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Kartagener's syndrome: unusual dental morphology

S. J. MERRETT & P. DURNING
Department of Orthodontics, Cardiff Dental Hospital, Cardiff, UK

Summary. The present paper describes the dental presentation of a female patient with Kartagener's syndrome. There are no reports in the literature identifying dental anomalies in patients with this condition. The subject presented with the congenital absence of an upper lateral incisor, enamel hypoplasia and aberrant tooth morphology.

Introduction

Kartagener's syndrome (KS) is described as a triad of situs inversus (mirror image organ placement), bronchiectasis and sinusitis [1–3]. It is included in the group of diseases caused by primary ciliary dyskinesia [4–8]. The syndrome was first mentioned in the literature in 1904 by Sievert, but it was in a series of papers published in the twentieth century that KS was defined [1–3]. Its incidence is reported at one in 15 000–20 000 Caucasians, and there is no known racial predilection [1–3]. The underlying factor in KS is a recessive gene defect inherited in an autosomal manner; however, the specific gene(s) conferring this condition has (have) not yet been identified [2,4].

Primary ciliary dyskinesia and associated disorders are characterized by the immotility or abnormal beating of cilia, which leads to insufficient mucociliary clearance, and defects of the cilia and spermatozoa [5,6]. Electron microscopic studies have shown an anomaly of the dynein arms which are present on the microtubules of the flagella of the cilia and sperm. Functional impairment of the ciliary apparatus involves the respiratory system, the alimentary tract and the reproductive system. Other disturbances coexisting with KS include disorders of the bone system, defects of the auditory organs, and defects of heart vessels [3].

Clinical diagnosis of KS is difficult since the symptoms are ill defined. Typically, rhinitis or nasal congestion begins on the first day of life; this remains constant over time and is difficult to treat. A persistent, moist-sounding cough within the neonatal period is unusual and suggestive; however, it is not conclusive proof of KS [7–9]. When diagnosis is late, the history of early respiratory problems is often unclear, but rhinitis, and/or nasal congestion and a moist-sounding cough will have been early and constant features. Chronic secretory otitis media is a very common problem amongst patients suffering with KS [6,9].

Diagnosis is usually achieved by studying mucociliary clearance, or ultrastructural examination of ciliary structures from samples obtained by bronchial and nasal brushing [4,5].

Radiological images are crucial in a diagnosis of KS; for example, X-rays of lateral sinuses, which typically demonstrate mucosal thickening, opacified sinus cavities and hypoplastic frontal sinuses. Chest radiographs and ultrasound of the abdominal cavity are also useful diagnostic tools [2,8,9].

There are numerous treatment methods for KS including airway clearance with daily courses of physiotherapy. Treatment modalities are aimed at preventing chronic infections of the respiratory system which can lead to severe respiratory problems in later life. Antibiotics are used to treat upper airway infections. Although prophylactic antibiotics should be used with great caution because of emerging antibiotic resistance, children with KS are especially good candidates for low-dose preventative antibiotics [9].

There is no evidence in the literature to suggest that people with KS have increased mortality. However, severe bronchiectasis is often a problem in patients who are diagnosed with KS later in life.

Correspondence: S. J. Merrett, Department of Orthodontics, Cardiff Dental Hospital, Heath Park, Cardiff, UK. E-mail: merrettsj@cardiff.ac.uk

The present case report illustrates dental anomalies found in a patient with KS. Some of the issues discussed can be applied to patients suffering from other respiratory problems, such as cystic fibrosis, since they also experience recurrent pyrexial episodes requiring antibiotics [10,11].

Case report

K. D., a 13-year-old female Caucasian, was referred by the community dental service for the opinion of a consultant orthodontist regarding her dentition. On presentation, the patient's complaint was 'pointed fang teeth' and irregular anterior teeth. The patient attended regularly and had previously received dental treatment under local anaesthetic. K. D. was diagnosed with KS at the age of 8 years, having suffered from recurrent chest infections and otitis media since birth. She had the classical triad of situs inversus, bronchiectasis and sinusitis. There was no family history of KS or dental abnormalities. The respiratory tract infections were treated with numerous courses of antibiotics. The antibiotics varied depending on sensitivity tests carried out at the time, and were administered either intravenously or as an oral suspension.

On clinical examination, K. D. was found to have a class 3 dental relationship on a mild skeletal 2 base, with an increased Frankfurt mandibular plane angle. Buccal segment relationships were class 1. K. D. had an anterior open bite with a posterior cross-bite in the premolar region.

