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Meningococcal septicaemia and dental complications: a literature review and two case reports

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Summary. The present report describes two cases of dental complications of meningococcemia at an early age. The meningococcal infection in these young children resulted in limb amputation and significant plastic surgery. Dental treatment and psychological considerations are described in both cases. Similar dental complications, especially in the premaxilla, were found. Some of the severely affected teeth were extracted.

Introduction

Neisseriae meningitidis is an anaerobic gram-negative cocci that is a frequent habitant of the human nasopharynx. It can bring about disease when it penetrates the tissue, enters the bloodstream and subsequently disseminates to other organ systems [1,2]. The clinical manifestations of the disease are associated with an acute inflammatory response and include: upper respiratory infection resembling the common cold; acute meningicoccemia with fever; malaise; myalgia and arthralgia deteriorating to petechial lesions; disseminated intravascular coagulation; hypotension and renal failure; meningitis; coma; and even death. This condition may also result in necrosis and infarction of organs such as the adrenal glands, kidneys and bone as a result of small vessel occlusion by microthrombi of fibrin and leukocytes. High morbidity and mortality rates are reported as a result of acute meningicoccemia [3,4]. The usual age for meningococal infection in children is the period from 3 months to 5 years [1–3]. At this age, there is rapid development of both jaws and the permanent teeth. Dental complications have been reported in a 2-year-old child following meningococcal infection involving diffuse vasculitis and disseminated intravascular coagulation (DIC) [4]. However, the dental complications were only revealed 8 years later, when the permanent central incisors failed to erupt. The dental abnormalities described included: tooth agenesis; denticles; impaction; delayed eruption and development; fusion of tooth germs; and hypoplasia of teeth in the premaxilla [4].

Viral and bacterial infections occurring in early childhood can disturb dental and orofacial development [5,6]. Serious childhood illnesses increase the prevalence of dental defects [7]. Children are more susceptible to developmental defects as a result of systemic infection than adults since they are in the critical stages of their development and maturation [5]. Disturbance at any time during the appositional development of the crown may arrest the activity of the group of functional ameloblasts, causing the enamel rods to develop less extensively than normal, and will be expressed as enamel hypoplasia. However, it is also well recognized that some systemic conditions may affect the enamel and dentin-forming cells either before or after the appositional phase of their development [8]. The same factors which cause enamel hypoplasia can also cause more significant crown defects (e.g. either enamel hypocalcification or hypomineralization), depending on the stage of tooth development, i.e. matrix formation, calcification or maturation [9]. It is important to note that lesions with similar appearances do not necessarily have similar causes, and conversely, the same aetiological

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factor can produce dissimilar lesions when active at different stages of tooth development [9].

The aims of the present paper are to review two cases involving dental abnormalities as a late consequence of a severe meningococcal infection in young children, and to discuss the possible dental and psychological complications which may be relevant to the paediatric dentist.

Case reports

Case 1

A.R., an 8·5-year-old girl, was referred to the Department of Pediatric Dentistry of the Hadassah School of Dental Medicine, Jerusalem, Israel. Her main complaints were about several short episodes of dentinal pain and the aesthetic displeasing appearance of her upper incisors.

Her medical history revealed that, at the age of 2 years, A.R. had experienced an episode of meningococcal infection with high fever (40 °C) and purpura fulminans, followed by complications of DIC which caused diffuse necrosis of large areas of her skin, especially over her limbs, fingers and face (i.e. her nose and upper lip), and a temporary dysfunction of her liver and kidneys. The immediate treatment included intubations and intravenous antibiotics, steroids and blood replacement (i.e. fresh frozen plasma, cryoprecipitate, platelets and concentrated factor VIII). A.R. later underwent amputation of both legs above the knees and the distal phalanges of her fingers. Since her recovery, the subject had undergone several operations to correct facial scarring.

Extraoral examination revealed scars over her nose and upper lip (Fig. 1).

Intraoral examination revealed a limited mouth opening of only 25 mm anteriorly and scars on the inner side of A.R.'s upper labial mucosa. The upper and lower permanent incisors (teeth 11, 21, 31, 41, 12, 22, 32 and 42) were hypoplastic, with yellow-brown internal discoloration of the enamel. The upper lateral incisors (teeth 12 and 22) were ectopically erupting, with the right one emerging labially and the left erupting lingually. All the permanent central incisors and first molars were decayed and hypoplastic. The remaining primary molars were near exfoliation. Severe anterior crowding and an underdeveloped premaxilla were also noted, along with poor oral hygiene and associated chronic marginal gingivitis (Figs 2 & 3).

