Case Report

Co-occurrence of Rickets and Scurvy- A case report

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Abstract:

Now a days, rickets and scurvy are considered rare except in certain situations. Co-occurrence of rickets and scurvy develops in a child when the risk factors of both conditions present simultaneously. Infant and young children are more vulnerable to micronutrient deficiencies. Appearance of clinical and radiological features of both conditions are dependent on the order and length of first disease prior to the development of the 2nd illness. One of the two diseases will dominate over the other and mask the features of the non-dominant disease. Screening for rickets should be considered for children with poor growth/ development, seizure activity/tetany, and children with chronic malabsorptive states and scurvy should be evaluated to exclude co-occurrence. Here, we report a 14 months old female child presented with cough and respiratory distress for 2 months and poor weight gain since early infancy. She was a preterm, low birth weight baby on exclusive breast feeding without any vitamin supplementation. She had history of inadequate sunlight exposure. Her current diet was lack of fruits and vegetables. Incidentally, she was found having rachitic changes. Her radiology showed features of both rickets and scurvy. So, finally she was diagnosed as a case of persistent pneumonia, co-occurrence of scurvy and rickets with failure to thrive. She was treated with vitamin D, Calcium, Vitamin C, antibiotics and other supportive measures.

Key words: Rickets, Scurvy, Micronutrient deficiency, Preterm, Low birth weight (LBW)

Introduction:

Scurvy and rickets occur simultaneously when a culture or society has risk factors for both conditions. Diet is one of the most important factors for developing a co-occurrence of both conditions. [1] Scurvy is the disease caused by a deficiency in vitamin C. Vitamin C, is a water soluble vitamin and an essential nutrient requiring to obtain from the diet. Vitamin C rich foods include fresh fruits, vegetables, and human breast milk, with small amounts in raw liver. A deficiency of this vitamin is caused by a deficient diet and/or poor nutrient absorption.[2] Vitamin D deficiency (called nutritional rickets) is the most common and typically due to a lack of ultraviolet B (UVB) ray exposure, dietary deficiency and malabsorption in the gut of vitamin D.[3] Infants and young children are more vulnerable to micronutrient deficiencies for rapid growth and use up their stores.[4] Now a days, rickets and scurvy are considered rare which develop only in peculiar situations. Lewis et al recently reported cases of scurvy and rickets have been documented in refugee and ethnic minority populations.[5] Moreover, infants and children are immunocompromised with increased risk of acquiring infectious or diarrhoeal diseases which can causes malabsorption of important nutrients. [4] Socioeconomic status is another risk factor as it may limit access to vitamin C and D by restricting diet and by adopting particular behaviours. After weaning, a diet rich in vitamin C and vitamin D is essential, with adequate sunlight exposure.[6] Here, we report a rare case of 14 months old girl who presented with persistent pneumonia and poor weight gain ,incidentally, she was found having rachitic changes and his radiological findings were correspond with both rickets and scurvy.

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Case summary

Mitul, a baby girl of 14 months, 3rd issue of non-consanguineous parents, immunized, belongs to a lower class family was admitted on March,2020 in pediatric ward in AWMCH hospital with the complaints of cough and respiratory distress for 2 months and poor weight gain from early infancy. Previously she was hospitalized and diagnosed as a case of persistent pneumonia. She was being treated with several antibiotics with no cure, hence was referred to our hospital for better evaluation.

She was delivered normally at preterm with low birth weight (1500 gm) with uneventful perinatal period but

she did not receive any vitamin and iron supplementation. She was given exclusive breast milk for 6 months. Then complementary feeding was started but poor in quality. She was never offered fresh vegetables and fruits, meat, fish ,egg etc. On query, her exposure to sunlight was also inadequate, most of the time she used to spend in indoor. Her milestone of development was age appropriate.

She had no h/O recurrent loose motion, skin lesions, easy fatiguability, limb pain, gum bleeding or contact with tuberculosis patients. All other family members were in good health.

