JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

SCURVY WITH MUSCULOSKELETAL PAIN IN INDONESIAN PEDIATRIC PATIENT: A CASE REPORT

Hilmi Muhammad[1], Meirizal[2], Yudha Mathan Sakti[2], Aditya Fuad Robby Triangga[2],
Zikrina Abiyanti Lanodiyou[2], Ginanjar Reza Putra[2], Yasser [2], Yuni Artha Prabowo[2]
[1]Pediatric Sub-Division, Orthopaedic and Traumatology Division, Department of Surgery, Dr. Sardjito General Hospital, Faculty of Medicine, Public Health and Nursing, Gadjah Mada University, Yogyakarta, Indonesia
[2]Orthopaedic and Traumatology Division, Department of Surgery, Dr. Sardjito General Hospital, Faculty of Medicine, Public Health and Nursing, Gadjah Mada University, Yogyakarta, Indonesia
Corresponding Author: E-mail: hilmimuhammadortho@gmail.com

ABSTRACT

Background: Scurvy is a potentially fatal disease caused by a severe deficiency of vitamin C, which can manifest as a musculoskeletal problem. Early diagnosis and treatment of vitamin C deficiency are crucial to prevent severe symptoms. Unfortunately, vitamin C level evaluations in Indonesia are very limited due to the lack of laboratory facilities. As a result, patients with scurvy are often underdiagnosed and do not receive adequate management. **Case Presentation:** 5-year-old boy with musculoskeletal pain and inability to walk. Radiographic findings showed a pathognomonic line of Fränkel, Trümmerfeld zone, and Wimberger ring sign, with laboratory evaluation showing an anemia and vitamin D deficiency. We did not perform a laboratory examination for vitamin C due to a lack of test facilities but the patient was given oral 250 mg of vitamin C daily instead. The patient showed significant improvement in symptoms and laboratory results after three months of vitamin C supplementation. To summarize, that vitamin C deficiency should be considered among patients with related musculoskeletal manifestations supported by imaging results.

Keywords: *Bone Pain, Musculoskeletal Pain, Pediatric, Scurvy, Vitamin C*

INTRODUCTION

Scurvy is a rare disorder caused by severe deficiency of vitamin C leading to life-threatening manifestations.[1] Vitamin C is an antioxidant and a cofactor for numerous enzymes and it plays a crucial role in many fundamental processes, such as collagen synthesis, wound healing, bone formation, and the biosynthesis of catecholamines.[2,3] In collagen synthesis, vitamin C is needed as a cofactor in the process of procollagen maturation. Its deficiency causes increased fragility of various tissues. The described mechanism is responsible for the classical signs of scurvy, such as an increased risk of bruising and bleeding, impaired wound healing, corkscrew hairs, gingival abnormalities, asthenia, and anemia.[3]

The primary source of vitamin C is daily dietary intake, making inadequate nutrition the main cause of vitamin C deficiency.[5] Some risk factors of vitamin C deficiency include dietary habits, type 1 diabetes, malabsorptive disorders, and physiological disorders which could cause significant dietary restrictions.[2,4,5] Studies reporting scurvy's prevalence are still very limited. Several studies have been carried out only to evaluate vitamin C deficiency, especially in high-income countries in Asia, but most of them were only limited to the adult population. In Japan, they showed a relatively low prevalence of deficiency of 2.4%, while Singapore was higher at 12%. Low social status limiting access to high vitamin C foods has been used to explain the presence of scurvy in lower-middle income countries. Indonesia showed a high prevalence of deficiency with 45% of the population experiencing hypovitaminosis C.[4]

Bone disease is a common presentation in pediatrics with vitamin C deficiency, which may be related to a deficiency in osteoid matrix development and cartilage reabsorption.[8] This defective matrix formation is responsible for both limb pain and the typical radiological findings.[3] Radiologic examination may show osteopenia, cortical thinning with periosteal proliferation, an irregular and thickened white line at the metaphysis that indicates a calcification of the cartilage matrix (white line of Fränkel), a zone of rarefaction beneath the Fränkel line that indicate subperiosteal hemorrhage (Trümmerfeld zone), a white line that surrounds epiphysis ossification nucleus (Wimberger ring sign), and metaphyseal spurs (Pelkan sign).[6,8]