Fig. 1. Facial view of A.R. on presentation. Note the scars.

Fig. 2. Intraoral frontal view of A.R.'s hypoplastic incisors. Note the anterior crowding.

Radiographic examination revealed that the roots of the upper lateral incisors were partially developed and the roots of the upper central incisors were short. Circular hypoplasia was evident in all the lower incisors. The unerupted upper permanent cuspids also appeared hypoplastic (Fig. 4). A radiolucent area in

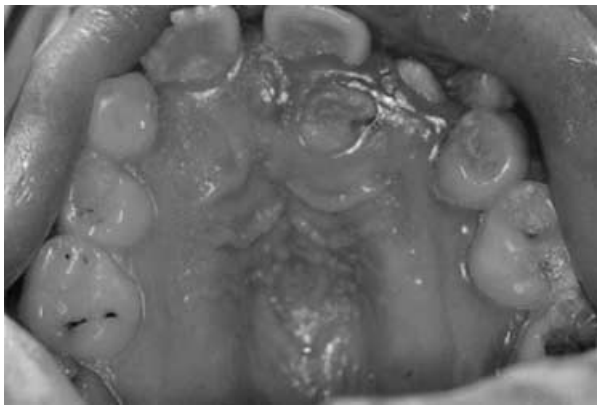

Fig. 3. Intraoral occlusal view of A.R.'s upper teeth. Note the hypoplastic lateral incisior and the crowding.

the coronal dentin of the lower left first premolar was diagnosed as a pre-eruptive intracoronal resorptive lesion, an example of the so-called 'occult caries' (Fig. 5).

A detailed treatment plan was proposed to the family and the definitive treatment was carried out under general anaesthesia. This included extraction of both crowded underdeveloped hypoplastic upper permanent lateral incisors. The primary cuspids were removed in order to provide space for the eruption of the permanent canines. The upper central incisors were restored with composite resin. The posterior teeth were restored either by amalgam (for large restorations) or with preventive resin restorations (for smaller lesions). A.R. and her parents were given oral hygiene instructions which included toothbrushing with fluoridated toothpaste and rinsing with fluoride-containing mouthwash.

Following treatment, the family was very pleased with their daughter's improvement in her dental function and aesthetics, socialization with her peers, and her self-esteem and body image. The compliance with the comprehensive preventive programme was excellent. At the initial review, it was evident

Fig. 4. Periapical radiograph of A.R.'s unerrupted upper canines revealing hypoplasia: (a) tooth 13 and (b) tooth 23. The hypoplastic incisors are also discernable.

Fig. 5. Panoramic view of A.R. at 8·5 years of age. Note the suspected lesion of pre-eruptive intracoronal resorption in the first left premolar (tooth 34).

Fig. 6. Intraoral view of A.R.'s newly erupted hypoplastic upper permanent canines. Note the composite restoration of the central incisors.

that the extraction of both hypoplastic upper lateral incisors had enabled the initial eruption of the permanent cuspids into the extraction space. A follow-up examination 2 years later revealed the full extent of the coronal chronological hypoplasia of the permanent upper cuspids and first premolars (Fig. 6).

Case 2

D.W., a 2·5-year-old girl, was referred for dental examination by her paediatrician to the Department of Dentistry of the Children's Hospital at Westmead, Sydney, New South Wales, Australia.

Her medical history revealed that, at the age of 14 months, she had experienced severe meningococcal infection with purpura fulminans, electrolyte disturbance, critical ischaemia involving all four limbs, coagulopathy, tachycardia and nephropathy. D.W. required multiple procedures: the gangrenous tissue was debrided; she underwent plastic surgery, including skin grafting; her left arm was amputated below the elbow; and all the digits on her right hand (with the exception of her thumb stump) and the tips of most of her toes, except for the left big toe, were also amputated. As a result of her illness and subsequent treatment, D.W. suffered from leg length discrepancy and significant body scarring. Her general level of intellectual functioning was within the normal range.

At her first dental visit 15 months after her illness, a delay in the completion of the eruption of her primary incisors was noted. The upper left central incisor (tooth 61) was mobile, and soon after, it was lost prematurely because of a lack of periodontal support. Over the next 3 years, the left lateral incisor (tooth 62) and cuspid (tooth 63) also exfoliated prematurely. The upper right central incisor (tooth 51) also failed to erupt fully and was removed whilst the patient was under general anaesthesia for her left big toe/right thumb graft at 7 years of age. At this initial stage, D.W. and her mother were given a general outline of the dental issues which would confront the subject in the future as well as the lifelong importance of following a thorough preventive regime.

