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Enamel hypoplasia of the primary dentition in a 4-year-old with intestinal lymphangiectasia

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Summary. Intestinal lymphangiectasia (IL) is a rare disorder, and its incidence and prevalence is unknown for either Australia or world-wide. It is characterized by diarrhoea, mild steatorrhoea, oedema, enteric loss of protein (protein-losing enteropathy) and abnormal dilated lymphatic channels in the small intestine. Whilst oedema and diarrhoea are the predominant clinical features, other observed features include hypoalbuminemia, hypogammaglobulinemia, trace metal deficiency, hypocalcemia and chylous pleural effusions. While medical presentation of the condition has been reported widely, few descriptions of oral findings have been published. A search of Medline found two reports of dental findings in the permanent dentition in patients with IL. To date, there have been no reports on dental findings in the primary dentition. The primary dentition of a 4-year-old boy with IL had teeth with enamel defects which reflected the timing of enamel development and the period in which the disease was active. The present report highlights the need for early involvement of the dental team in the dental management of children with IL.

Introduction

Intestinal lymphangiectasia (IL) is a relatively rare disorder that manifests itself through intestinal malabsorption. Its incidence and prevalence in Australia and world-wide is unknown. The disease affects males and females equally, and there is no racial predilection. The disease can be primary (congenital), where there is malformation within the lymphatic channels leading to their blockage, and the condition is usually diagnosed within the first decade of life. The condition can also be secondary to other disease states, such as constrictive pericarditis, lymphoma, sarcoidosis and scleroderma, and can affect older adults. The first presentation is usually persistent diarrhoea and peripheral oedema. The oedema can be unilateral or bilateral, and macular oedema on funduscopic examination has been reported (blindness caused by this is reversible). There is frequently steatorrhoea, lymphocytopenia, hypogammaglobulinemia, hypoproteinaemia and malabsorption [3,4]. Patients can also develop hypocalcemia secondary to failure to absorb fats and fat-soluble vitamins. Because of lymphocytopenia, hypoproteinaemia and hypocalcemia, oral findings of gingivitis and defects of the enamel in the permanent dentition have been reported [1,2].

Ultrasound and computed tomography scans may identify dilated intestinal loops, regular and diffuse thickening of the intestinal walls and folds, and mesenteric oedema. Jejunal biopsy usually establishes a definitive diagnosis, and displays dilation of mucosal and submucosal lymphatic channels without any evidence of inflammation. The hypoplastic lymphatics result in an obstruction in lymph flow, leading to increased pressure within the lymphatics that causes dilation of the lymphatic channels in the intestine, and finally, rupture of the channels and resultant discharge of the lymph into the lumen of the bowel [3,4].

Management of the condition involves dietary changes, pharmaceutical therapy and behavioural modifications. Dietary modification is primarily directed at replacing long-chain fatty acids with

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medium- and short-chain fatty acids because, unlike the long-chain fatty acids, the medium- and short-chain fatty acids are more easily absorbed through the portal venous channels. Medications used include over-the-counter drugs to bulk up the stool and control diarrhoea. In more severe cases, the use of octreotide, a synthetic analogue of a naturally occurring hormone somatostatin (a potent inhibitor of the release of growth hormone, serotonin, gastrin, glucagon and insulin) has been found to be successful [5,6]. Surgical management is rarely indicated for patients with primary IL, but may be used for conditions predisposing to IL.

There are just two reports of the dental manifestations of IL cited in the Medline database [1,2]. The report by Dummer and Cardiff [2] described dental findings in a 13-year-old boy. The child displayed poor oral hygiene with abundant plaque and calculus, and marked pitting and staining of all the permanent incisors and canines. The first permanent molars were reported as absent and the primary teeth were reported as having been normal. The report by Ralph and Troutman [1] described oral findings in a 14-year-old boy. The child had poor oral hygiene and severe marginal gingivitis, and all the permanent teeth had pitting hypoplasia and brown discoloration. There have been no reports of findings in the primary dentition of patients with IL. The present case report describes oral findings in the primary dentition of a 4-year-old boy with IL.

Case report

A boy aged 4 years and 9 months attended for routine examination at a school dental therapy clinic with his mother. The parent had completed a medical information questionnaire that indicated that the child had IL that had been diagnosed soon after birth. Medical information was sought from the consulting paediatric gastroenterologist. The child was born through Caesarean section and weighed 4·08 kg at birth, but he failed to thrive and was 3·18 kg within a few weeks. The boy displayed signs of malabsorption soon after birth, with frequent malodorous stools with fatty deposits within them. The child was diagnosed with IL through biopsy and placed on a modified diet (Portagen®) from 2 months of age until he was 12 months old. The blood assay demonstrated reduced levels of immunoglobulins, but the boy did not experience any episodes of major infections. He currently enjoys a fairly normal diet with no major restrictions, and can consume full fat milk and yoghurt. The child's current height for weight is appropriate for his age, being at the seventy-fifth percentile for weight and the fiftieth percentile for height [7]. A review assessment by the gastroenterologist that included a blood test within a week of the dental examination showed that all his biological parameters were within normal limits.