Physical examination: Mitul had distressed look with intercostal recession, respiratory rate-50/min, Pulse-180/min, Temperature 98^0F, SPO2-86%, incidentally she was found having rachitic changes such as box shaped head with wide open anterior fontanelle, widening of costochondral junctions of ribs, widening of both wrists and ankles without sign of inflammation. Her weight 6.4 Kg, height 68cm, upper segment- 40 cm, lower segment-28 cm, U:L – 1.4(normal), occipitofrontal circumference (OFC) -46 cm (50^{th} centile), mid upper arm circumference (MUAC) – 115 mm, WAZ -3.8 (severe wasting), HAZ- 2.7 (moderate stunting). On auscultation, there were crepitations on left lung field but no murmur. Other systemic examination revealed no abnormalities.

Investigations:

Complete blood count : Hb- 12.0g/dl, white blood cell-16,000/cmm, neutrophil-65%- neutrophilic leukocytosis reflected infection S. calcium- 10mg/dl(normal), S.Phosphate - 2.20mg/dl (normal) , alkaline phosphatase - 610u/L(↑), parathyroid hormone(PTH) 132 p g /ml (↑) suggestive of rickets, however parents refused to perform vitamin D level which is a costly investigation.

X ray of wrist joints and lower limbs revealed features of both rickets and scurvy-

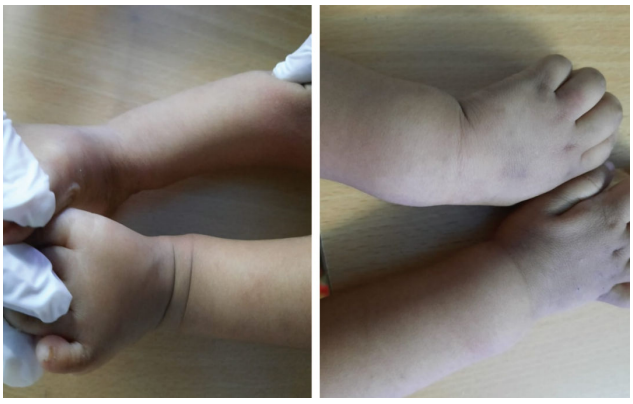

Fig.-1: *Widening of both wrists and ankles*

Fig. 2: Picture of the child

Rickets- Fraying, widening and cupping of metaphysis of lower ends of radius, ulna and femur and upper and lower ends of tibia and fibula (Fig-3,4)

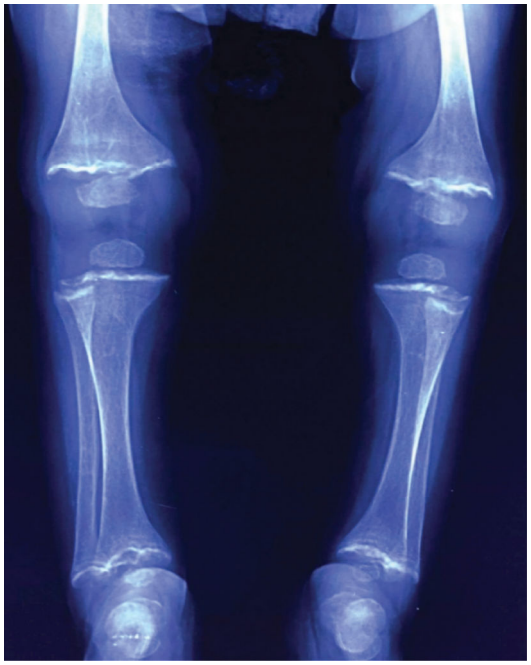

Fig 3. Rickets- Fraying, widening, cupping of metaphysis of long bones, Scurvy- White line of Frankel, scurvy line, wimberger ring sign of epiphysis and tarsal bones and osteopenia

