DENTAL MANIFESTATIONS OF HEREDITARY HYPOPHOSPHATEMIC RICKETS CLINICAL, RADIOGRAPHIC AND BIOCHEMICAL STUDY.

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The X-linked hypophosphatemia VDRR is the most common type of rickets. In this study, ten children patients with different age and sex and clinically manifested VDRR were selected. Clinical, radiographic and biochemical investigations were performed. From the result it was evident that the most common dental manifestations were gingival abscesses and fistula. Also, enamel hypoplasia, looseness of teeth and early shedding of the deciduous teeth.

REVIEW OF LITERATURE:

Parathyroid hormone and vitamin D are the principal hormone products of vertebrate evolution that regulate extracellular phosphorus and calcium homostasis(1). Vitamin D acts on the small intestine to enhance absorption of both calcium and phosphate. Parathyroid hormone acts on the kidneys to conserve calcium and reject phosphate anion in glomerular filtrate(4). Inadequate calcium or vitamin D in diet or insufficient exposure to sunlight or both leading to rickets of alimentary origin and can be easily treated by supplying calcium and phosphate in diet and by administering an adequate amount of vitamin D(3).

There is another form of rickets which did not respond to vitamin D therapy. This condition was first described by Albright et al. 1937(5) as vitamin D resistant rickets (V.D.R.R.) or a refractory condition. It is called also, hereditary hypophosphatemic rickets or rickets of renal origin(2). Hypophosphatemic trait is inherited through an X linked dominant gene where the abnormal mutant allele appears on the X chromosome(15,16).

The prime features of this disorder are familial occurrence, lowered serum phosphate level associated with decreased renal reabsorption of phosphate and lack of response to physiological doses of vitamin D(5).

The clinical manifestations of the disease often appear in the second year of life, at the time affected children begin to walk. The children are of short stature and often have significant bowing of the legs. The extra-oral radiographic evidence revealed an increase anteroposterior diameter of the skull rachitic rosary and enlargement of wrists.

Dental alterations are the first observed sign of VDRR as spontaneous gingival and periapical abscesses without concurrent evidence of caries or root fractures. Due to the abnormalities in the dentin formation that evident in the histopathologic examination especially in the region of pulpal horns that reveals tubular defects of the dentin and elongated into pulp horns under incisal edges(8,9).

In radiographic examination of the dental structures often reveals enlarged pulp chambers and pulpal horns of primary dentition, extending to the dentinoenamel junction and extensive osseous lesions of rarification which involve the entire root and suggesting a periodontal in origin(10). Many others(11-14) reported that there are many abnormalities affecting both deciduous and permanent teeth as looseness, large root canals and pulp chambers, enamel hypoplasia, attrition and loss of lamina dura.

The phyophosphaturia usually persists after administration of vitamin D and is probably due to an inborn error or phosphate homeostasis, manifested by a selective defect in secondary active transport of phosphate in the brush border membrane of the proximal nephron(17). The abnormal low inorganic serum phosphate, normal serum calcium levels and elevated serum alkaline phosphatase are common biochemical characteristic of this disorder(11).

AIM OF THE STUDY:

The purpose of this study is to reveal the dental abnormalities in hereditary hypophosphatic rickets or vitamin D resistant rickets (VDRR) by clinical, radiographic and biochemical examination.

MATERIAL AND METHODS:

From in and out patient hospital, Cairo University, ten children patients with age range (3.5 to 12 years) with history of hereditary in origin Clinically manifested VDRR were selected. They had received dose therapy of 600,000 IU. of vitamin D for three injections with no improvement as noted from radiographic findings and laboratory data of serum calcium and phosphorus and alkaline.

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البحث العلمي الحادي عشر

الظواهر السنية في مرض الكساس المحوروث المقاوم لفيتامين (د)
دراسة كلينيكية - معملية لكمياء الدم وشعاعية

DENTAL MANIFESTATIONS OF HERIDITARY HYPOPHOSPHATHEMIC RICKETS,
CLINICAL, BIOCHEMICAL AND RADIOGRAPHIC STUDY

الملخص العربي

إن الكساس المقاوم لفيتامين (د) هو مرض آيضي يحدث في الطفولة، وقد تعرفنا عليه
اكتئابيا منذ ثلاثين عاما، وقد أضح أن يقاوم الجرعات المعتادة من فيتامين (د) وأنه لا يشفى
لا باستخدام جرعات كبيرة جدا منه. وقد تم فحص ثلاثين مريضا بالكساخ الظاهر اكتئابيا
وتراوح أعمارهم من 3-14 سنة، وقد اختبروا من مستشفى أبواليش للأطفال. وقد
تم فحص دقيق اكتئابيا للمرضى، وأخذ الأشعة اللازمة لكل منطقة في الفم، وكذلك الرأس,
واصل فحوصات الكيميائية الهيائية لتقدير مستوى الكالسيوم وفسفور الدم والاتزيم
الفسفاتازيم القلبي.

وقد كانت نتائج البحث تدل على: -

1) أن تقوس السيفان وإلتواء العمود الفقري لا يحدث إلا عند الجلوس أو المشي،
وكذلك عدم وضوح النتائج العملية الخاصة بهذا المرض، في البداية نتيجة مدى المنخفض
للترايب-المنخفض في الطفولة وهو الذي يمنع ظهور الفوسفاتات في البول وبالتالي يمنع
انخفاض الفوسفات في الدم كعلامة مميزة لهذا المرض.

2) وقد وجد أن طبيب الانسان هو أول مكتشف للأعراض المبكرة للمرض لأن هذا
المرض عادة يبدأ بأعراض ارتياحية وسنية، ولهذا يجب دوام الطبيب في ارشاد المريض إلى العلاج
الطبى السليم وبالتالي تجنب العواقب الخطيرة للمريض.

3) الأعراض السامة الشديدة في المرض أمكن مشاهدتها بقليل من الأمراض
العامة بصورة ارتياحية، والفحوصات العملية الدم بذلك أمكن التوصل للتشخيص السليم
للمريض.