In follow-up appointments when she was 8 years old, D.W.'s upper permanent molars were sealed, and initial treatment planning was undertaken for her hypoplastic upper left central incisor (tooth 21) and hypoplastic/hypomineralized lower right permanent central incisor (tooth 41). The upper right primary right cuspid (tooth 63) and upper left second primary molar (tooth 65) were also hypoplastic. Gingival inflammation and recession around the upper right primary lateral incisor and cuspid were observed (Fig. 7).

Radiographic examination revealed that the tooth germs of the permanent upper lateral incisors and

Fig. 7. Intraoral view of D.W.'s hypoplasic left upper central incisor (tooth 21). Note the absence of the right upper central incisor (tooth 11).

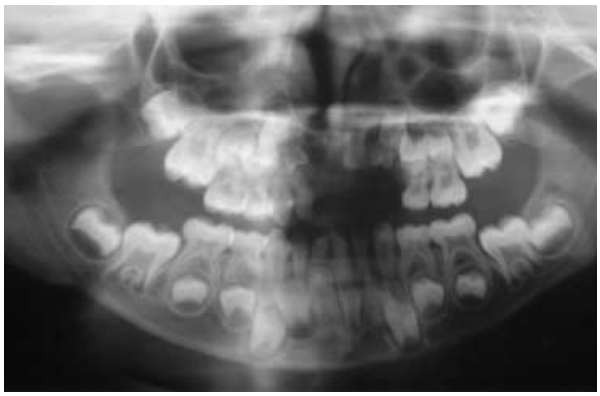

Fig. 8. Panoramic view of D.W. at 8 years of age.

Fig. 9. Periapical radiograph of D.W.'s maxillary incisors revealing hypoplasia of the left upper central incisor (tooth 21) and aplasia of the right upper central incisor (tooth 11).

cuspids were underdeveloped and hypoplastic, and there was agenesis of the upper right permanent central incisor (Figs 8 & 9).

Recent restorative treatment for D.W. has included composite resin restorations of the facial surfaces of teeth 21, 53, 41, 31 and 32. Her frequent hospitalization, most recently for leg-lengthening procedures, has resulted in a situation where she finds treatment difficult and invasive dental procedures are a real challenge. Consequently, D.W.'s work has been carried out using oral sedation (midazolam 0·3 mg kg^{-1} up to a maximum dose of 10 mg). The hypoplastic anterior teeth were, not surprisingly, extremely sensitive, and the use of local anaesthesia was greatly facilitated by the use of oral sedation.

D.W. has recently had an upper acrylic partial denture fitted to replace her missing upper anterior teeth. She is very pleased with the results, and has proved adept at keeping her mouth and denture clean. The subject is under strict instructions to always leave her dentures out overnight.

At her next review, D.W.'s orthodontic needs will be of prime concern, especially given the apparent hypoplasia of her developing, unerupted, upper anterior dentition.

Discussion

The possible consequences of severe infection by *Neisseriae meningitidis* in early childhood, as described in the present cases and the one reported by Walton *et al.* [4], may be divided into three areas: dental, orofacial and general.

The possible dental defects may include: tooth agenesis; morphologic aberration; denticles; tooth impaction; delayed eruption and development of teeth; premature exfoliation of primary teeth; fusion of teeth germs; hypoplasia; hypocalcification; caries; and malocclusion.

The orofacial complications may consist of: restricted mouth opening; scarring; bony deficiency; and poor aesthetics.

The general complications may include: psychological and behaviour management problems as a result of repeated episodes of surgery and experience of painful procedures; mental and motor developmental delay; crippling disabilities; and a financial and psychological burden on the family [10].

Some similarities between these two case reports, and that of Walton *et al.* [4], may be noted:

1 All three cases involved children who were 14–24 months old.

2 All three cases presented with coagulopathy and bone involvement.

3 The teeth in the area of the premaxilla suffered most from severe hypoplasia and morphological abnormalities. There was also hypocalcification and

mild hypoplasia of the lower permanent incisors, upper canines and premolars in one case.

In addition to its systemic damage, DIC may have also contributed to local metabolic deficiencies in the ameloblasts. It was most likely that both the systemic and local factors contributed to the dental effects.

