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CASE REPORT

Calculus in a toddler with end-stage renal disease due to prune-belly syndrome

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Prune-belly syndrome is a congenital kidney and urinary tract anomaly which may lead to end-stage renal failure (ESRF). The present case describes an infant suffering from end-stage kidney disease due to prune-belly syndrome, undergoing chronic hemodialysis, with excessive calculus deposits which disappeared following kidney transplantation. Possible explanations are discussed. The first mechanism is associated with lack of oral function which may have caused pooling of saliva around the teeth enhancing precipitations of minerals. The second possible mechanism is associated with the child's uremic state. The third mechanism could be a disturbance in calcium-phosphor metabolism. It is possible that in the present case, the gastrostomy and the electrolyte disturbances characterizing ESRF had an additive effect.
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Keywords: prune-belly syndrome; calculus; end-stage renal failure; oral hygiene

Introduction

The prune-belly syndrome occurs almost invariably in males with an incidence of 1 in 30 000–40 000 live births (Gonzalez, 1996). This syndrome has a wide spectrum of clinical manifestations including the kidneys and urinary tracts as well as the musculo-skeletal system and the testes (cryptorchidism). Prune-belly syndrome is an example of congenital anomalies of the kidneys and urinary tract which may lead to end-stage renal failure (ESRF) (Gonzalez, 1996). The term originates from the clinical appearance of poor abdominal musculustructure which resembles the shape of a prune.

In early stages of renal failure, serum phosphate levels rise, causing reduction of serum calcium concentrations. This, in turn, will lead to secondary hyperparathyroidism and ultimately to bone lesions. Secondary hyperparathyroidism can be treated with active vitamin D compounds, together with phosphate-binding agents thus reducing bone disease involvement. When conservative measures fail to halt renal damage progression, renal replacement therapy (RTx), either dialysis or renal transplantation is needed. All dialysis modalities may provide approximately 20% of normal two-kidney renal function, meaning that dialysis patients remain in severe chronic renal failure (CRF). A successful RTx is the ultimate goal in treating children with end-stage renal disease (ESRD) (Avner *et al*, 2003).

Chronic renal failure and ESRF, as well as RTx are characterized by soft and hard tissue involvement of the oral cavity. The oral manifestations include: petechiae, ecchymoses, uremic stomatitis, gingivitis, periodontitis, reduced prevalence of caries, enamel hypoplasia and dental pulp obliterations (Wolff *et al*, 1985; Galili *et al*, 1991; Kho *et al*, 1999).

Flow rates of unstimulated whole saliva and stimulated parotid saliva are decreased in these patients, while increased salivary urea concentration has been detected (Blum *et al*, 1979; Epstein *et al*, 1980; Peterson *et al*, 1985; Gavalda *et al*, 1999). The pH and buffer capacity of unstimulated whole saliva were greater in CRF patients: pH of 6.4 compared with 5.6 in healthy individuals (Peterson *et al*, 1985).

Higher plaque and calculus indices and lower salivary secretion are prevalent among patients treated with chronic hemodialysis compared with healthy individuals (Epstein *et al*, 1980). Furthermore, quantitative assessment of salivary flow rate and alterations in the composition of stimulated and unstimulated saliva revealed that dialysis patients formed heavier calculus (Peterson *et al*, 1985; Gavalda *et al*, 1999).

In preschool children, calculus occurs much less frequently than in adults. Supragingival deposits of calculus often situated on the buccal surfaces of the maxillary molars and the lingual surfaces of the mandibular anterior teeth adjacent to the openings of the major salivary glands. Calculus which possesses masses

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of organisms is an important factor in the development of gingival and periodontal disease and its removal is therefore crucial as it possesses masses of organisms, hence is an important factor in the development of gingival and periodontal disease (White *et al*, 1997). Children with gastrostomy particularly need to decrease the accumulation of calculus. Calculus in these patients is associated with increased bacterial load and therefore be a risk factor to aspiration pneumonia (Jawadi *et al*, 2004).