Evaluating vitamin C levels is crucial for both diagnosing the condition and monitoring the disease

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

course. Meanwhile, laboratory tests could only show nonspecific abnormalities that may suggest the presence of malnutrition, including anemia, iron deficiency, folic acid deficiency, and low levels of cholesterol and albumin, a detailed physical and radiology examination is needed.[6,7] Treatment for scurvy is vitamin C supplementation. The recommendation for children is 100-300 mg of vitamin C daily for 1 month which can be continued until full improvement of symptoms.[10] A full recovery is expected after approximately 3 months of regular vitamin C supplementation.[9,10]

Currently, in clinical settings in Indonesia, laboratory facilities to measure vitamin C levels are unavailable. As a result, scurvy is often underdiagnosed, and patients do not receive adequate management, such as vitamin C supplementation. This study reports the successful vitamin C supplementation in patients diagnosed without specific laboratory examination of vitamin C levels. This patient was diagnosed with scurvy based on history taking, physical examination, and specific findings from radiological examinations.

CASE PRESENTATION

A 5-year-old Indonesian boy was referred to our orthopedic division due to his inability to walk over the past three weeks with no reported history of trauma. No history of the same condition in his family. He was unable to support his body weight and exhibited lower limb pain. He also has knee pain especially when his knees were passively flexed. Initially, the pain disabled him from walking, limiting him to crawling. The pain eventually worsened and made him bed-bound. He had no other symptoms, such as fevers, rashes, rhinorrhea, cough, sore throat, vomiting, or diarrhea. The patient was a "picky eater" though his growth was within normal and achieved developmental milestones. Thorough history taking revealed no dietary source of vitamin C and only very limited amounts of iron and vitamin D-rich food.

The musculoskeletal examination showed some discomfort upon palpation of both lower extremities. The patient showed restricted mobility in both knee joints, resulting in his inability to walk. Upon examining the oral cavity, he had swollen and bleeding gums. Initially, the patient was suspected of osteomyelitis, but after initial treatment further evaluation of physical examination, and radiological evaluation the results were normal. From laboratory workup, we found microcytic anemia with a hemoglobin level of 3.8 g/dL. The patient received packed red cell transfusion upon admission and the hemoglobin level improved to 8.8 g/dL. The patients also showed a marked increase in platelet count at level 674 10^3/µL and CRP Protein level of 8.11 mg/L. after 3 months of treatment, laboratory workup shows a platelet count increase in 729 10^3/µL and improvement with CRP Protein level at the level 2.74 mg/L. Patient's laboratory work-up before and after treatment are shown in **Table 1.**

The hip and knee plain radiographs showed lytic and sclerotic lesions (scorbutic zone and dense zone), sclerotic epiphyseal line (Ivory epiphyseal), and sclerotic lesion with lucent area on metaphysis of at both femoral neck, distal femur and proximal tibia **(Fig. 1).**

Correction for Vitamin C and Vitamin D deficiency was initiated at 250 mg Ascorbic Acid and 0.5 mcg Calcitriol given daily. Monitoring was performed monthly. On his third-month follow-up, we repeated the hip and knee x-rays to evaluate the course of the disease and found an improvement in his condition **(Figure 2).** These findings were backed up by a gradual improvement in his overall condition, which included a decrease in bone discomfort, weight gain, and the restoration of his ability to walk. **(Figure 3).** The patient's timeline is shown in **(Figure 4).**