All erupted canines were pointed in appearance with horizontal enamel grooves. The lower incisors also had an aberrant shape, with notching being particularly evident on the lower lateral incisors (Fig. 1). The upper central incisors had incisal notching with an area of cervical demineralization. The upper right lateral incisor was diminutive in size and the upper left lateral incisor was congenitally absent, with a retained upper left primary lateral incisor. Horizontal ridges were evident halfway up the clinical crown on all first permanent molars. The mild enamel hypoplasia exhibited on these teeth resembled linear enamel hypoplasia (LEH). There was no history of trauma and no record of abnormalities was found in the primary dentition.

An orthopantomogram showed the patient to be in the mixed dentition with developing second premolars, upper second molars and a lower right third permanent molar. No other teeth were congenitally missing and no other abnormalities were found.

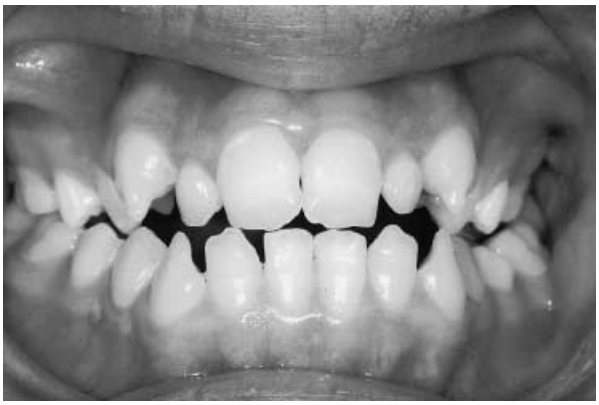

Fig. 1. Photograph illustrating the aberrant tooth morphology, notching and grooves on the patient's anterior teeth.

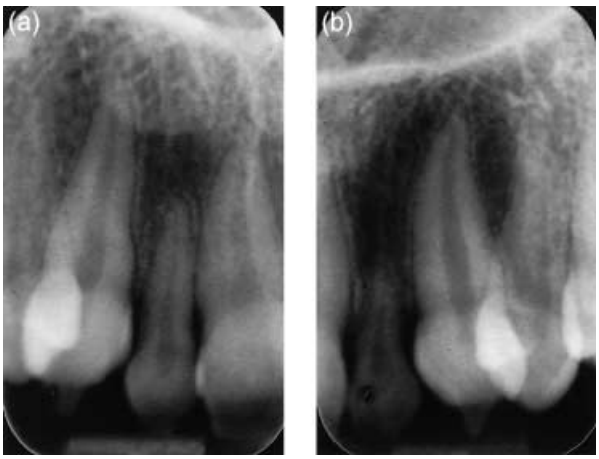

Fig. 2. Periapical radiographs showing a diminutive root on the upper right lateral incisor with a retained upper left primary lateral incisor.

Caries were found in the lower first permanent molars and both upper second primary molars. Periapical radiographs showed a diminutive root on the upper right lateral incisor with a retained upper left primary lateral incisor (Fig. 2). There was poor bone support remaining around the retained primary lateral incisor with 60% bone levels around the upper right permanent incisor.

The patient was referred back to the community dental service for restorations in the lower molars coupled with preventative advice. An appointment was arranged for a joint orthodontic and restorative opinion at the University of Wales Dental School, Cardiff, UK.

At this appointment, it was decided that the upper left primary lateral incisor had a limited prognosis because of the diminutive root and bone support. K. D. was willing to accept the loss of the primary lateral incisor; however, she was keen to retain the upper permanent lateral incisor, with the advice that it could be lost in the future.

Because of her previous high caries incidence, K. D. received a course of oral hygiene instruction with an orthodontic hygienist prior to commencing treatment. As a result of the cervical demineralization exhibited on the anterior teeth, it was decided to keep treatment to a minimum and only address her presenting complaint. Prior to commencing fixed orthodontic treatment, the retained primary incisor would be extracted and simple fixed upper and lower appliances used to align the anterior segments. The lateral spaces would be increased to allow provision of a resin-retained bridge to replace the upper left lateral incisor and also to allow a composite build up on the upper right lateral incisor. Following alignment of the anterior segments, correction of the posterior cross-bite would only be addressed if requested by K. D.