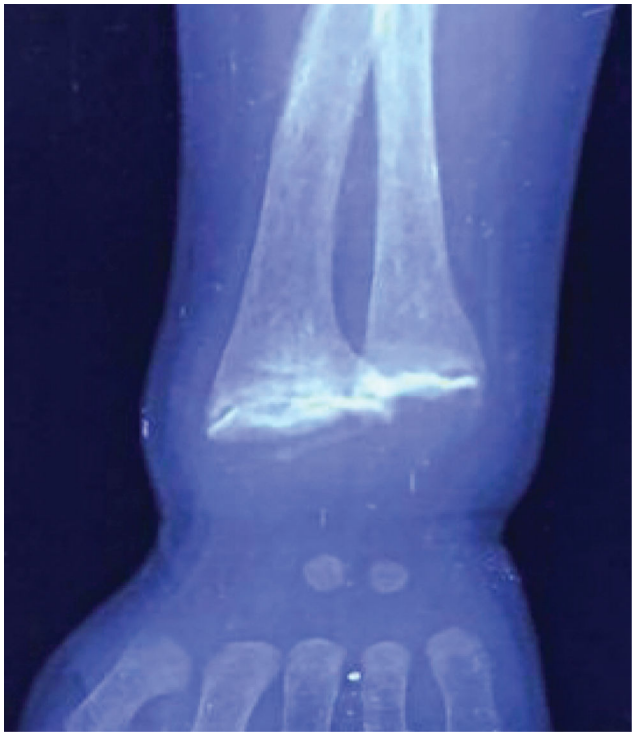

Fig 4. Rickets- Cupping, fraying and widening of radius and ulna, Scurvy- White line of Frankel, scurvy line

Scurvy- White line of Frankel, scurvy line, wimberger ring sign of epiphysis and tarsal bones also (just started), osteopenia(Fig-3,4)

CXR- patchy opacity in left mid zone (pneumonia)

To find out the cause of persistent pneumonia, we have excluded tuberculosis by mantoux test (MT test) and gastric lavage for acid fast bacilli (AFB) and GeneXpert and congenital heart disease by echocardiography ECHO). However her S. Antibody levels showed S.IgM-12.20 gm/l↑(range 0.40-1.43gm/l), IgA 1.21gm/l ↑(0.27-0.66 gm/l), S.IgE 117.70IU/L↑(<60.00 Iu/ml) and IgG 8.41gm/L (N).

To exclude other than nutritional rickets we have done liver function test, renal function test, s. electrolyte, arterial blood gas analysis. All the reports were within normal range.

Diagnosis:

Therefore, finally on the basis of clinical, biochemical and radiological parameters she was diagnosed as a case of rickets and scurvy with persistent pneumonia with failure to thrive.

Treatment:

After counselling she received oral vitamin D (4000IU) daily for 6 weeks with Calcium(250mg), vitamin C (1000 mg) daily , oral antibiotics and other supportive measures. When her respiratory symptoms settled down after 10 days, she was discharged with advice and requested to come for follow up after 1 month.

Discussion:

Rickets is an example of extreme vitamin D deficiency, with peak incidence in infancy.[7] Nutritional rickets is present more commonly in Africa, the Indian subcontinent, Asia, Latin America and the Middle East.[8]

Vitamin D is essential for absorption of calcium from the intestines. There are two well-known sources: exposure to sunlight and dietary intake, which accounts for less than 10%. Besides poor dietary intake and inadequate sunlight exposure, further increased risk is associated with dark-skinned individuals, solely breastfed infants and prematurity.[9] Most women in developing countries have vitamin D deficiency which is an important risk factor for congenital rickets .[10] Screening should be considered for children with poor growth/ development, seizure activity/tetany, and children with chronic malabsorptive states.[9]

On the other hand, scurvy, a disease of vitamin C deficiency, has been increasingly reported in recent years.[11] Musculoskeletal manifestations are present in 80% of patients with scurvy and are prominent in pediatric population.[12] Scurvy occurs in disadvantaged populations with poor intake of fresh fruit or vegetables. As mentioned earlier, co-occurrence of rickets and scurvy develops when the society has risk factors for both conditions. This baby was born at preterm with low birth weight and was on exclusive breast feeding without any multivitamin supplementation. She also had lack of exposure to sunlight and fresh fruit and vegetables.

The presentation of co-occurrence is highly variable, and co-occurrence is not associated with any pathognomonic features. Appearance of clinical features of both conditions are dependent on the order and length of first disease prior to the development of the 2nd illness.[13]

In rickets there is defective mineralization of osteoid tissue whereas scurvy produces pathological osteoid and thereby reduces osteoblastic activity but mineralization is unaffected. The majority of classic rachitic features require some normal osteoid tissue for remodeling (e.g., softening and bending of the limbs, flaring of the metaphysis) that is inhibited by the co-occurrence of scurvy. When rickets is dominant it could mask the classic scurvy feature of new bone formation due to poor mineralization, and reduce the clinical symptoms of pain and tenderness caused by

scurvy.[14] Our patient presented with poor growth and rickets seemed to be the dominant one as she had box shaped head, wide open anterior fontanelle , widening of the wrist and ankle joints and growth failure but none of the clinical features of scurvy(limb pain, gum hypertrophy ,etc) was present, but only few radiological changes of scurvy.

