Rickets review and patterns as seen at Orotta Pediatric National Referral Hospital

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Abstract

Objective: To review cases of rickets as seen at Orotta Pediatric National Referral Hospital and identify risk factors, common associated diseases, treatment outcomes and come up with recommendations.
Methods: All patients with clinical and radiological diagnosis of rickets as seen at Orotta Pediatric National Referral Hospital during the period of 2005 and 2006 were enrolled. Data on age, sex, maternal educational status, nutritional status using the weight for age, clinical manifestation of rickets, x-ray findings associated medical illnesses and treatment outcome were recorded and analyzed.
Results: Overall 120 children, 75% of who were from Asmara with the diagnosis of rickets were analyzed. Majority of the cases were children in the age group 12-23 months and 6-11 months accounting for 43.3% and 34.2% respectively. The distribution in the age group 0-5 months was 7.5%. More than 75% had at least one or two associated medical diseases such as pneumonia, malnutrition, anemia, delayed growth and development. Over 60% of the patients were not exposed to sunlight adequately and 20.8% were exposed while a glass window was closed. Clinical and radiological cure rate was over 95%. Four patients with severe chest deformities, severe malnutrition and associated severe pneumonia died on the 3rd day of their admission. Two patients with severe bowed legs responded unsatisfactorily to medical treatment and were referred for surgical corrections.
Conclusion: Rickets is not forgotten disease as presumed but rather it is still a formidable cause of morbidity and mortality in Eritrea a country where there is plenty of sunlight. Adequate information and communication will alleviate the problem.

Introduction

Rickets in childhood is manifested by softening and weakening of the bone resulted from deficiency of vitamin D. In the absence of organic disease vitamin D deficiency is due to sunlight deficiency. Rickets is not a fatal disease but complications and intercurrent infections are more likely to cause death in rachitic children [1, 2].

To date there is no systematic data collection to show the prevalence of rickets in Eritrea. However it is not unusual to see cases of rickets and its complications presenting with different associated clinical disease entities. Data from Health Management Information System (HMIS) and hospital statistics do not reflect the magnitude of the disease as the diagnosis is coded under other vitamin deficiencies and/or reported under the associated medical illnesses such as pneumonia, anemia, malnutrition, and musculoskeletal disorders. In this article a brief descriptions of the disease entity, risk factors and clinical manifestations as seen at Orotta Pediatric National Referral Hospital are described.

Methods

All patients with clinical and radiological diagnosis of rickets as seen at Orotta Pediatric National Referral Hospital during the period of 2005 and 2006 were recorded and analyzed. Data on age, sex, maternal educational status, nutritional status using the weight for age, clinical manifestations of rickets, x-ray findings associated medical illnesses and treatment outcomes were recorded and analyzed. Cases that did not come to the attention of the author were not included in this analysis.

Results

Over all 120 children with the diagnosis of rickets were analyzed with age and sex distribution. The peak incidence was in the age of groups 12-23 months accounting for 43.3% followed by age group 6 –11 months with 34.2%, 24 – 35 months with 14.2% and 0-5 months with 7.5%. A 10 years old child with a diagnosis of chronic renal failure and renal Osteodystrophy with typical radiological picture of rickets was included in the study. There was no single case in the age group 36-59 months. The male to female ratio was 1:1.3.

Three quarters of the patients were from Zoba Makeel predominantly from Asmara, 19% from Zoba Debub mainly from Mendefera, Dekemhare, Adikeih, Dibarwa and the rest 5.8% from Northern Red Sea, Anseba and Gash Barka.

On assessing the educational background of caretakers (mothers) only two mothers were illiterate 15.8% were between 1-4th grade, 15.8% mothers were between 5-8th grade, 38.3% mothers between 9-12th grade and 3.3% of mothers were with grades above 12th.

Most of the care takers (mothers) 62.5% accepted that they did not expose their child to adequate sunlight, 20.8% claimed they use to expose their babies regular and another 16.7% used to expose while the window glass was closed.

