

# **SCURVY IN A BOY WITH KNEE PAIN AND A LIMPING GAIT: A CASE REPORT**

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## **ABSTRACT:**

Scurvy is a disease caused by chronic vitamin C deficiency which has become uncommon in the modern age. Several cases have been reported in children with restricted dietary habits with neurodevelopmental issues. It is often overlooked as an initial diagnosis in view of its vague presentations mimicking other systemic diseases which lead to extensive investigations and delay in diagnosis. A previously healthy 9-year-old boy with left knee pain and a limping gait presented to a local health clinic. The child subsequently developed worsening bilateral knee pain and was unable to walk without support. His restricted diet and radiographic findings were suggestive of scurvy which was confirmed by the extremely low levels of ascorbic acid. Treatment with ascorbic acid replenishment and maintenance resulted in remarkable improvement of his symptoms. This case highlights the importance of a high index of suspicion in at-risk children so as to avoid unnecessary invasive investigations and procedures.

**Keywords:** Scurvy, vitamin C, ascorbic acid, limping gait, knee pain

## **INTRODUCTION:**

Scurvy is a vitamin C deficiency which has existed in ancient times but has become very rare especially in the modern developed countries. In Malaysia, nutritional deficiency such as scurvy is relatively uncommon and least expected as the country has the highest prevalence of obesity in South-East Asia followed by Brunei and Thailand, according to the World Population Review 2019(1). Scurvy is even more uncommon in the pediatric population without any underlying medical conditions(2). However, it has been reported that children with developmental and behavioral abnormalities such as Autism Spectrum Disorder tend to be at higher risk of developing scurvy due to their food selectivity(3). The diagnosis of scurvy is often overlooked as its clinical features may mimic other systemic diseases such as rheumatological disorders in Juvenile Idiopathic Arthritis or Systemic Lupus Erythematosus, infectious diseases, haematological malignancy such as acute lymphoblastic leukaemia, soft tissue or bone tumors(4). This would lead to extensive laboratory and radiographic investigations in order to rule out other common diseases. Therefore, we aim to highlight the importance of recognizing vitamin C deficiency as one of causes of limping gait and bony tenderness especially in paediatric population.

## **CASE REPORT:**

A previously healthy 9-year-old boy presented to government health clinic with two weeks history of left knee pain and a limping gait. He denied any history of fever, sore throat, fall or trauma. He was admitted to the nearest district hospital for further evaluation and management. He was seen by visiting Orthopedics team and referred to physiotherapy. He was put on a left above knee back slab for one week and underwent a physiotherapy session but there was no improvement. After one week, he developed right knee pain and unable to ambulate at all. His bilateral knee pain worsened over four weeks period until he was unable to get out of bed or walk without assistance. Subsequently, he was referred to a tertiary hospital and was admitted to Paediatric ward for further management.

On examination, he was alert with normal vital signs. He had no apparent joint swelling or joint tenderness, but there was pain with motion of his knees and hips. Both knees were in flexed position and he was unable to fully extend both knees due to pain and stiffness. Muscle wasting of bilateral hamstrings was noted slight reduction of muscle power of both lower limbs. Both knees were not erythematous and there were no

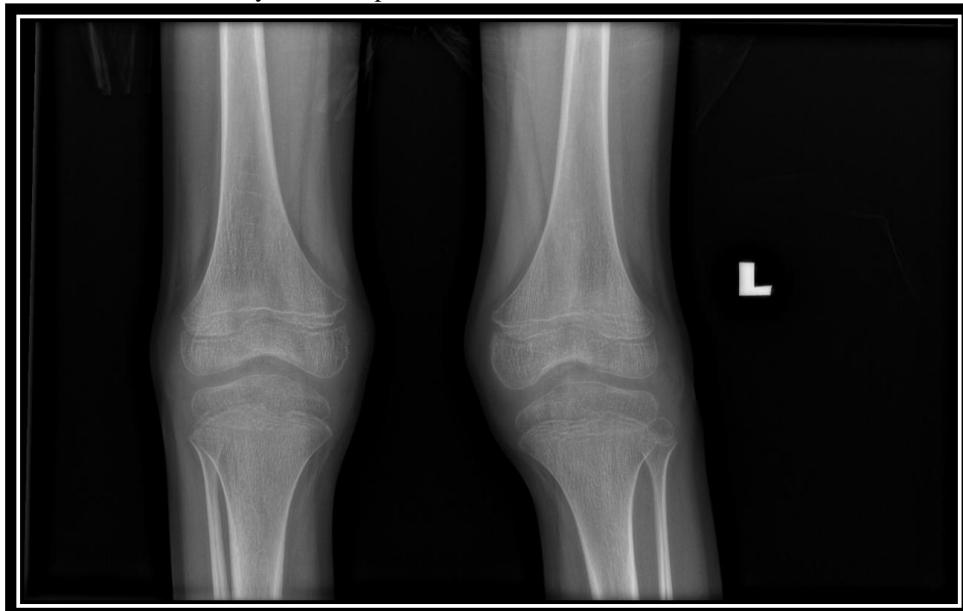
surrounding skin changes. Scattered perifollicular purpuric macules were noted on his lower extremities. He had normal abdominal examination results, without organomegaly. The remainder of his neurologic examination results were normal.