The reported underdevelopment of the premaxilla in the first case (A.R.) and in the case described by Walton *et al*. [4] may be a result of bone infarction and necrosis during the acute phase of the disease. In addition, the scarring of the upper lip puts added pressure on the upper teeth, thus contributing to the severe crowding of the anterior teeth.

Walton *et al*. [4] suggested that the mechanism of the dental defects is osteomyelitis. Even though it may be possible that the alveolar bone had subclinical osteomyelitis, the present authors can still find no explanation for the hypersensitivity of the premaxillary region as compared to the posterior teeth in the mandible. It seems likely that tooth germs in the posterior mandibular area would be more prone to osteomyelitis because of diminished mandibular vascularity [11].

Coagulopathy and DIC may cause infarction, which is transient in nature and does not necessarily involve osteomyelitis. Nevertheless, impairment of the vascular supply around the teeth germs may result in the death of the osteoblasts and odontogenic cells. It may be that the premaxilla is more susceptible to occlusion of the small vessels, and may be prone to infarction and necrosis during DIC, as are other body extremities (e.g. the fingers and the tip of the nose). The posterior mandibular bone and tooth germs appeared to be less sensitive to impaired vascular supply in all three cases.

Children and parents often realize the existence of dental complications at 7 or 8 years of age when the permanent teeth fail to erupt or erupt with an unaesthetic appearance. Facial aesthetics at this age are essential [12,13]. A smile is very important for socialization (i.e. as a first impression and for attractiveness), for the child's self-esteem, and for establishing a positive basic reality [13–15]. Improved facial appearance may facilitate the integration of a child with a disability into society [16].

Aesthetic treatment, along with orthodontic counselling, has contributed to the restitution of these children's self-confidence and body image. This, along with improved prostheses for the amputated limbs, had a crucial effect on their motivation and compliance with oral hygiene instructions.

Mental and psychological difficulties may be more common in these children [10]. They may suffer from intellectual and motor developmental delay, severe dental anxiety, objective physical difficulties even with simple tasks like toothbrushing, and a high caries rate because of their hypoplastic teeth [17]. Thus, an individually tailored dental care plan is essential so that all these special factors are taken into consideration. An orthodontic care plan may involve some lateral thinking to best manage the unique situations which one is confronted with, such as the extraction of the upper permanent lateral incisors, as described in the first case (A.R.) [18].

The preferred dental management in some of these cases involved treatment under general anaesthesia. In the first case (A.R.), it was mainly because of severe dental anxiety, the complexity of the dental treatment, the restricted mouth opening and the hypersensitivity of the girl's lips to touch, and her wish to restore the aesthetics as soon as possible. In the second case (D.W.), the more demanding treatment of extracting the impacted primary incisor was performed whilst the child was under general anaesthesia for another surgical procedure.

Conclusions

The two cases described in this present report illustrate some of the severe dental complications which can occur subsequent to meningococcal infection. The paediatric dentist should be aware of the developmental disturbances which may occur following such a devastating infection and be prepared to perform the appropriate dental treatment as part of an overall healthcare plan for these very special children.

Résumé. Cet article présente deux cas de complications dentaires de meningococcémie à un âge précoce. L'infection à méningocoque chez ces jeunes enfants a entraîné l'amputation d'un membre et plastie chirurgicale importante. Le traitement dentaire et les considérations psychologiques sont décrits dans les deux cas. Des complications dentaires similaires, spécialement dans le prémaxillaire ont été trouvées. Certaines des dents sévèrement atteintes ont été extraites.

Zusammenfassung. Die vorliegende Arbeit stellt zwei Fälle von zahnärztlichen Komplikationen einer früheren Meningokokkensepsis vor. Die

Meningokokkeninfektion der jungen Kinder war von Amputationen im Gliedmaßenbereich und erheblichen nachfolgenden plastisch chirurgischen Eingriffen gefolgt. Zahnbehandflung und psychologische Überlegungen in beiden Fällen werden beschrieben. Es wurden ähnliche zahnärztliche Komplikationen gefunden, vor allem in der Prämaxillaregion. Einige schwer betroffene Zähne wurden extrahiert.

Resumen. Este informe describe dos casos de complicaciones dentarias de una meningococemia en una edad precoz. La infección meningocócica en estos niños pequeños produjo amputación de una extremidad y la realización significativa de cirugía plástica. En ambos casos se describe el tratamiento dental y las consideraciones psicológicas. Se encontraron complicaciones dentarias similares, especialmente en la premaxila. Se extrajeron algunos de los dientes severamente afectados.

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