When asked the reason for not exposing sunlight, mothers who did not or inadequately exposed their babies responded that they were afraid of cold exposure and had no knowledge of sunshine as source of vitamin D and its importance for bone development. The caretakers who expose their babies while the window glass was closed claimed that they thought they were doing the right thing and know the benefit of sunlight as source of vitamin D.

The majority of the patients had some degree of malnutrition ranging from mild to severe forms, with severe malnutrition and under weight each present in about 30% of cases (Table 1).

Table 1 Nutritional status of children with rickets using Weight for age classification

Weight for Age	Sex		Total	%
	M %	F %		
> 80% STD	24	28	52	43.4
60-80 % STD	14	20	34	28.3
< 60% STD	9	12	21	17.5
60-80 % STD with edema	4	5	9	7.5
< 60% STD with edema	1	3	4	3.3
Total	52	68	120	100%

All the children had at least 4 of the following skeletal manifestations such as craniotabes, caput quadratum, frontal bossing, rachitic rosary, Harrison's groove, widen wrist, double malleoli, bowleg kyphoscliosis and green stick fractures (Table 2).

Table 2 Skeletal manifestations of children with rickets

Skeletal manifestations	Num. of Cases	%
Craniotabes	50	41.7
Caput quadratum	20	16.7
Frontal bossing	20	16.7
Rickets Rosaries	120	100
Harrison's groove	54	45
Widen Wrist	120	100
Double malleolus	120	100
Bowleg	43	35.8
Green stick fracture of tibia and fibula	2	1.7

Only 40% of patients presented with isolated skeletal deformities while 60% had at least one or two associated systemic medical illness (Table 3). About half the patients had some degree of malnutrition or recurrent pneumonia, 15% had anemia and 15% had delayed growth and development (Psychomotor delay) mainly failure to stand unsupported at the expected age (Table-3). Clinical evidence of tetany and seizure disorder was seen in 5% or less of patients.

Table 3 Case of rickets and associated medical illness

Associated illness	No of case	% of cases
Malnutrition	68	56.6
Recurrent pneumonia	56	46.6
Anemia	18	15
Delayed growth &development	18	15
Tetany	6	5
Seizure disorder	4	3.3
Isolated skeletal manifestation	48	40

Radiological investigations done were consistent with the diagnosis of rickets showing beading at the costochondral junction, cupping and fraying at wrist joint and ankle joint; bone softening and evidence of bowing and/or fracture bones. A thorough biochemical test was not done. Liver function test done in 20 patient showed, elevated alkaline phosphatase. About 15% children had clinical evidence of anemia confirmed with low hemoglobin value. Cell morphology and red blood cell indices were consistent with iron deficiency anemia. Calcium level in those children manifested by tetany and seizure disorder were low than the normal reference value.

All the children were treated either as inpatient 40% or outpatient 60% depending on the severity of the illness and associated disease. Two thirds of the patients received single high dose vitamin D intramuscularly while the rest of the patients were treated with vitamin D3 drops for 3 to 4 weeks depending on the availability. Children with severe malnutrition were treated with required therapeutic feeding and those who had anemia with iron syrups. Among the 56 patients with recurrent pneumonia 26 had clinical evidence of severe pneumonia deserving hospitalization and the rest were managed as an outpatient with antibiotics. Calcium gluconate was given to those patients who presented with tetany during the attack and maintained with oral calcium gluconate. The children who manifested with seizure disorder where initially managed with anticonvulsants. The diagnosis of rickets was made late in the course of the disease. After treatment with vitamin D and calcium supplement, seizure resolved and anticonvulsant discontinued.

Follow-up after treatment with clinical and radiological response was done 1-2 monthly for a minimum of 4 visits. Cure was achieved in 95% of the children. Four patients had severe chest cage deformities, severe malnutrition and severe pneumonia and died within three days of admission. Two cases with severe bowed legs that did not show marked improvement of the degree of deformity after medical treatment were referred for orthopedic corrections.