He was born via spontaneous vaginal delivery at term with uneventful perinatal period. He completed his vaccinations and parents claimed his developmental milestones were normal and appropriate with his age. There was no history of autoimmune disease, rheumatological diseases, hematological disorders, or malignancy in the family.

The initial differential diagnosis was autoimmune diseases such as Juvenile Idiopathic Arthritis (JIA) and Systemic Lupus Erythematosus (SLE), rheumatic fever, infective cause and hematological malignancy especially leukemia. Evaluation included a total white cell count of  $6.4 \times 10^9/L$  with a normal differential count, a platelet count of  $379 \times 10^9/L$ . He has normal C-reactive protein of 2.84mg/dL. Antinuclear Antibody (ANA), and Anti-Double Stranded DNA were negative. However, his erythrocyte sedimentation rate was elevated with a reading of 56 mm/h. Complements including C3 and C4 were in normal range. Infective causes screening including Antistreptolysin O (ASO) titer and mycoplasma antibodies were negative. He had hypochromic microcytic anemia, with a hemoglobin level of 9.2g/dL, hematocrit level of 0.28, and mean corpuscular volume of 77 fL. The diagnosis of iron deficiency anemia was made based on her iron studies which showed low serum iron of 4.7, serum TIBC of 56.7 with T-saturation of only 8.3%. Xray of bilateral femur, tibia and fibula showed no obvious fractures seen but subtle osteopenia changes. MRI whole spine and lower limb showed no significant abnormalities detected.

Since the investigations had ruled out most of possible autoimmune diseases, infective causes and hematological disorders, the alternative diagnosis of scurvy was considered. A detailed review of the boy's dietary history revealed severely limited variety of food since he started weaning. He was only breastfed until the age of two months and continued with formula milk feeding until the age of five. His parents claimed that the patient has been a picky eater since the age of two. He refused to eat all fruits and vegetables. Since the age of five, he only preferred to eat rice together with fried egg and soy sauce until the age of nine. Further history revealed that he had one episode of gum swelling and bleeding at home noted by his parent but the bleeding resolved spontaneously and did not cause any pain to patient. He was referred to Dental team and the diagnosis of chronic gingivitis and dental caries were made. Serum ascorbic acid level was taken and showed less than 0.1mg/dL (reference range 0.4-2.0 mg/dL).

The laboratory and clinical findings supported the diagnosis of scurvy. He was started on oral ascorbic acid 100mg three times daily and syrup multivitamin 2.5mls daily. He has been managed by multidisciplinary teams besides Paediatrics including Rheumatology, Orthopedics, Rehabilitation, Physiotherapy, Occupational therapy and Dietician. After 4 weeks of ascorbic acid supplementations with regular physiotherapy and rehabilitation, his pain improved rapidly, and he began to walk normally again. After 6 weeks of admission, he was discharge from the hospital with oral ascorbic acid 100mg twice daily and syrup multivitamin 2.5mls daily. Complete resolution of his pain occurred within 2 weeks, and his skin lesions faded simultaneously. With behavioral modification, his dietary intake improved, and he remained well.



**Figure 1:** Radiograph of bilateral knee joint showing subtle generalized osteopenia

**DISCUSSIONS:**

Vitamin C or L-ascorbic acid is an essential micronutrient for the maintenance of intercellular connective tissues, dentine, osteoid and collagen formation in humans. Vitamin C performs a large number of physiologic functions include assisting with iron absorption, synthesis

of carnitine and norepinephrine, and a number of antioxidant properties(5). Vitamin C is a water-soluble vitamin that is fully absorbed from the gastrointestinal tract and distributed in both intracellular and extracellular tissues. Any excess of vitamin C is excreted in the urine. Vitamin C content in human milk is higher than in cow's milk(4). Citrus fruits and vegetables are the best sources of vitamin C. Cooking, storage, and oxidation will reduce the vitamin C content of many foods. The daily intake of vitamin C has an impact on plasma concentration. As in this case, the child was only breastfed for short duration of two months old and refused to eat any fruits or vegetables which causing the vitamin C deficiency.