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

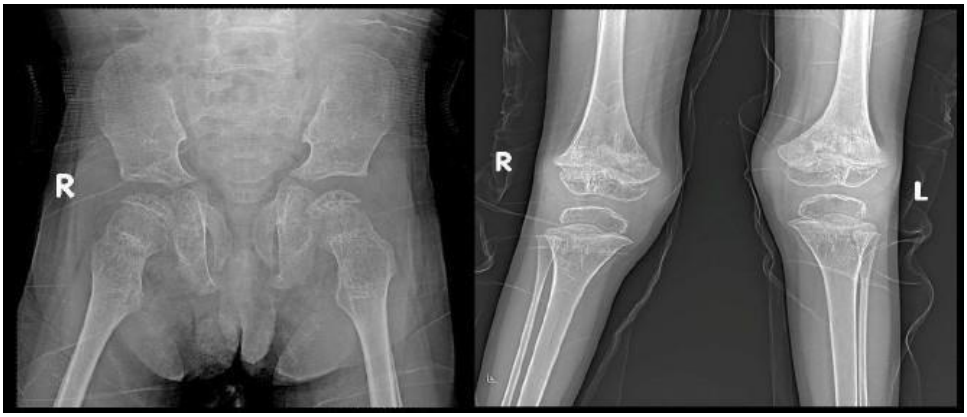

Figure 1. *Pelvic and bilateral knee X-rays show characteristics of the Scurvy Wimberger ring Sign, Frankel's line, and Trümmerfeld zone*

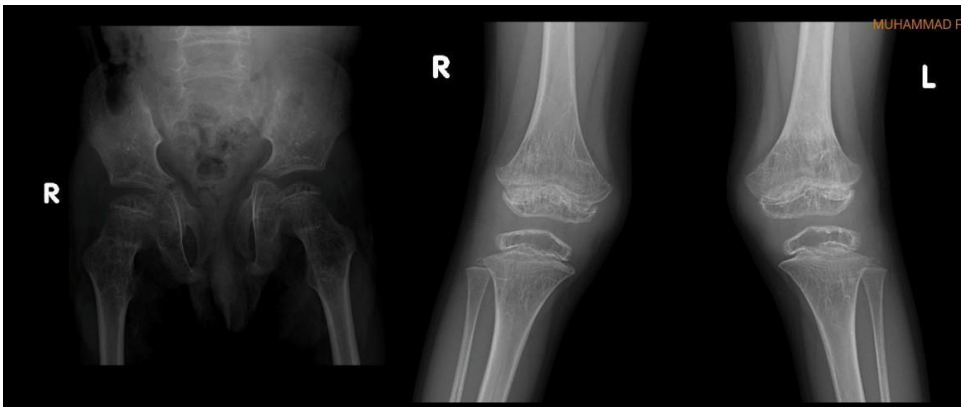

Figure 2. *Pelvic and bilateral knee x-rays after 3 months show improvement*

Figure 3. *Clinical Presentation after 3 months of treatment patients can stand and walk without pain.*

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

Hilmi Muhammad, Meirizal, Yudha Mathan Sakti, Aditya Fuad Robby Triangga
Zikrina Abiyanti Lanodiyou, Finanjar Reza Putra, Yasser, Yuni Artha Prabowo