Oral findings

The patient had reasonably good oral hygiene, with minimal debris and plaque accumulation. The gingival tissues were healthy, with no sign of overt gingivitis. All primary teeth were present. Some teeth were noted to have carious lesions (lower left first and second primary molars) and some had already been restored (both the upper second primary molars), and on closer examination, it was found that all the second primary molars had enamel defects and it was likely that caries was a secondary occurrence. The teeth affected by enamel defects were all the second primary molars (55, 65, 75, 85) and the primary canines (53, 63, 73, 83), and the extent of the defect ranged from a well-defined demarcated opacity on the lower primary canines to an absence of enamel on the incisal edge of the upper primary canines. The lesions present on the first primary molars did not have an appearance of enamel defects, but this could not be definitely excluded, while the lesions on the second primary molars showed demarcated opacities associated with enamel defects as well as the carious lesion. Figure 1 shows the enamel defects on the right upper and lower primary canines, and Fig. 2 shows the enamel defects on the upper and lower left primary canines. Figure 3 shows caries and enamel defects on lower left first and second primary molars. Bitewing radiographs also show clearly the enamel defects present on the canines and the molars (Figs 4 & 5).

Discussion

The present paper describes dental findings in a 4-year-old boy with IL. The location of the enamel defects (i.e. sites and teeth) were suggestive of the episode occurring when medical complications caused by IL were active, i.e. from birth until 8 weeks of age. The chronology of dental development suggests

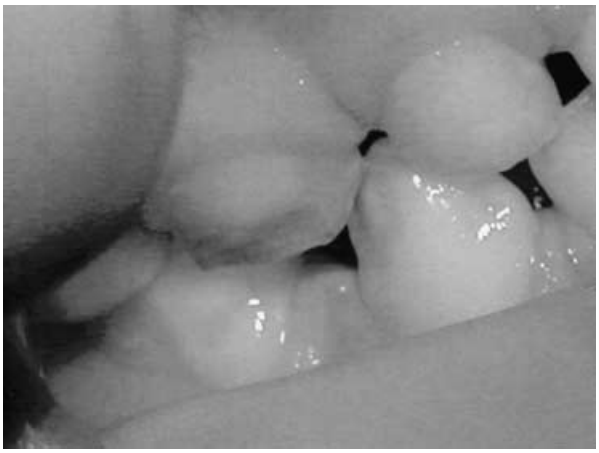

Fig. 1. Defects on 53 and 83 (c / c).

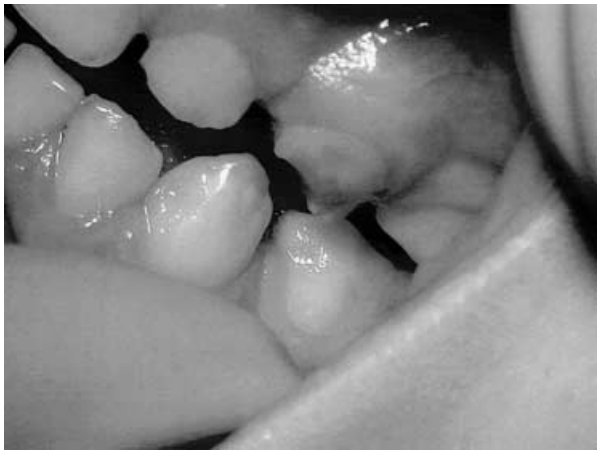

Fig. 2. Defects on 63 and 73 (c / c).

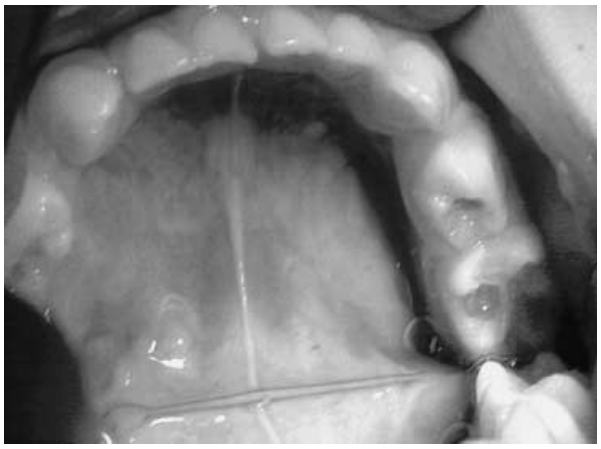

Fig. 3. The mandibular left first and second primary molars, (DE) (mirror image).