Like clinical presentation, bony features of either disease are not clearly visible unless one condition occurred first and had some period to develop bony changes prior to the appearance of the secondary disease.[13] In cases of co-occurrence, classic features associated with either severe rickets (e.g., bending) or severe scurvy (e.g., line of Fraenkel, scurvy line, corner signs, elevated periosteum) are often absent and 'masked' by the processes of counter action of either disease.[14]

In some cases, one of the two diseases will dominate over the other and mask the features of the non-dominant disease. Valentini et al. reported cases of dominant scurvy and where radiographs did not provide any evidence of concomitant rickets unless rickets was well developed.[15] Fouron and Chichoine presented four cases where rickets was the dominant disease. In these cases, radiological features of rickets were predominant and white dense line of Frankel and one example of a Wimberger's ring were present in scurvy cases. However, advanced features of neither disease were present, for example bending of rickets and periosteum elevation of scurvy. [14] Similarly, in this case fraying and widening of metaphysis of long bones, white line of frankel, scurvy line, wimbergers ring were present without advanced features of either disease as we know scurvy and rickets are always counteracting. Cases of scurvy and rickets co-occurrence have been mentioned in a number of recent articles.[16,6]

The diagnosis of any forms of vitamin D deficiency rickets is usually established by clinical, biochemical, and radiographic criteria.[17] In our case in addition to clinical and radiological criteria biochemical parameters such as high alkaline phosphatase and parathormone level with normal calcium and phosphate levels were also suggestive of nutritional rickets. A level of 25 hydroxy vitamin D (25-OH D) less than 12.5 nmol/L (5 ng/mL) is suggested for the diagnosis of rickets.[9] However, we could not perform vitamin D level for financial constraint of the parents.

Though serum Vitamin C concentration in the plasma is specific for the diagnosis of scurvy; vitamin C <11micro-mole/l suggests scurvy, actually it is not necessary to confirm the diagnosis as a meticulous dietary history, physical examination and radiological findings are enough to reach a correct diagnosis easily.[18] In case of co-occurrence, radiographs appear to be the most helpful. Radiographic methods can produce a high percentage of accuracy in identifying co-occurrence of rickets and scurvy.[19]

There are two strategies for administration of vitamin D. In stoss therapy, vitamin D(300,000-600,000IU) is administered orally or intramuscularly as 2-4 doses over one day. The alternative strategy is daily vitamin D with a minimum dose of 2000IU/ day for a minimum of 3 months. Either strategy should be followed by daily vitamin D intake of 400IU/day if < 1 year, or 600IU/day if >1 year along with adequate dietary calcium and phosphorus intake. Vitamin D3 is preferable to D2 because of longer half life. Hypocalcemia should also be treated with parenteral administration of calcium gluconate in case of manifest tetany or convulsions followed by oral calcium supplements.[7] In case of scurvy, supplementation with 100-200mg/day oral or parental vitamin C is the usual treatment. Clinical improvement is seen within 1 week in most cases, however treatment should be continued for up to 3 months.[20]

For adequate exposure to sunlight, a fully clothed child would have to spend two hours outside weekly and darker skinned individuals may require exposures up to 6–10 times this amount. Breast milk is the ideal nutrition for infant, however, it only contains 15 – 50 IU/L of vitamin D. So, American Academy of Pediatrics (AAP) has recommended vitamin D supplementation in all breastfed infants from the first day of life and continue through childhood/adolescence.[9]

Conclusion:

In the existing literature of Bangladesh, co-occurrence of scurvy and rickets has not been found being reported. Nutritional assessment is very crucial for every child. When children present with growth failure and developmental delay, rickets should be evaluated and coexistence of other nutrient deficiencies such as scurvy must be ruled out. Several risk factors are associated with co-occurrence. Education on proper nutrition during pregnancy, lactation, infancy and supplementation of vitamin D during pregnancy, lactation period, in preterm &low birth weight babies, in all exclusive breastfeeding babies and adequate exposure to sunlight necessary to prevent its growing resurgence. Proper childhood maintenance visits with growth and development screenings are critical for early detection of these easily treatable condition.

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