ORIGINAL ARTICLES

Types and Clinical Profile of Rickets in a Tertiary Care Hospital

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Abstract

Background: There are different types of rickets. Rickets presents with various clinical signs and symptoms. Familial X linked hypophosphatamic rickets (XLHR) is reported to be the commonest one. **Objectives:** To find out the types of rickets and the presenting features of rickets. **Setting:** Department Paediatrics, Bangabandhu Sheikh Mujib Medical University (BSMMU). **Design:** Descriptive type of study. **Methods:** A total number of 20 children with rickets were included in this study during the period of January 2004 to July 2008. A questionnaire was used for compiling the information. Data were compiled manually and expressed as frequency distribution table. **Result:** Male: Female ratio was 13:7. Mean age of the patients was 49.36 months. Nutritional and familial hypophosphatamic rickets constituted the majority, each of the type being 40%. Common clinical presentations included limb weakness (90%), growth failure (90%) and repeated respiratory tract infection (80%). Widening of the wrist were present in 90% of patients. Rachitic rosary and protruded abdomen were present in 80% of cases. Radiological findings of rickets were present in 100% of patients. **Conclusion:** Nutritional and XLHR rickets were the common type. Common clinical features were weakness of limbs, growth failure and widening of wrists.

Key wards: Rickets, nutritional, XLHR, RTA.

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Introduction

Rickets result from the failure of osteid tissue to calcify in growing bone. Counter part of this abnormality in adult is called osteomalacia.[1] The term is derived from the old English word "twist" or "wrick" and throughout the history, children with rickets could be identified by their bowed legs and knocked knees.[2] The typical clinical pictures of rickets include delayed growth, widening and bowing of weight bearing bones, tooth enamel hypoplasia, muscle hypotonia and even tetany.[3-5] There are various types or rickets, including vitamin D disorders, familial cause, renal cause and others[6]. Familial X linked hypophosphatamic rickets (XLHR) is reported to be the commonest type[7]. XLHR is synonymous with vitamin D resistant rickets. Sometimes, rickets is caused by the lack of vitamin D in diet and lack of exposure to sun light when it is called nutritional rickets.[8] Factors that may limit sun exposure in children include use of sunscreens, increased indoor activities, industrial pollution, living in slum areas, wearing long dresses etc.[8] Present study was undertaken to find out the types of rickets and the presenting features of it among the admitted patients in the paediatric department of Bangabandhu Sheikh Mujib Medical University (BSMMU).

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Patients and Methods

It was a descriptive type of study conducted in the department of paediatrics, Bangabandhu Sheikh Mujib Medical University (BSMMU). All the infants and children admitted in the paediatrics ward with various types of rickets between the period of January 2004 to July 2008 were included in the study. Total number of patients were 20. Patients with rickets already on treatment were excluded from the study.

Procedure

A questionnaire was prepared which contained, detailed history including**:** nutritional intake, exposure to sunlight, repeated diarrhoeal episodes, urinary problem, hepatic problem, history of convulsion, detailed drug history and family history. Thorough physical examination for clinical

signs of rickets. Anthropometric measurement including weight and height/length. Routine blood examination including Hb%, total WBC count, differentials and ESR. Routine examination of urine. For evidence of rickets, X-ray chest and affected parts of the body. Biochemical investigations including**:** serum calcium, phosphate, alkaline phosphatase. Urinary phosphate and calcium, serum creatinine, serum alanine aminotransferase (ALT) and electrolytes.

After taking informed consent, detailed history was taken and a thorough clinical examination was done for filling up the questionnaire. Relevant investigations were also recorded in the questionnaire. Data were compiled manually and expressed as frequency distribution.

Results

Table -I shows the demographic profile of the patients. Mean age of the patients was 49.36 months and male: female ratio was13:7. There was lack of sun exposure in 15% patients, and 75% of patients had vitamin D deficient diet. Three patients (15%) had family history of rickets (sibs had rickets). Majority of patients came with weakness of limbs (90%) and growth failure (90%). There was history of repeated respiratory tract infection in 80% of patients (Table-II).

Widening of the wrist was found in 90% and rachitic rosary was present in 80% of patients. Protruded abdomen was also present in 80% of cases. Fifty percent patients had pectus excavatum & 40% had pectus carinatum. Harrison sulcus was present in 40% of the children. Table-III shows the types of rickets found in this study. Nutritional and familial hypophosphatamic rickets constituted the majority, each of the type being 40%.