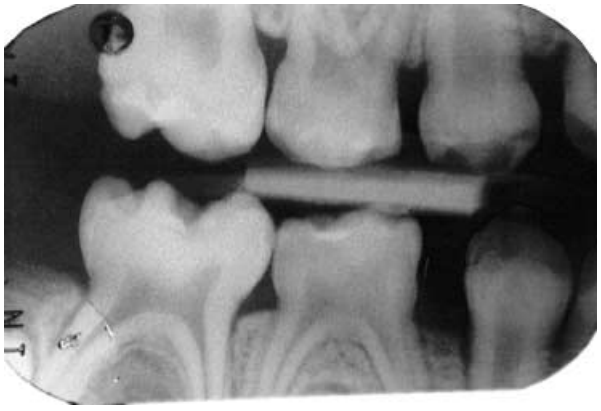

Fig. 4. Right-hand-side bitewing radiograph.

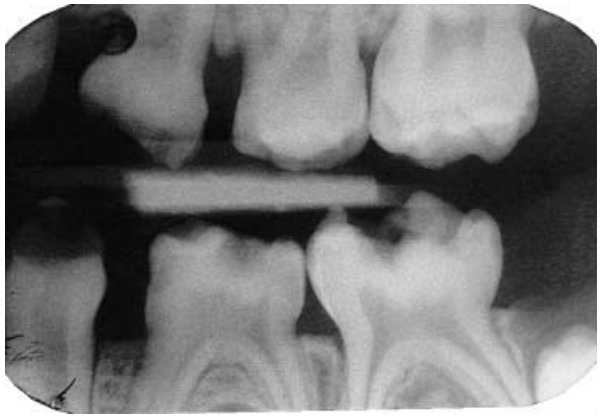

Fig. 5. Left-hand-side bitewing radiograph.

that the time of occurrence of the illness and expected stage of enamel calcification closely reflected the defects observed. At birth, about one-third of the enamel is mineralized in the primary canines and the cusps of the second primary molars would be mineralized, but the cusps would still be separate [8]. In the present subject, the incisal third of the maxillary canines was more severely affected than that of the mandibular canines, and the lesions in the molars were confined to the grooves and fissures. A careful assessment did not reveal any familial history of amelogenesis imperfecta or excessive exposure to fluoride. No other primary teeth were affected. No other radiographs were taken apart from the bitewing radiographs, and it will be of interest to observe the enamel status of the permanent dentition, especially the first permanent molars, when they erupt. The report by Dummer and Cardiff [2] noted the absence of first permanent molars, but did not explain the reason for their absence, i.e. whether the teeth were congenitally absent or had been extracted because of caries, maybe secondary to severe hypoplastic defects.

The mechanism through which intestinal lymphangiectasia causes enamel defects is not clear. It has been postulated that systemic factors which cause hypocalcemia can interfere with tooth calcification [9]. Enamel defects have been observed in individuals with other malabsorption diseases such as coeliac disease [10]. An association between coeliac disease and intestinal lymphangiectasia have been reported, and the effects of these conditions may be similar; both conditions can lead to hypocalcemia [11]. Although, in this instance, the consulting gastroenterologist reported that there did not appear to have been hypocalcemia, the child was not absorbing vitamin D. Deficiency in vitamin D and various disturbances associated with vitamin D metabolism have been implicated in defects of enamel formation [9,12].

The present case report illustrates that dental defects may arise in the primary dentition of a child with intestinal lymphangiectasia, especially if the disease was active during periods of enamel mineralization. The defects ranged from demarcated opacities to a frank absence of enamel, although pitting hypoplasia (as has been reported for the permanent dentition) was not observed [1]. The parent of this child, through concern and perhaps lack of information, treated the child to sugary foods as compensation for the child's inability to consume a normal diet. The frequent availability of sugar in the diet probably compounded the problem of enamel defects and led to the occurrence of caries on tooth surfaces with enamel defects on sites where oral hygiene may not have been as good as on the more accessible surfaces of the primary canines. The carious lesions were restored temporarily with a glass-ionomer restorative, while more definitive stainless steel crowns will be considered for the future. The noncarious enamel defects have been left untreated at this stage. The parent was involved in the entire management process, and was fully informed of the dental findings and the care required to maintain the remaining primary teeth in a healthy condition. The child has been placed on a regular review to monitor the developing dentition.

The two published reports on dental findings associated with intestinal lymphangiectasia highlighted the defects present in the permanent dentition [1,2]. The present report shows that defects can also occur in the primary dentition. The author has highlighted the need for dental considerations in cases of malabsorption among children, and in particular, the early involvement of a dental professional in the overall care of the patient, not just for restorative care, but in the dietary counselling of parents and to assist in establishing appropriate preventive care for the developing dentition.