Discussion

Rickets is a disease of the growing bones characterized by defective mineralization of the osseous matrix and epiphyseal cartilage, resulted from lack of calcium and phosphorous metabolism. In the absence of underlining organic causes such as liver, kidney diseases and/or prematurity, rickets is caused by lack of vitamin D resulted from failure of adequate direct sunlight exposure or deficiency of vitamin D in the diet [1,2 3]. Vitamin D deficiency in childhood can cause rickets whose sign and symptoms include bone deformity, bone fracture, muscle weakness, developmental delay, short stature, failure to thrive respiratory distress, recurrent pneumonia, tetany and heart failure [1,2,3,4,5]. The effect of sub-clinical rickets are unknown. Vitamin D deficiency in the first few months of life is relatively rare but has occurred congenitally in infants of mothers who were severely deficient of vitamin D during pregnancy. [6, 7, 8, 9]

Epidemiologically, the prevalence of rickets is not

known globally and nation wide. It was epidemic in industrialized cities in the 20th century and as of the year 1960 virtually eliminated through vitamin D supplementation and milk fortification with vitamin D. Rickets remain serious health problem in some developing countries such as China and Turkey where studies showed in selected areas the prevalence as high as 40% and 33% respectively 9. Due to the convergence of risk factors, rickets is actually most common in those regions of the world where there is plenty of sunlight; such as India, Ethiopia, Libya, Morocco and Tunisia [2, 3, 4, 10]. The situation in Eritrea is unknown as to date there is no systematic data collection to study the magnitude of the disease. The only reports are the ones from facility and most of the time is believed to be underreported in most instances. The clinical sign except for those with the severe form are also believed to be overlooked by health personnel. In this study it was common in the age group 6-24 months, similar to data done elsewhere [2, 3].

There are some factors that contribute to the development of rickets like maternal vitamin D deficiency during pregnancy which is often asymptomatic and lead to insufficient fetal vitamin D store 4, 8. Breast milk has low vitamin D and babies on breast milk unexposed to other source of vitamin D are likely to develop rickets. These are the groups who develop rickets during their early infancy. In this series 7.5% developed rickets in the 1st six months of their life with the youngest at age 3 months. The magnitude of the illness in this age group is as high as 19-26% in studies done elsewhere [2]. Several studies have shown without postnatal exposures to sunlight vitamin D of fetal origin was depleted by 8 weeks in a child on exclusive breast feeding. Breast milk produces much less than the daily recommended (200-400 iu/day). Only few hours of total summer sunlight produces enough vitamin D to avoid deficiency for several months. Sunlight exposure of 30 minutes per week diaper only or 2 hours per week fully closed without hat appears to be sufficient to prevent deficiency [4, 6, 10].

Indoor confinement during daylight hours is a leading cause of rickets. The majority of patients were not exposed to adequate sunlight. The reason given by care takers for not exposing their children to sunlight was fear of cold exposure (cold climate), sun light does not enter their rooms and were afraid to take their children out doors; use of excess clothes and covering even when they were outdoors in some occasions as well as poor knowledge despite their educational background on the benefit of sunlight as source of vitamin D. Ultraviolet light with a rad of 288nm required for synthesis of vitamin D does not cross ordinary glass as a result babies exposed to sunlight while the window glass is closed are at higher risk to develop rickets1. In this study 16.6% children whom their mothers claimed they had exposed their children to sunlight regularly via the closed window glass still developed rickets.

High latitude contributes to reduced exposure to sunlight [1, 2]. This is not a problem for Eritrea as the country is located north of the equator between latitudes 12°22'N-18° 02'N, an area where there is 12 months of sunshine [11].

Living in areas with many high building that block sunlight to nearby houses may contribute to less exposure to sunlight and development of rickets. Though there are few very high buildings even in the big city to block sunlight in our situations some living in the palaces and crowded areas reported that they have poor sunlight access to their rooms. A pollution that blocks sunlight in some industrial cities may lead to poor exposure to sunlight. This is not a problem in our situation as there are no heavy industries to cause such pollutions. Use of sun screen may also limit the entry of sunlight to the body. Increased birth order probably due to poor asymptomatic mother with depleted store may lead to increased risk of rickets in children [12, 13, 14]. In our case though further study is recommended to examine the distribution among siblings the illness is seen even among the 1st born children.