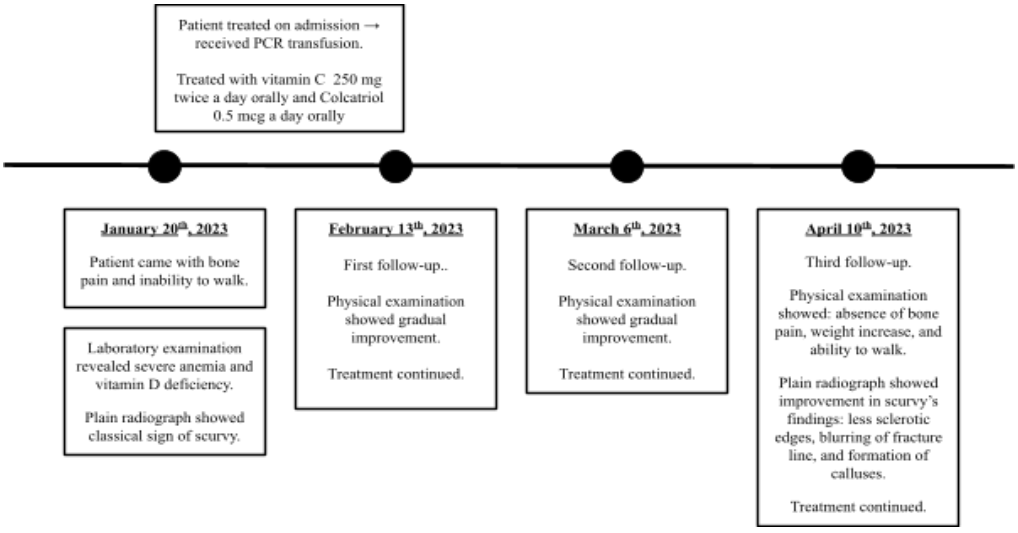

Figure 4. *Patient's timeline*

Table 1. *Patient's laboratory examination before and after treatment. Values which considered abnormal are marked with an asterisk (*).*

Laboratory test	Value	
	Before treatment	**After 3 months of treatment**
Hemoglobin	3.8 g/dL*	**8.8 g/dL**
Platelet count	674 10^3/µL*	**729 10^3/µL***
White blood cell count	8.7 10^3/µL	**6.7 10^3/µL**
C reactive protein	**8.11 mg/L***	**2.74 mg/L***

DISCUSSION

Vitamin C is required for the human body's basic functions, particularly the maintenance of intercellular connective tissue and the formation of collagen.[10] Although the actual prevalence of scurvy is unknown, some groups, particularly those with mental, neurological, physical, or developmental problems such as autism, are at a higher risk.[9]

Musculoskeletal symptoms, such as arthralgia, myalgia, hemarthrosis, and muscle hematomas, are observed in 80% of scorbutic cases and are more common in pediatric scurvy.[6,7] Pain and swelling related to subperiosteal hemorrhages and hemarthrosis frequently occur as a result of synovial blood vessel injury and microfractures.[2] Patients tend to show a limping gait, a limited range of motion, and an inability to stand or walk.[6,] In our case, the inability to walk is among youths' most prevalent musculoskeletal signs of scurvy.

A meticulous history taking, including dietary history and physical examination, can aid in confirming the diagnosis of scurvy.[6,8] Our patient's hips and knees plain radiographs showed a sclerotic lesion, consisting of scorbutic zone and dense zone, sclerotic epiphyseal line at metaphysis, Wimberger ring sign, lines of Fränkel, and Trümmerfeld zone which are all common in scurvy.

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

Laboratory findings in scurvy are nonspecific. Anemia, iron deficiency, hypoalbuminemia, and other vitamin deficiencies may be present in scurvy.[2,6,7] As in our case, the patient has vitamin D deficiency and severe anemia which required a blood transfusion. The causes of anemia in scurvy are numerous but mostly are iron deficiency due to decreased iron absorption caused by vitamin C deficiency. Other than that, the increased likelihood of spontaneous bleeding and concomitant vitamin deficiencies also contribute to the anemia seen. Assessing vitamin C levels is necessary when dealing with scurvy disease. A low plasma ascorbate concentration of less than 0.2 mg/dL indicates a vitamin C insufficiency.[7] Laboratory evaluation for vitamin C is not commonly accessible in Indonesia, limiting the diagnostic process.

Treatment of scurvy is based on replenishing the level of vitamin C needed, although there is no standardized supplementation regimen and the need varies based on the age of the child. Multiple treatment regimens have been advocated. The typical regimen involves supplementation of 1 g/day oral vitamin C for 2 weeks. Other studies found that daily vitamin C supplementation of between 100 and 200 mg might be given for a longer period.[6] They usually administered 100- 300 mg/day of vitamin C orally or 1 g IV for a week, followed by 100 mg daily for 1 to 3 months for infants and toddlers.[7] The key evidence confirming the diagnosis of scurvy is the alleviation of symptoms following treatment.[8]

In our case, we administered a three-month course of oral vitamin C supplementation at 250 mg/day. Monthly monitoring revealed symptom relief throughout the treatment period. By the end of three months, the patient experienced significant improvement, including the resolution of bone pain, weight gain, and a return to normal gait. To prevent recurrences and address any underlying issues, it is crucial to ensure the patient continues to meet their daily vitamin C requirement even after completing the treatment. At the six-month follow-up, the patient showed no signs of recurrence, because of ongoing vitamin C supplementation.[6]

From this case, we are confident that the combination of musculoskeletal manifestations and specific findings of scurvy from radiological examinations is valuable for diagnosing scurvy, even without specific serum vitamin C measurements.

