complete agenesis of the tooth germ.

When there are defects on concurrently forming teeth, this signifies a systemic metabolic stress as opposed to genetic or traumatic origin. The mandibular canines being affected so severely and in isolation may have resulted from the systemic stress occurring at a key stage of development; they are in fact one of the latest forming incremental structures completing calcification at around 6–7 years of age. No other teeth appear to be affected, however, microscopic analysis of the surrounding dentition may reveal local enamel hypoplasia defects on other teeth still forming at the time of insult that are not visible upon macroscopic observation.

Although the exact aetiologies of the described dental anomalies are not well understood, it would seem sensible to suggest that the observed association between hypoplasia, dilaceration and fusion may have resulted from an alteration in the local environment caused by the initial systemic metabolic stress.

The management of malocclusions complicated by such anomalies is challenging and benefits from multidisciplinary input. With severe developmental defects, CBCT may assist in showing in detail the crown morphology internally, however, this should only be requested if it is likely to influence the treatment plan.

Additionally, adult patients with capacity have every right to decline intervention for elective procedures despite their decisions being contrary to parental/family wishes or contrary to perceived social 'norms'. This may well involve frank discussions with the patient and accompanying family members.

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