The notching, grooves and aberrant shape of the incisors and canines would be modified using composite techniques. Both the patient and her parents were happy with the proposed treatment plan.

Discussion

To date, there are no reports in the literature of an increased incidence of LEH, aberrant tooth shape or an increased risk of congenital absence of teeth in patients with KS. It is difficult to draw conclusions on the basis of one case report. However, there has been mention of disorders of the bone system associated with this condition, which could implicate disorders of the dentition. Developing tooth germs are contained within the bone architecture, and any alteration in the bone system can affect the developing dentition [3].

The high incidence of caries could be explained by the risk of infections experienced by the individual. Frequent infections imply a risk for dental caries since, when the child is suffering from otitis media or pneumonias, there is an increased risk of cariogenic dietary habits, with frequent sweet drinks, meals and snacks [9–11]. The patient also received antibiotics as an oral suspension, and if this was not in the form of sugar-free medication, then this could be a risk factor for caries. Furthermore, infections may affect salivary function and any decrease in salivary flow often leads to an increased caries rate. It is also likely that oral hygiene procedures are not carried out as effectively when a child is ill.

One of the most unspecific indicators of stress is LEHs [12,13]. These are defined as horizontal lines or grooves of decreased enamel thickness on the external surface of the tooth crown. They probably result from disturbance of the enamel formation during tooth crown development. Such disturbances can be caused by factors which affect the child's growth; for example, inherited and infectious disease, pyrexia, changes in dietary habits, and malnutrition [12,13]. These factors are believed to interrupt enamel formation by affecting ameloblast activity. After recovering, formation of the tooth crown continues, leaving a line of thinner enamel and ridges. The hypoplasia illustrated in the present case could be explained by childhood illness (with associated pyrexia), hospitalization (which is often stressful) and recurrent courses of antibiotics.

It is known that enamel defects such as hypoplasias or hypomineralization with disintegrated enamel retain dental plaque, increasing the risk of dental caries. Therefore, if the enamel of the primary dentition in the present case was hypoplastic, then this could explain the high caries incidence in the primary molar teeth of this individual [9].

The literature shows that there is evidence that mouth breathing and an anterior open bite have been associated with chronic nasal and sinus obstruction. This has been used to explain the high incidence of anterior open bite in patients suffering from cystic fibrosis [10,11]. Cystic fibrosis has a similar respiratory presentation to KS, and hence, the presence of anterior open bite in the present case report could be explained by respiratory problems suffered by the individual.

The treatment plan that was agreed included the upper right lateral incisor in the full-mouth fixed appliance therapy. Recent research has shown that patients with peg-shaped and small lateral incisors are not at higher risk of root resorption [14]. Furthermore, the patient had not received any trauma to her anterior teeth, had no lip or tongue dysfunction, and did not have any impacted canines; this also placed her in a lower risk category for root resorption [14,15].

The above case report illustrates the deficiencies in the current literature and research with regard to the reporting of disorders of the dentition in patients

with KS. Future research could be channelled into studying the dentition of children and adults affected with KS. Should further investigations reveal hypodontia, LEH and aberrant tooth shape in KS patients, then this would indicate that KS sufferers should have their developing dentition closely monitored. Radiographic examination should be undertaken at an appropriate age to exclude hypodontia and preventative advice reinforced at subsequent appointments.

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Résumé. Cet article décrit l'aspect dentaire d'une patiente avec un syndrome de Kartageners. Il n'existe aucun rapport dans la littérature identifiant les anomalies dentaires chez les patients atteints de ce syndrome. La patiente présentait une absence congénitale d'une incisive latérale supérieure, une hypoplasie de l'émail et une morphologie dentaire aberrante.

Zusammenfassung. Dieser Artikel beschreibt Zahnärztliche Befunde einer Patientin mit Kartagener Syndrom. In der Literatur sind bislang keine Fälle dieses Syndroms mit Zahnveränderungen beschrieben. Bei der hier beschriebenen Patientin zeigten sich das Fehlen eines seitlichen Oberkiefer Schneidezahns, Schmelzhypoplasie und Zahnformveränderungen.

Resumen. Este artículo describe las anomalías dentales de una paciente con síndrome de Kartageners. No hay informes en la literatura que identifiquen las anomalías dentarias en pacientes con este síndrome. La paciente presentaba ausencia congénita de un incisivo lateral superior, hipoplasia de esmalte y morfología dentaria aberrante.

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