CONCLUSION

From our case, we conclude that patients with specific clinical and radiological findings of scurvy can benefit from vitamin C supplementation, which has shown significant clinical improvement.

ETHICAL APPROVAL AND CONSENT

This publication was approved by the Ethical Committee Faculty of Medicine, Nursing and Public Health Universitas Gadjah Mada.

CONSENT FOR PUBLICATIONS

Written informed consent was obtained from the patient's parents for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

AVAILABILITY OF SUPPORTING DATA

Supporting data will be available upon reasonable request

CONFLICT OF INTEREST

The authors declare no conflict of interest

FUNDING

This research received no external funding

AUTHOR CONTRIBUTIONS

Conceptualization: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Validation: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Formal Analysis: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Investigation: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Resources: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Data Curation: H.M., M.M., Y.M.S, A.F.H.; Writing-original draft preparation: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Writing-review and editing: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Visualization: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Supervision: H.M., M.M., Y.M.S, A.F.H., Z.A.L., Y.A.P., G.R.P., Y.Y; Project Administration: H.M., M.M., Y.M.

JURNAL KEDOKTERAN DIPONEGORO
(*DIPONEGORO MEDICAL JOURNAL*)
Online : http://ejournal3.undip.ac.id/index.php/medico
E-ISSN : 2540-8844
DOI : 10.14710/dmj.v13i5.44840
JKD (DMJ), Volume 13, Number 5, September 2024 : 271-276

REFERENCES

1. Regehr J, Kriwiel M, Roth E, Ofei-Dodoo S. A case report of a modern-day scurvy. Kans J Med.2021;14. doi:10.17161/kjm.vol1414609.
2. Lund RM, Becker ML, Shapiro S, Allison T, Harris JG. Scurvy presenting with limp and weakness: a case report. BMC Pediatr. 2019;19(1):228. doi:10.1186/s12887-019-1605-5.
3. Ceglie G, Macchiarulo G, Marchili MR, et al. Scurvy: still a threat in the well-fed first world? Arch Dis Child. 2019;104(4):381-383. doi:10.1136/archdischild-2018-315496.
4. Rowe S, Carr AC. Global Vitamin C Status and Prevalence of Deficiency: A Cause for Concern?. Nutrients. 2020;12(7):2008. Published 2020 Jul 6. doi:10.3390/nu12072008
5. Fain O. Musculoskeletal manifestations of scurvy. Joint Bone Spine. 2005;72(2):124-128. doi:10.1016/j.jbspin.2004.01.007.
6. Alqanatish JT, Alqahtani F, Alsewairi WM, Al-kaizen S. Childhood Scurvy: an unusual cause of refusal to walk in a child. Pediatr Rheumatol.2015;13(1):23. doi:10.1186/s12969- 015-0020-1.
7. Miraj F, Abdullah A. Scurvy: Forgotten diagnosis, but still exists. Int J Surg Case Rep. 2020;68:263-266. doi:10.1016/j.ijscr.2020.03.002.
8. Thiemann S, Cimorelli V, Bajwa NM. Case Report: Uncommon cause of limp in the 21st century. Front Endocrinol. 2022;13:968015. doi:10.3389/fendo.2022.968015.
9. Trapani S, Rubino C, Indolfi G, Lionetti P. A Narrative Review on Pediatric Scurvy: The Last Twenty Years. Nutrients. 2022;14(3):684. Published 2022 Feb 6. doi:10.3390/nu14030684.
10. Gallizzi R, Valenzise M, Passanisi S, Pajno GB, De Luca F, Zirilli G. Scurvy may occur even in children with no underlying risk factors: a case report. J Med Case Reports. 2020;14(1):18. doi:10.1186/s13256-020-2341-z.