The earliest clinical presentations of scurvy can be nonspecific constitutional symptoms such as fatigue, anorexia, and weight loss. Scurvy can present with musculoskeletal symptoms, which include arthralgia, myalgia, hemarthrosis, and muscular hematomas in 80% of the cases(6). In our case, the child presented with a limping gait and inability to walk normally which is a very common presenting musculoskeletal symptom in children with scurvy. Pain in lower limbs particularly over the distal end of the femur and tibia may be caused by subperiosteal hematomas(7). Gingival hyperplasia, corkscrew hairs, and weakened blood vessel walls, leading to bleeding in the skin, joints, and other organs are due to poorly formed collagen in vitamin C deficiency(7). Common dermatologic findings are follicular hyperkeratosis and perifollicular hemorrhages especially over the lower limbs(8). These explained the chronic gingivitis and scattered perifollicular purpuric macules noted on the child's lower extremities.

Anemia is the most common hematologic manifestation of scurvy and seen in up to 80% of the cases(9). Reduced iron absorption and concurrent vitamin deficiencies especially folate deficiency are believed to be the reason of anemia in patients with scurvy. Ascorbic acid promotes iron absorption by reducing it from the ferric (Fe+3) to ferrous (Fe+2) state which is more absorbable in the body.(9). Therefore, concomitant micronutrient deficiencies, including levels of serum iron, folate, and vitamin B12 should be ruled out. In this case, hypochromic microcytic anemia and low serum iron were seen in this patient.

Furthermore, our patient also presented with an elevated erythrocyte sedimentation rate (ESR) which is commonly seen in many patients presenting with vitamin C deficiency, making it difficult to differentiate scurvy from other rheumatologic disorders (5). There is a possible relationship between the role of vitamin C in the inflammatory process. In an animal study conducted by Horio and coworkers, it was noted that ascorbic acid deficiency leads to increased levels of interleukin-6, haptoglobin,  $\alpha$ -1-glycoprotein, and cytokine-induced neutrophil chemoattractant-1 which are seen in an acute inflammatory state (6). However, the physiologic role of these findings is unclear.

Osteonecrosis, osteopenia, and cortical thinning with periosteal proliferation are all possible radiological findings in scurvy. Some radiological signs such as The Frankel sign (zone of calcification at the margin of the growth plate), Wimberger sign (calcification around the epiphysis), and scurvy line (lucency adjacent to metaphyseal sclerotic line) were seen in other studies(7)(10). However, all those signs were absent in our patient as his radiograph of bilateral knee joints only showing subtle generalized osteopenia. This could be due to a good level of vitamin D that might prevent these changes.

Scurvy may be diagnosed with a thorough medical history, including a dietary history and a physical examination. Workup is usually not needed to confirm a diagnosis of scurvy. At least 10 mg/day is needed as a daily intake of vitamin C in order to prevent scurvy(11). The recommended daily intake varies depending on the age of the child. The recommended daily allowance (RDA) of vitamin C is 15-45 mg for age 1 to 13 years and 65-75 mg for age 14-18years (6). There is higher requirement for vitamin C during infections or inflammatory states, pregnancy and lactation (6). There is no specific regimen in the treatment of scurvy. The aim of treatment is to replenish the level of vitamin C to improve the symptoms. The usual treatment is 1 g/day of oral vitamin C supplementation for 2 weeks or 100 to 200 mg/day for a longer duration(2)(6). Spontaneous bleeding, oral symptoms, and constitutional symptoms may improve within few days of receiving vitamin C treatment, while bony changes and ecchymoses can take several weeks to resolve(9).The improvement of symptoms after ascorbic acid administration is a confirmatory approach to the diagnosis of scurvy.

**CONCLUSIONS:**

Scurvy has a variety of clinical manifestations that may overlap other common childhood illnesses. Therefore, healthcare providers especially the primary care physicians who commonly encounter the patient first at the clinic should have a high index of suspicion of scurvy. A thorough clinical history including a detailed dietary history is required especially in a child presented with musculoskeletal complaints. The diagnosis of scurvy and the initiation of appropriate treatment should be made promptly by a suggestive clinical history supported by radiographic evidences and laboratory investigations. Vitamin C supplementation in children with dietary selectivity can prevent complications related to scurvy. Even in this modern era, awareness of vitamin C

deficiency in children who present to clinic with musculoskeletal pain is necessary to avoid invasive and unnecessary investigations.

**CONFLICTS OF INTEREST:**

All authors have none to declare any conflict of interest.

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