

Title: Scurvy : A case report and review of the literature

Short title: Scurvy

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Abstract:

Scurvy is a clinical manifestation caused by vitamin C deficiency. Musculoskeletal manifestations such as inability to walk (pseudoparalysis), arthralgia, myalgia, hemarthrosis, muscular hematomas, many subcutaneous hemorrhages, swollen and bleeding gums, perifollicular hemorrhage, petechial hemorrhage and massive subperiosteal hemorrhages of the humerus and femur are the classic symptoms. Scurvy can happen to any children of any age who have an inadequate intake of fresh fruits and vegetables. Although scurvy is rarely found, it still exists in Thailand. Objective: to report a case and review the literature of scurvy. The case presented is a 2 years and 5 months old boy with pain and inability to walk for 1 month. His dietary history revealed he had drunk only UHT soy milk and low intake of rice, fresh fruits and vegetables. The bilateral knee radiograph showed diffuse osteopenia, the thickened and dense zone of provisional calcification (“white line of scurvy”) and a prominent white line surrounding the internally rarefied epiphysis producing the Wimberger ring. These are the common characteristic findings of scurvy. The patient had very low serum ascorbic level at 0.076 mg/dL (reference 0.2-1.4 mg/dL). This test is done in order to confirm the diagnosis. Conclusion: the deficiency of vitamin C should be aware for all physicians because it can be cured easily, and has rapidly and noticeable response to treatments.

Keyword: scurvy, Wimberger’s sign, ascorbic acid, pseudoparalysis, UHT milk.

คัดย่อภาษาไทย

โรคลักปิดลักเปิดคือภาวะขาดวิตามินซี ซึ่งจะแสดงอาการได้แก่ ไม่ยอมเดิน อ่อนแรงที่ขาทั้งสองข้าง ปวดข้อ ปวดกล้ามเนื้อ มีเลือดออกในข้อ มีเลือดออกเหงือก เหงือกบวม เลือดออกใต้ผิวหนัง เป็นต้น ภาวะขาดวิตามินซีเกิดได้กับเด็กทุกอายุที่รับประทานอาหารไม่ถูกต้อง และรับประทานผัก ผลไม้ไม่เพียงพอ ซึ่งในประเทศไทยพบได้น้อยมาก แต่ก็พบได้ประปราย โดยจุดประสงค์คือ รายงานผู้ป่วยและบททวนวรรณกรรมที่มีภาวะขาดวิตามินซี ซึ่งเป็นเด็กผู้ป่วยชายอายุ 2 ปี 5 เดือนมาด้วยไม่ยอมเดินมา 1 เดือนโดยมีประวัติดื่มแต่นม ยู เอช ที นมถั่วเหลือง และรับประทานข้าว ผัก ผลไม้เล็กน้อย ได้ตรวจเอ็กซเรย์เข้าทั้งสองข้างพบ diffuse osteopenia, the thickened, dense zone of provisional calcification, Wimberger ring ซึ่งเป็นลักษณะพบบ่อยในโรคนี้ ผู้ป่วยได้ตรวจระดับวิตามินซีพบว่า มีระดับต่ำมาก 0.076 มิลลิกรัมต่อเดซิลิตร โดยสรุป ภาวะขาดวิตามินซีควรได้รับการระลึกไว้เสมอโดยแพทย์ที่ทำการรักษาเนื่องจากสามารถรักษาได้ง่ายและตอบสนองต่อการรักษาอย่างรวดเร็วและชัดเจน

คำสำคัญ: ขาดวิตามินซี, Wimberger's sign , วิตามินซี, อ่อนแรงไม่ใช่จากโรกระบบประสาท, นมยู เอช ที

Introduction:

Scurvy is a rare disease that's a constellation of clinical manifestation caused by vitamin C deficiency.<sup>1-3</sup> Vitamin C, also known as ascorbic acid, is necessary in hydroxylation reactions, including procollagen production. Risk factors for scurvy are poor nutrition, social isolation, poverty, and homelessness.<sup>1-5</sup> Musculoskeletal manifestations may be the presenting symptoms, consisting of inability to walk (pseudoparalysis), arthralgia, myalgia, hemarthrosis, and muscular hematomas.<sup>1,3,6,7</sup> Hemorrhagic symptoms have also been found including manifested through many subcutaneous hemorrhages, swollen and bleeding gums, perifollicular hemorrhage, petechial hemorrhage and massive subperiosteal hemorrhages of the humerus and femur.<sup>4,5,8-10</sup> Scurvy can affect children of any age but is seen mostly in preschool children who have an inadequate intake of fresh fruits and vegetables and lack of vitamin C.