In children with CRF and ESRF the urea nitrogen concentrations in the saliva are twofold higher than in healthy individuals. In these patients plaque pH was significantly more alkaline and directly correlated with salivary urea nitrogen concentration (Peterson *et al*, 1985). Phosphate concentration was found to be significantly elevated in the saliva of uremic patients (dialyzed and non-dialyzed) compared with healthy individuals. In turn, salivary calcium concentrations were similar to those documented in healthy subjects. As increased salivary phosphate was found also in primary hyperparathyroidism, the authors attributed their findings to the secondary hyperparathyroidism of the uremic state (Blum *et al*, 1979).

The present case describes an infant suffering from end-stage kidney disease caused by prune-belly syndrome, undergoing chronic hemodialysis, with excessive calculus deposits which disappeared following kidney transplantation. Possible explanations are discussed.

Case report

A 2-year-old boy was referred to the Department of Pediatric Dentistry at the Hebrew University-Hadassah School of Dental Medicine in Jerusalem by his pediatric nephrologist for evaluation. Medical history revealed that the boy had suffered from dysplastic kidneys and advanced chronic kidney failure from birth caused by prune-belly syndrome, which required dialysis already at the age of 1 month. He was initially treated with dialysis. Due to poor appetite, and in order to provide enough calories to assure adequate growth and development, the boy was fed via a gastrostomy tube. He consumed only little water (<100 ml) by mouth. The boy's main medical problems stemmed from a state of CRF and included hyperkalemia, secondary hyperparathyroidism and anemia. Oral examination disclosed extensive calculus deposits mainly on the lingual and buccal aspects of the mandibular incisors (Figure 1), and on the occlusal surfaces of the primary first molars despite good oral hygiene (Figure 2). Soft tissues were intact. Radiographs revealed clear pulp chambers and root canals (Figure 3a,b). Calculus and plaque removal were carried out (Figure 4). Chlorohexidine gel (2%) was applied on the teeth, and strict oral hygiene instructions were given. A follow-up examination 4 months later revealed extensive calculus in the very same sites (Figure 5). Again, the calculus deposits and the plaque were removed. Oral hygiene instructions were reemphasized. One month later the boy underwent kidney transplantation. He was examined in the dental clinic 4 and 8 months following transplantation. No new deposits of calculus were evident, although massive plaque accumulation was observed (Figure 6).

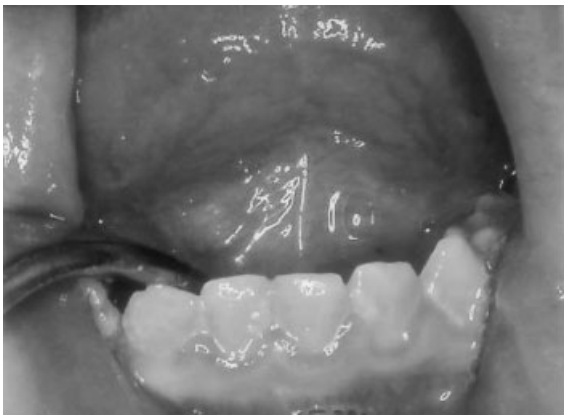

Figure 1 Extensive calculus deposits are shown on the lingual and buccal aspects of the mandibular incisors at the initial visit

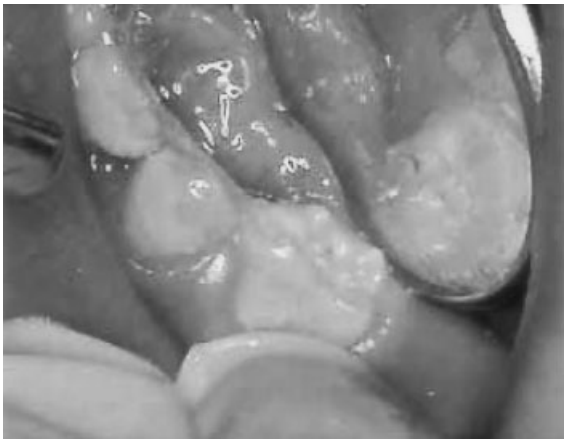

Figure 2 Extensive calculus deposits on the occlusal surfaces of the primary first molar

Discussion

The present case describes the oral findings in a child who suffered from ESRF as a part of prune-belly syndrome. The most significant finding was abundant amount of calculus which were mechanically removed but reappeared to the same extent 4 months later while he was on chronic hemodialysis treatment. Interestingly 4 months after renal transplantation no signs of dental calculus were observed. Moreover, the boy demonstrated fair oral hygiene during the time he was treated with dialysis, and poor oral hygiene after transplantation.