Table-I

Demographic profile of the patients

Finding	Number (n=20)	Percentage
Age in months (with rage mean)	49.36 (9-156)	-
Male: female ratio	3 : 7	-
Family member >4	17	85
Living in slum	16	80
Lack of Vitamin D in diet (more carbohydrate)	15	75
Lack of exposure to sunlight	3	15
Family history of rickets	3	15

Table-II

Clinical findings of the studied patients (n=20)

Symptoms/Signs	Number	Percentage
Symptoms		
Limb weakness	18	90
Growth failure	18	90
Repeated respiratory tract infection	16	80
Limb pain	10	30
H/O malabsoption	5	25
Urinary complaints	4	20
Signs		
Widening of the wrists	18	90
Rachitic Rosary (beaded ribs)	16	80
Protuded abdomen	16	80
Pectus excavatum	10	50
Pectus carinatum	8	40
Cubetus varus (Knocked knee)	9	45
Cubetus valgus	5	45
Harrison salcus	8	40
Dental caris	6	30

Table-III

Types of Rickets (n=20)

Name	Number	Percentage
Nutritional	8	40
Familial hypophosphatamic	8	40
Renal tubular acidosis (type-2)	2	10
Renal osteodystrophy (CRF)	1	5
Unclassified	1	5

Table-IV shows that 100% children in this study had radiological findings of rickets including, fraying, cupping, widening of distal end of metaphysis as well as beading of the ribs. Other radiological findings included generalized osteopenia and increased distance between the epiphysis and diaphysis. Serum calcium was found low in all the types of rickets but was more so in distal renal tubular acidosis, renal osteodystrophy and in unclassified category (Table-IV). Whereas, serum phosphate was low in familial hypophosphataemic type and high in renal osteodystrophy (Table-IV). Alkaline phosphatase was increased in all the types. Mean serum potassium was low in distal RTA cases (2.7 mmol/l) and high (5.3 mmol/l) in renal osteodystrophy. Serum chloride was raised in renal tubular acidosis. Serum bicarbonate was low in renal osteodystrophy and renal tubular acidosis. Urinary phosphate was raised (215.8 mmol/l) in X linked familial hypophosphataemic rickets (Table-IV).

Table-IV

Investigation profile of the cases

Findings	Nutritional n-8	Familial hypo phosphatamic	RTA (Distal) n=2	R.osteody strophy n=1	Unclassified n=1
Radiological evidence n=20	8	8	2	1	1
Ultrasonogram n=3	-	-	Neph calcinosis (1)	Small kidney	-
S. Calcium mg/dl	8.36	8.9	7.8	7.5	8
Mean (range) n=20	(6.8-9.8)	(7.3-9.4)	(7.2-8.2)		
S. Phosphate (mg/dl)	3.91	2.07	3.5	5.2	3.2
Mean (range) n=20	(2.75-5.0)	(1.8-3.5)	(3.2-3.8)		
S.ALK phosphatase (mmol/dl)	908.25	899.7	1565	4230	881
Mean (range) n=20	(408-1248)	(544-1460)	(1198-1980)		
S. Sodium (mmol/l)	n=4	n=4	n=2	n=1	n=1
Mean (range) n=12	141.5	141	131	138	145
	(135-151)	(138-145)	(127-132)		
S. potassium (mmol/l)	4.9	4.42	2.7	5.3	4.2
Mean (range) n=12	(4.6-5.3)	(3.9-4.9)	(2.4-3.1)		
S. Chloride (mmol/l)	102	92.5	110.5	109	99
Mean (range) n=12	(98-104)	(88-110)	(105-116)		
S. Bicarbonate (mmol/l)	24.26	23.74	16.4	19	23.8
Mean (range) n=12	(20-26.2)	(20-25)	(25.70-18)		
Anion gap (mean) n=3	-	-	n=2	n=1	-
			0.95-1	12	
Urinary phosphate (mmol/dl)	n=2	n=7	n=2	n=1	-
Mean (range) n=12	1.15	215.8	13.5	12	
	(0.10-2.2)	(100-320)	(10-17)		
Urinary calcium (mg/dl)	n=3	n=2	n=2	n=1	
Mean (range) n=8	-	0.53	0.9	2.1	1
	(0.4-0.7)	(0.8-1)	(2-2.2)		
Serum creatinine mmol/l	n=5	n=3	n=2	n=1	n=1
Mean (range) n=12	0.6	0.5	0.95	1. 5	0.4
	(0.5-0.9)	(0.4-0.8)	(0.95-1)		
Urinary pH	n=4	n=3	n=2	n=1	-
Mean (range) n=10	6.75	6.5	6.5	6	
	(5.0-7.0)	(6-7)	(6.1-6.5)		
Mean 24 hrs urinary	n=3	n=2	n=2	n=1	-
protein(gm/dl) n=8	0.35	0.36	0.4	0.5	