Epidemiology of scurvy in Thailand had seen a few because there are plenty of tropical fresh fruits and vegetables available around the year. However, scurvy still occurs because of improper feeding. There are five review reports of scurvy in Thailand. The first report was a case in northern

Thailand in 1983<sup>11</sup>. Secondly, 6 pediatric patients from Thai J Parenteral Enteral Nutr in 1990<sup>12</sup> was reported. Thirdly, there were 21 pediatric patients, their aged between 14-42 months old, seen in 1995<sup>13</sup>. Forthly, there were 28 pediatric patients from 1995 to 2002 at Queen Sirikit National Institute of Child Health (QSNICH). The most common clinical presentation were involved with limp or inability to walk (96%).<sup>7</sup> Lastly, there was a case report of refutation to walk at Chiang Mai university in 2006.<sup>14</sup>

The purpose of this presentation is to demonstrate a typical case of scurvy, which is frequently forgotten. Moreover, it is to review the relevant literatures of this particular disorder of childhood that all physicians should be aware of.

Case report:

A 2 years 5 months old boy was admitted to the hospital with clinical symptoms of disordered limping and unable to walk independently for 1 month. Whenever he moved for example lying flat or the act of flexion of hips and knees, he always cried. He had no fever, no ataxia and no tremors. His mother took him to the provincial hospital. The attending physician treated him by weight bearing traction for a period of 1 week. He was treated as a case of osteomyelitis and septic arthritis. He was re-admitted in the same hospital when he had progressed to a complete refusal to walk and low-grade fever. The patient was then referred to Naresuan University Hospital. First, he was admitted to the orthopedic ward where he continued to receive treatment similar to the previous hospital. He was then sent to the pediatric ward. After carefully evaluated his dietary history, he had been feeding UHT soy milk (600 ml per day) since

he was 7 months old and had low intake of rice, fresh fruit, vegetables or meat. The patient's history of vaccination, growth and development was normal. On examination, his body temperature was 38 ° C, pulse 120 beats/min, respiratory rate 25 /min, BP 112/74 mmHg, present body weight was 11 Kg (P10), and length of 96 cm.(P 90). He had severe pain on passive range of motion of knees and on bilateral hip extension. However, there were no signs of effusion, swelling or redness. The patient had generalized weakness in the lower extremities. He was able to lift his hips and knees against gravity but cannot lift with a resistance. He refused to bear any weight. His deep tendon reflexes and sensation were intact. His skin and gingiva were unremarkable.

Laboratory studies showed complete blood count; Hct 38% WBC 13,560 cell /mm<sup>3</sup> , MCV 67.3 fL, MCH 23.2 pg, RDW 14.1%, Neutrophil 49%, Lymphocyte 46%, monocyte 5%, platelet 349,000 cell /mm<sup>3</sup> , no anisocytosis and poikilocytosis and elevated erythrocyte sedimentation rate of 40 mm/h (reference range, 0-20 mm/h) and C-reactive protein rate 32.9 mg/L (reference range, 0-3 mg/L).

Hemoculture showed no organism growth. Plain radiographs of the knees revealed thickened and dense zone of provisional calcification ("white line of scurvy") and a prominent white line surrounding the internally rarefied epiphysis producing the Wimberger's ring (Figure 1). The MRI findings of both hips are not well described. A differential diagnosis of nutritional deficiency specifically for vitamin C deficiency was carried out. The patient's serum vitamin C level showed 0.076 mg/dL (reference 0.2- 1.4 mg/dL) by HPLC method. He also had latent iron deficiency anemia. He was then started on vitamin C 300 mg/day and an iron supplement (5 mg/kg/day). He was discharged 1 week later with alleviation of leg pain but he still refused to walk. One month after discharge, he began to walk and run. Follow up film legs at 4 month and 8 month,

respectively after treatment show improvement of osteopenia, reduction in metaphyseal lucencies and disappearance of white line of scurvy (Figures 2 A.B). He has begun chewing vegetables and fresh fruits, as well as drinking cow's milk everyday.