What this case report adds

- This paper describes dental findings in a 4-year-old boy with intestinal lymphangiectasia.
- Enamel defects were evident in primary teeth with a pattern corresponding to the timing of tooth development and concurrent disease activity.

Why this paper is important for paediatric dentists

- As in the case of many other causes of systemic upset, enamel defects may arise as a consequence of intestinal lymphangiectasia. These may affect primary as well as permanent teeth.
- Paediatric dentists need to be aware that the defects may add to any predisposition to caries, particularly when diet needs to be restricted and contains a high proportion of cariogenic items.

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Résumé. *Introduction.* La télangiectasie intestinale (IL) est une pathologie rare dont l'incidence et la prévalence sont inconnues en Australie et dans le reste du monde. Elle est caractérisée par des diarrhées, une légère stéatorrhée, un œdème, une perte entérique de protéines (entéropathie avec perte de protéines) et des canaux lymphatiques anormalement dilatés dans le petit intestin. Tandis que l'œdème et la diarrhée sont les caractéristiques cliniques prédominantes, d'autres éléments peuvent être observés tels une hypoalbuminémie, une hypogammaglobulinémie, une déficience en métaux, une hypocalcémie et des effusions pleurales chyleuses. Des répercussions dentaires ont été décrites en denture permanente (1,2) mais pas pour la denture temporaire.
Méthodes. Tandis que la présentation médicale de la maladie a été largement rapportée, peu de choses l'ont été sur les caractéristiques buccales. Une recherche sur Medline a donné deux articles évoquant les caractéristiques dentaires en denture permanente.

A ce jour, aucun rapport ne concerne la denture temporaire.
Résultats. Le denture temporaire d'un enfant de 4 ans avec IL présentait des défauts de l'émail reflétant le stade de développement de l'émail au moment où la maladie était active.
Conclusions. Cet article illustre la nécessité d'une implication précoce de l'équipe dentaire dans la prise en charge dentaire des enfants avec IL.

Zusammenfassung. *Hintergrund*. Intestinale Lymphangiektasie (IL) ist eine seltene Erkrankung, Inzidenz und Prävalenz sind sowohl in Australien als auch weltweit nicht bekannt. Die Erkrankung ist charakterisiert durch Diarrhoe, Steatorrhoe, Ödeme, enteralen Proteinverlust und abnormal erweiterte Lymphgefäße im Dünndarm. Während Ödeme und Diarrhoe die dominierenden klinischen Befunde sind, können auch andere Auffälligkeiten wie Hypoalbuminämie, Hypogammaglobulinämie, Spurenelementmangel, Hypocalcämie sowie Chylopleura beobachtet werden. Dentale Befunde wurden für die bleibenden Zähne berichtet, aber bisher gibt es keine Angaben zu den Auswirkungen auf Milchzähne.
Methoden. Während die Allgemeinsymptome der Erkrankung bereits ausführlich publiziert wurden, gibt es nur wenig zu den Zahnbefunden. Eine Medline-Recherche ergab zwei Veröffentlichungen mit Zahnbefunden an bleibenden Zähnen bei Patienten mit IL. Bisher gab es noch keine Berichte zu Zahnbefunden bei Milchzähnen.
Ergebnisse. Die Milchzähne des 4jährigen Patienten zeigten Defekte, welche die Zeit der Krankheitsaktivität reflektierten.
Schlussfolgerungen. Der Bericht zeigt die Bedeutung einer frühzeitigen Einbeziehung des zahnärztlichen Teams zur Betreuung der Zahngesundheit bei Kindern mit IL.

Resumen. Linfagiectasia intestinal (LI) es una afección rara y su incidencia y prevalencia en Australia y en el mundo es desconocida. Se caracteriza por diarrea, ligera esteatorrea, edema, pérdida entérica de proteínas (enteropatía perdedora de proteínas) y canales linfáticos anormalmente dilatados en el intestino delgado. Mientras que el edema y la diarrea son signos clínicos predominantes otros signos observados incluyen hipoalbuminemia, hipogammaglobulinemia, deficiencia de indicios de metales, hipocalcemia y derrames pleurales quilosos. Se han descrito los efectos dentales de LI para la dentición permanente (1,2), pero no se ha informado de su efecto en la dentición primaria.
Métodos. Mientras que se ha informado ampliamente de la presentación médica de la afección, se han publicado pocos informes de los signos bucales. Una búsqueda por Medline señaló dos informes con signos dentales en la dentición permanente de pacientes con LI (1,2). Hasta la fecha no ha habido informes sobre signos dentales en la dentición primaria.
Resultados. La dentición primaria de un niño de 4 años con LI, tenía los dientes con defectos del esmalte que reflejaban el momento del desarrollo del esmalte y el período en que la enfermedad era activa.
Conclusiones. El informe subraya la necesidad de la implicación temprana del equipo odontológico en el tratamiento dental de niños con LI.

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