Discussion

Rickets usually occurs during the periods of rapid growth, when the body demands high levels of calcium and phosphate. Usually rickets starts to appear at 4-24 months of age and is uncommon in newborn. Clinical sings may occur later.[8] In our study mean age was 49.36 months with the age range of 9 months to 156 months. Fischer et al in their study found the mean age of rickets as 69 months with age range of 36-98 months[9].This mean age was more or less consistent with our series. But one patient of renal osteodystrophy presented to us with rickets at the age of 13 years and extended the age range of our series up to 156 months. The male: female ratio in the present study was 13:7 which is also consistent with Fischer et al study where 64% patients were male[9]. This may also be a reflection of our male dominating society where female child is usually given less care and not brought to hospitals.

Soliman et al showed that rachitic patients were significantly shorter and growth velocity of them was low[3]. We did not see the growth velocity but growth failure was present in 90% of patients in our series. In the present series majority of the patients presented with weakness of limbs (90%), repeated respiratory tract infections (80%) and limb pain (25%). But in the Fischer et al study,[9] there was limb pain in 93% patients and weakness of limbs in 86% patients. Ninety percent of our children had widening of wrist and 80% had beaded ribs, 45% had cubetus varus (knocked knee) and 25% patients had cubetus valgus which were more or less similar to Fischer et al series in which 71% children had beaded ribs, widening of wrist and knocked knee. We did not find any tooth enamel hypoplasia and tetany among our studied patients.

Although familial X linked hypophosphatamic rickets (XLHP) is reported to be the commonest one,[6,7] equal number of nutritional and familial X linked hypophosphatamic rickets (XLHP) was found in the present study each type being 40%. Small sample size and geographical location may be a factor for this difference. More over one patient in the nutritional rickets group was a diagnosed case of Wilson's disease and another patient was a case of cerebral palsy. This could be another reason for more numbers of nutritional rickets in our series.

Majority of the patient came from the income group of 4000-5000 Taka/month and 80% of rachitic children in our study came from slum area. This may suggest some relationship of rickets with economic status but it is also to be noted here that, perhaps this class of people mostly attend this hospital. In this study we found one unclassified rickets who presented at the age of 10 months with history of adequate dietary intake and sun exposure. He had low calcium (8mg/dl), normal phosphate level (3.2mg/dl) and high alkaline phosphates level (881mmol/l). His electroytes, serum creatinine and urinary phosphate excreation were normal. As because necessary hormones and enzyme studies could not be done due to inadequate logistic support, actual diagnosis could not be made and we diagnosed this case as unclassified rickets.

While analyzing radiological findings of the patients, the study found that 100% of patients had radiological evidence of rickets (Table IV). In fact this was the first line diagnostic tool of our cases. Biochemical changes seen in the study were consistent with documented reports [6,10,11] and helped us to classify the types of rickets (Table IV).

As expected, serum potassium was low in RTA, and high in renal osteodystrophy. Hyperchloraemia was also present in RTA cases (mean 110.5 mmol/l). Substantial systemic acidosis was present in RTA (mean serum bicarbonate 16.40 mmol/l) and mild acidosis was present in renal osteodystrophy (19mmol/l). Despite having substantial acidosis, urinary pH of our RTA patients was more than 6 (mean 6.5, range 6-7). So diagnosis of distal RTA could be done with strong emphasis.[12] In addition absence of anion gap was also found in our RTA cases further strengthening the diagnosis.[12] Urinary phosphate excreation was very high in familial hypophosphatamic rickets (mean 215.8 mmol/day). In other types of rickets it was in the normal range. This finding was consistent with established data.[6] Although urinary calcium was not high but its excretion was found in the upper normal level in distal RTA patients (mean 2.1)[13]. It is established that urinary calcium excreation is higher in RTA patients[12]. In other types, it was much lower (Table-IV).

Serum creatinine was found high (1.5 mg/dl) in the only renal osteodystrophy case of the study. Twenty four hours urinary protein excretion (UTP) was also high in that particular patient. In other types serum creatinine and UTP was normal. These findings were consistent with extablished findings.[11,14] On ultrasonography of the, abdomen 1 of our 2 RTA cases had nephrocalcinosis and the only renal osteodystrophy patient had small shrunken kidneys substantiating our diagnosis.

Conclusion

This study concluded that nutritional rickets and familial hypophosphatamic rickets are the common types of rickets found in our setting. Clinical features and laboratory findings are similar with the documented findings. But it

is to be noted here that this was a tertiary care hospital based study. So the real picture of our country may not be reflected in this study. A community based study with adequate sample size is needed for getting real information about rickets in our country.

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