#### Discussion:

Scurvy or vitamin C deficiency is rare in all ages, especially in developed countries. Scurvy develops from improper nutrition. This patient's diet was devoid of fresh fruits e.g. orange, grape, guava, papaya, vegetables, juices, or cow's milk. The patient drank an ample amount of UHT soy milk since he was 7 months old which caused him nutrition insufficiency, particularly vitamin C. Vitamin C is a water-soluble compound vulnerable to heat, ultraviolet radiation, and oxygen. The total pool in the body is 1,500-2,500 mg and the daily turnover is 45-60 mg. Vitamin C has a half-life of 10-20 days.

Absorption of vitamin C occurs in the ileum via an active transport mechanism that becomes saturated when the oral intake exceeds 180 mg/day.<sup>1</sup> More than 90% of vitamin C is derived from fruits and vegetables, including potatoes, tomatoes, berries, green vegetables, and citrus fruits<sup>10</sup>. In general, ordinary cooking of these items reduces their vitamin C content by 20% to 40%. Using little water or employing a microwave oven will decrease these losses. This patient had scurvy. The symptoms of scurvy appeared between at least 29 to 90 days after deprivation of vitamin C.<sup>5</sup> A prominent feature is an inability to walk. It's the most common presentation (96%)<sup>7</sup> in a retrospective study of 28 children, 2003. But no had other clinical manifestations in this case. The other clinical manifestation would be tissue bleeding with nonpalpable purpura,<sup>9</sup> petechial

hemorrhage, spontaneous ecchymoses, gum changes, hyperkeratosis, and coiled hairs. A low plasma level of vitamin C is diagnostic of scurvy, (plasma ascorbate concentration of < 0.2 mg/dl usually is considered deficient) which was seen in this case. However, this plasma vitamin C level was taken 8 days after being admitted at this hospital. The plasma vitamin C level seen in this case was not really accurate because he may have had a recent intake of food with vitamin C content. The levels also may be normal if there has been recent vitamin C supplementation in any form. Thus, determination of plasma vitamin C levels remains an insensitive laboratory test for vitamin C deficiency. In practice, the diagnosis of scurvy is based on a combination of clinical and radiographic findings.<sup>15,16</sup> Scurvy can have a differential diagnosis when a dietary history indicates insufficient intake of vitamin C for at least 1-3 months and related clinical signs and symptoms are present. A differential diagnosis of scurvy is shown in this case depending on the clinical presentation such as osteomyelitis, septic arthritis, transverse myelitis and cord compression. However this patient did not have any neurological sign involving. Also, he did not have any leukocytosis, elevated erythrocyte sedimentation rate and C-reactive protein presented which is similar to the review in Thailand.<sup>11-14</sup> The diagnosis can be missed easily as the disease presents vague symptoms in the initial stages and cases are relatively rare. To prevent scurvy, the daily intake of vitamin C must be no lower than 10 mg/day and the body pool not below 350 mg. The recommended daily intake of vitamin C in children varies with age (Table 1).<sup>17</sup> Scurvy is treated immediately by oral vitamin C and orange juice. Observing that a daily dose of vitamin C 300 mg is given is necessary until full recovery of clinical signs and symptoms.<sup>8,18</sup> One month after treatment, he looked well and normal walking and running was observed. Although,

symptoms of scurvy have improved, this child must continue to drink orange juice and eat, rice, fruits and vegetables. There are certain foods that contain vitamin C, e.g, guava, malacca, papaya, orange and ivy gourd. The result of vitamin C treatment is dramatic with gradual improvement of all physical signs in a few days. Subjective improvement commonly begins within 24 hours of treatment, and lethargy, anorexia, and pain diminish in 2 to 3 days. Joint swelling resolves in a few days. The purplish hue of the skin lesions pales quickly and then subsides in 2 to 4 weeks, leaving areas of brown pigmentation that slowly disappear. The hyperkeratosis decreases in 1 to 2 weeks, and by 4 weeks the hairs have uncurled, returning to their normal appearance. The gums change from purple to red in 1 to 2 weeks, with more gradual resolution of the gingival edema, complete recovery being apparent by 3 months.<sup>10</sup> In this case, symptoms after treatment resolved in about one month. He is walking and running very well.

