

Dental Problems in Hypophosphatemic Rickets, a Cross Sectional Study

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Abstract

Objective: Hypophosphatemic rickets is an uncommon metabolic bone disorder which affects all ages and both sexes. It is characterized by low concentration of serum phosphate levels, impairment of mineralization of bone matrix and teeth with variable etiology. Dental problems in this disorder have not been described well in previous studies.

Methods: All hypophosphatemic rickets patients who came to a referral clinic during 2008-2010 enrolled in this study. All patients had low phosphorous and high ALP, normal PTH and 25-hydroxy-vitamin D and normal or low level of serum calcium. After diagnosis all patients were examined by a dentist for enamel hypoplasia, taurodontism, dental abscess, gingivitis, dental caries, and dentition delay.

Findings: Nineteen patients were enrolled in this study. The average age of the patients was 10 (± 4.23) years (range 3-17). Seventy-nine percent of patients had regular follow-up after diagnosis of background disease. Dental caries and delay in the dentition were most prevalent (each one 47.7%) followed by enamel hypoplasia in 42.1% of the patients. Other problems were taurodontism in 15.8% patients, dental abscess and gingivitis in 10.9%.

Conclusion: Hypophosphatemic rickets is a disease with different clinical features; one of them is dental problem, dental caries is the most common problem.

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Key Words: Dental caries; Hypophosphatemia; Rickets; Enamel hypoplasia

Introduction

Hypophosphatemic rickets (HR) is a metabolic bone disorders manifested by musculoskeletal disorders specially in the lower extremities, growth retardation and dental problems^[1,2]. This disorder affects all ages and both sexes and is characterized by a low concentration of serum phosphate, elevated alkaline phosphatase, normal

or decreased serum calcium and inappropriate levels of 1,25-dihydroxy-vitamin D [1,25 (OH)₂ vit D]^[1-3]. Musculoskeletal disorders are due to mineralization impairment of the bone matrix and teeth with different etiologies^[3]. It is caused by renal tubular loss of phosphate into urine^[1]. X-linked hypophosphatemic rickets (XLHR) is the most common form of the genetic disorders causing rickets due to hypophosphatemia. This

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disorder results from mutation in the PHEX gene and the product of this gene has a role in phosphatonin inactivation^[2].

HR is more common in females with positive family history observed in 35.3% patients^[3], nevertheless, it seems this disorder is more frequently and more severe among males in XLH form^[2]. Common features are a short stature, backache, body deformities, joint pain, and fractures^[2]. Dental abnormalities include dental taurodontism with increasing mean crown-body to root ratio, ectopic permanent canines^[4], dental abscesses^[2,5] and enamel hypoplasia^[6].

Treatment of rickets with vitamin D (vit D) and phosphate supplements has been shown to prevent and cure the dental anomalies in some but not all patients. From 1970, 1-hydroxylated vit D has progressively superseded vit D itself for treatment of HR^[7].

Dental problems were reported in 23-67% XLHR patients^[3,6,7], but these problems have not been described well in previous studies. The aim of this study was evaluation of the dental problems in XLHR patients.

Subjects and Methods

This study was a cross sectional investigation that was conducted in a referral pediatrics endocrinology clinic in Iran during 3 years (2008-2010). All cases of suspected hypophosphatemic rickets were enrolled in this study. The diagnosis of hypophosphatemic rickets was made on the basis of clinical and radiological findings of rickets in a patient with low level of serum phosphate, normal or low level of serum calcium (<8.2 mg/dl) and normal levels of 25(OH)₂-vit D (<15 pg/mL), and normal or mildly elevated PTH with high level of serum alkaline phosphatase (>55 pg/mL). Patients who did not have a clear diagnosis or refused to participate in the study were excluded. After taking a dental history, all subjects had a dental examination (including dental caries, enamel hypoplasia, taurodontism, dental abscesses and gingivitis); OPG was performed if necessary.

Written informed consent was obtained from all patients or their parents. Clinical examination was performed to look for dental deformities by a trained dentist for all patients. Information about dental problem including: dental abscess, enamel hypoplasia, dental caries, gingivitis, taurodontism, and delay in the primary or permanent dentition was obtained. For permanent teeth, decay, missing, or filled teeth (DMFT) index and for primary teeth, decay or filled teeth (DFT) index was assessed in all patients. Dental caries was compared with healthy control age and sex matched children. Delay in eruption of teeth was based on times of eruption of the primary and permanent teeth from Logan and Kronfeld^[8]. Any defect in the continuity of the enamel surface was considered enamel hypoplasia^[9]. Taurodontism was suggested if there was any change in tooth shape and was confirmed by orthopantomography (OPG).