Exposure to lead and anticonvulsants contributes to low absorption and interfere in the metabolism of calcium respectively [15]. Further studies are required to see the impact of lead and anticonvulsant in the development of rickets in our situation. Though theoretically it is believed that rickets only develop in a growing child and therefore less likely to happen in a child with malnutrition, studies had shown that there is an association. Studies from Bangladesh show that most case of rickets are stunted and 25-50% show signs of protein energy malnutrition, vitamin A, vitamin, C and calcium deficiencies, and contribute to significant morbidity burden [14]. Similar studies done in Ethiopia have also shown strong association of protein energy malnutrition and rickets [2, 3]. In this series more than half of the cases had some degree of malnutrition, a quarter of who had severe malnutrition were managed with therapeutic feeding. Less than one quarter of the children had clinical and laboratory evidence of anemia. Other studies done else where has also demonstrated considerable associations between rickets and anemia [2, 3].

The biochemical changes seen in patients with rickets are calcium level will be normal or low, phosphorous is low, parathyroid hormone is high, alkaline phosphatase is high, vitamin D25-OH and vitamin D1, 25-OH are usually low [1, 2, 3]. In this analysis detailed biochemical analysis was not done, alkaline phosphatase done in 20 patients show elevated level, more than 3 times the normal value. Calcium level was low in those children who manifested with seizure disorder and/or tetany.

The resorption of calcium and phosphorous from the bone will result to progressive softening and widening of the bone structures, delayed closure of fontanels or wide fontanel, caput quadratum, frontal bossing, craniotabes, particularly in the very young, rachitic rosary, prominence of the costochondral junction of the ribs with olive shaped enlargement, Harrison's grove, kyphoscliosis, widen wrist with knob like appearances, double malleolus of the ankle joint, ankle deformity of the epiphysis, splaying deformity, genovarus, genovalgus, bowleg, knock knee, green stick fracture and delayed teeth eruption are the

common skeletal manifestations[1,2,3,14,16,17]. The clinical manifestation may range from simple sub clinical to very severe deformity. In this series majority of the patients presented with more than 4 different skeletal manifestations with varying degree of severity.

Radiology is usually not only diagnostic but also shows the degree of severity from bone softening to the different degree of bone fraying, cupping of the growing end plate and radiological evidence of deformities like bowing, kyphoscliosis, and fracture. X- ray coupled with clinical assessment is also important to assess the treatment outcome. In this series all the skeletal x-rays done were consistent with the diagnosis of rickets and X- ray done 4-6 weeks post treatment had shown radiological improvements to complete healing.

Majority of the patients with rickets had associated medical illnesses upon presentation, such as recurrent pneumonia, delayed growth and development, malnutrition, seizure disorder fracture[3,14,16,17,18]. In this series only 40% had isolated skeletal manifestation, the rest presented with at least one or two associated medical illnesses.

Treatment outcomes depend on early recognition early treatment and treatment of the underlining or associated disease entity and close follow up until full recovery. In this series except for the four children with severe chest deformity, severe malnutrition and clinical evidence of severe pneumonia who died within 3 days of admission the cure rate was over 95% evidenced both clinically and radiologically. Two children with severe bowleg who had unsatisfactory result to the medical treatment were referred for surgical correction.

In conclusion this series of studies documented that rickets is still a significant cause of morbidity among the children in Eritrea. Vitamin D deficiency in children can easily be prevented through very simple and effective interventions such as exposure to adequate sunlight and provision of diet as recommended to the specific age groups. There is urgent need to provide health promotion to mothers so as to encourage them to expose their children to adequate sunlight from early life in order to eradicate this easily preventable disease.

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