Conclusion:

Scurvy, a rare and always forgotten disease, can be found in developed countries.<sup>18</sup> Other risk factors include chronic and wasting diseases, intentional dietary restrictions, and lack of education about dietary needs. Acute treatment of scurvy followed by appropriate dietary adjustments or supplementation carries an excellent prognosis. Patients who have progressed rapidly resolve to normal condition and good health with appropriate treatments. All physicians should be aware of the clinical presentations of vitamin C deficiency, because the presentation of the patient with scurvy may be subtle. Thus, scurvy is an easily treatable condition.

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Table 1 Recommended Dietary Intake of Vitamin C (mg/day) in Children Based on Age Group<sup>3</sup>

Age	Vitamin C (mg/day)
Infants: 0-6 mo.	40
6-12 mo.	50
Children: 1-3 y	15
4-8 y	25
9-13 y	45
14-18 y: male	75
Female	65

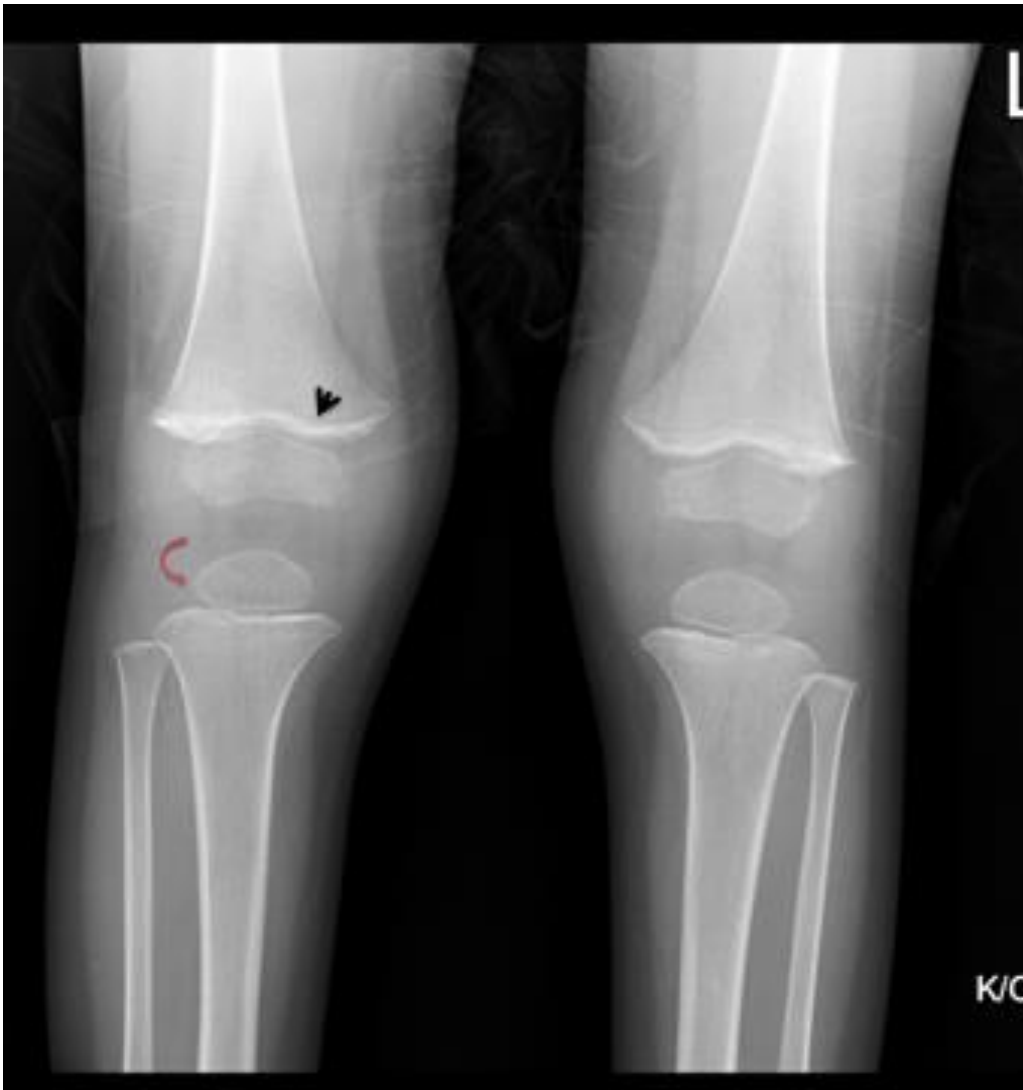


Figure 1

Figure legends:

Figure 1: Initial AP plain radiography of both knees showed diffuse osteopenia with the dense and thickened zone of provisional calcification “or white line of scurvy” (black arrow) and the white line surrounding the internally rarefied epiphysis producing the Wimberger’s ring (curved arrow)

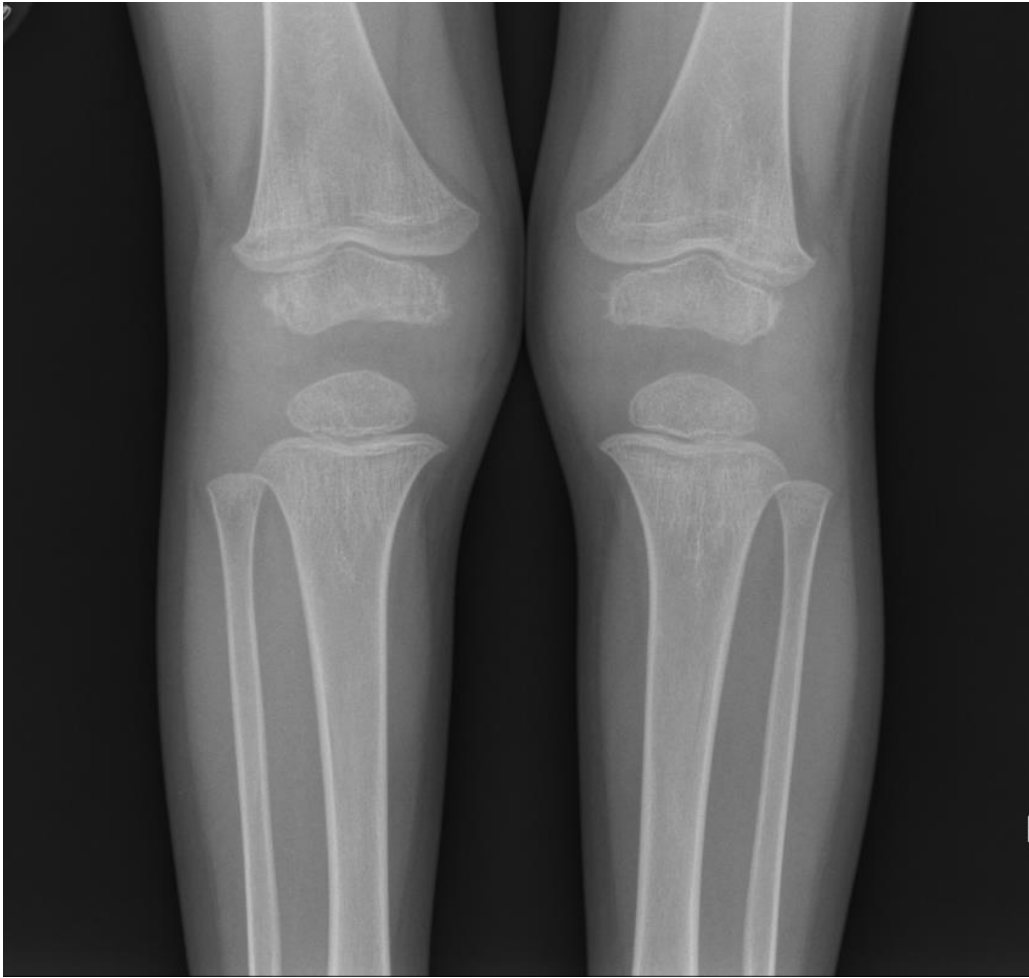


Figure 2 A



Figure B

Figure legends:

Figure 2: Follow up films at 4- months (A) and 8-months (B) after treatment show improvement of osteopenia, reduction in metaphyseal lucencies and disappearance of the white line of scurvy.