A number of pathogenic mechanisms are proposed. The first mechanism is based on the fact that until the kidney transplantation the boy was primarily fed via a gastrostomy tube. Previous researches have documented excessive accumulations of calculus in patients with gastrostomy (Jawadi *et al*, 2004). One study found that the presence of gastrostomy predisposed one to form

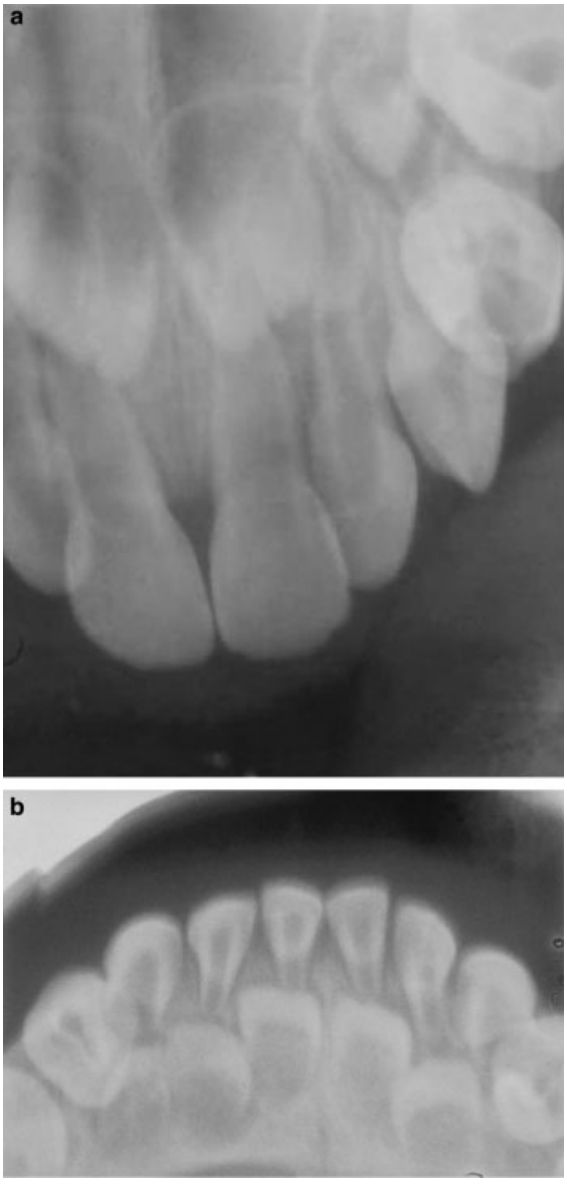

Figure 3 Radiographs revealed clear pulp chambers and root canals with no calcifications in the maxillary (**a**) or the mandibular incisors (**b**)

calculus, and oral hygiene procedures to reduce calculus were ineffective. It has been demonstrated that in some patients with gastrostomy, despite excellent oral hygiene, calculus had accumulated (Dicks and Banning, 1991). They suggested that even minute amounts of food consumed orally (despite the gastrostomy), might have served as a substrate to support plaque accumulation in the absence of oral function. The suggested explanations that are associated with plaque accumulation cannot be applied in our case as calculus was found even when no food entered the mouth, thus no plaque was formed. Therefore, it may be that the lack of oral function may have caused pooling of saliva around the teeth enhancing precipitations of minerals and thus, calculus formation.