The statistical program for social sciences SPSS was used for data analysis. This study was approved by ethical committee of Tehran University of Medical Sciences.

Findings

In this study we enrolled 19 patients with hypophosphatemic rickets. Mean age of these patients was 10±4.23 years (range 3-17). Eleven (57.9%) patients were female and 8 patients (42.1%) male (Female to male ratio: 2/1.3). Fifteen (79%) patients had regular follow-up after diagnosis of background disease and 17 (89.5%) patients had at least one dental problem. The most common dental problems were dental caries and delay in eruption of the dentition in 9 (47.7%) patients. Enamel hypoplasia was found in 8 (42.1%) patients and 3 (15.8%) patients had taurodontism and all last group were male. Both dental abscesses and gingivitis were found in 2 (10.5%) patients. Six (54.5%) female and 3 (37.5%) male patients had dental caries. The prevalence of dental caries was significantly more frequent in case group ($P=0.04$) that was 10.5% in healthy control matched group.

Discussion

This study showed that HR has a variable age of presentation and it was more common in females^[2,3]. Dental caries was the most common finding in this study (47.7%) that was similar to Chaussain-Miller's study^[7]. Dental caries is one of the most common factors for pulp infection, so oral hygiene has an important role for caries prevention^[10]. Dental abscesses were found in 10.5% of the patients compared to 25-65% in other studies^[4,6,12]. In patients with HR the dentition is highly susceptible to dental caries and bacteria can invade easily from the oral cavity to the dental pulp by means of structural defects in the enamel. So these patients should be followed for prevention of dental abscesses. Dental abscesses may occur in the presence or absence of dental injury or decay. Difference of prevalence of dental abscess may be due to early diagnostic and treatment of these patients, although Shroff et al believe that prophylactic pulpotomy therapy may have been no benefit in preserving the primary dentition in XLHR patients^[13].

In our study, enamel hypoplasia was found in 42.1% patients which was higher than in previous reports. Enamel hyperplasia has been reported in 16.6% patients with permanent teeth by Bender and Naidorf, however they said the incidence of this disorder is 36.8% (14/38) in cases reported in the literature^[14], this is similar to our study^[11]. This disorder is more common in the Fanconi syndrome^[14].

Delayed dentition was found in 47% of patients. Although this phenomena is expectable in HR patients, no significant differences was reported in dental age compared with the respective chronological age in a study^[5]. Genetic mechanisms have a main role causing defective dentition and dentine mineralization^[15].

In this study taurodontism had a significantly increased tendency in male XLHR subjects. Similar findings were reported by Seow et al^[4]. In a controlled, longitudinal study, they showed taurodontism had a significantly higher tendency in male XLHR patients^[16].

Patients respond well to a combination of oral phosphate and 1,25(OH)₂-vitD (calcitriol)^[4,6]. The beneficial dental effect of phosphate supplements

after birth and the possible influence of maternal phosphate homeostasis suggest that most abnormal dental features are linked to hypophosphatemia and deficient renal production of 1,25(OH)₂-D^[7]. Some effects of 1,25(OH)₂-D on dentin may be indirect, through an increased intestinal absorption of calcium and phosphorus. 1,25(OH)₂-D may also exert some direct effect on dental cells, as odontoblasts and ameloblasts express the vitamin D receptor and respond in vitro to this vitamin^[17]. This supplement therapy can prevent dental anomalies by good dental mineralization and development^[4]. However, development of permanent teeth may persist despite therapy. It seems, the best results were achieved when medication started during early childhood^[4,18]. Dental care of these patients should consist of periodic examination, topical fluoride application, pit and fissure sealants and maintenance of good oral hygiene^[18].

Conclusion

Patients with XLHR show some peculiar dental abnormalities. In our study, dental caries as common as delay in dentition were the most prevalent problems in XLHR patients that could be reduced by proper dental care and good oral hygiene. So the dentist as well as pediatrician should be made aware of the features of disorder and early intervention.

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Conflict of Interest: None

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