The second possible mechanism is associated with the child's uremic state. Previous reports suggested that high

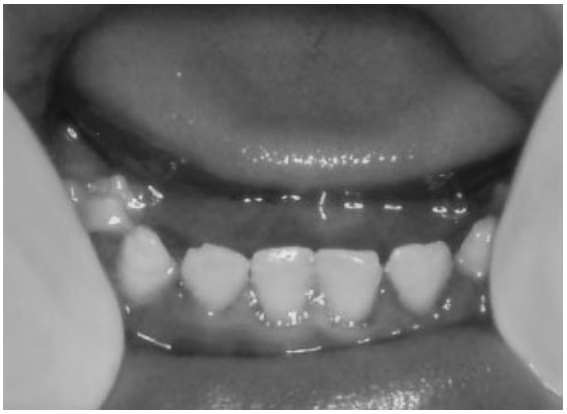

Figure 4 The mandibular incisor after calculus and plaque removal

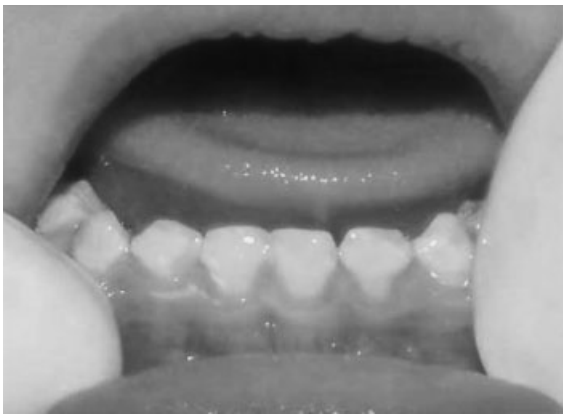

Figure 5 The mandibular incisors at the 4-month follow-up showing extensive calculus

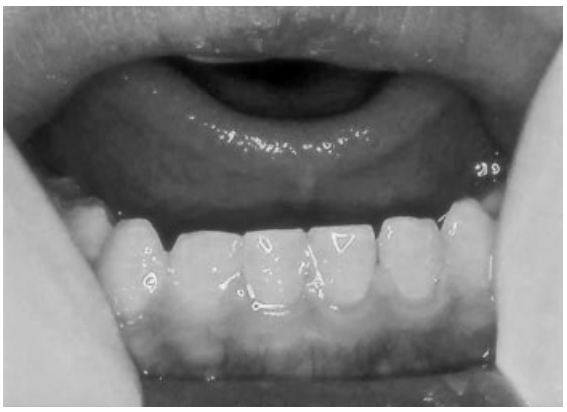

Figure 6 The mandibular incisors 4 months after transplantation. Demonstrating no new deposits of calculus, and massive plaque accumulation

amounts of urea in the saliva could enhance its buffering ability through urea hydrolyzation which consequently results in high concentration of ammonia (Blum *et al*, 1979; Epstein *et al*, 1980; Peterson *et al*, 1985; Gavalda *et al*, 1999). Both salivary flow rate and plaque pH

influence the saturation degree of calcium phosphates. It may be speculated that the elevated salivary pH led to a disturbance in calcium-phosphate products in the saliva. The decrease in the salivary pH after transplantation may have prevented calculus formation (Wolff *et al*, 1985; Kho *et al*, 1999).

The third mechanism could theoretically be a state of secondary hyperparathyroidism which is prevalent in children with ESRF. This child's serum parathyroid hormone (PTH) levels had not been outstandingly elevated and his plasma phosphate concentrations as well as the calcium-phosphate products were within normal range. However, occasional blood tests may not reflect prolonged impairment in calcium-phosphore metabolism. Plasma calcium-phosphate product levels in a child treated by peritoneal dialysis are generally higher than in a child with normal kidney function. Thus, calcium and phosphate may have been secreted in the saliva, thus enhancing calculus formation. Renal transplantation usually corrects a uremic state and its derivate electrolyte disturbances.

It is possible that in the present case, the gastrostomy and the electrolyte disturbances characterizing ESRF had an additive effect. The fact that this boy used his mouth only very little for essential functions such as eating, drinking or talking did not allow mechanical cleansing of his teeth and caused the saliva to accumulate without the normal washing in the oral cavity. This condition may have enhanced the formation of calculus despite the excellent oral hygiene that the boy demonstrated during dialysis. Following a successful renal transplantation, as the uremic state and the derivate electrolyte disturbances resolved, there was a significant developmental improvement.

The present case demonstrates the importance of multidisciplinary approach in treating children with ESRD and the need for further studies examining the influence of complex pathogenic mechanisms arising from renal failure on processes in the oral cavity. Further examination of saliva composition is needed to confirm the hypothesis argued here.

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