

Scurvy Presenting as Painful Gait With Bruising in a Young Boy

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A case of scurvy occurred in an apparently well-nourished 5-year-old boy with normal growth parameters. Only after the diagnosis of scurvy was raised on clinical grounds did we discover the peculiar dietary habits that were responsible for his deficiency of ascorbic acid. His case is a reminder to the clinician that nutritionally based disease may occur in any socioeconomic setting and that nutritional screening remains an important part of every child's general health care.

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Scurvy has been clinically recognized for centuries. One of the earliest historical documentations was by Jacques de Vitz in 1218, which described epidemics among populations affected by war.¹ In his *Treatise of the Scurvy* in 1747, James Lind described one of the earliest clinical trials. He showed that out of 12 scorbutic sailors, only 2 who ate citrus fruits were cured of their symptoms, providing the first link between diet and the disease.² Since the discovery of the antiscorbutic property of ascorbic acid in the 1930s, the worldwide incidence of scurvy has declined. However, as dietary patterns vary, isolated cases continue to occur. We report a case of scurvy occurring in a 5-year-old boy who had regular access to medical care. His presentation with rheumatic complaints and skin lesions resulted in confusion with possible immunologic or neoplastic disease.

REPORT OF A CASE

A previously healthy 5-year-old white boy with an unremarkable medical history developed a limp. He had bilateral peripatellar bruising and tenderness, suspected to be traumatic. His pain and ecchymoses resolved. However, 4 months later his painful limp returned. Pain worsened over the ensuing 4 weeks, until he was unable

to get out of bed or walk without assistance. His physician documented the return of ecchymoses around both knees and over the lower legs. On the day before his referral to a tertiary care children's hospital, he developed gingival bleeding.

On examination, he was alert and playful with normal vital signs. Height and weight were in the 85th percentile for his age and sex. Scattered petechiae and perifollicular purpuric macules were noted on his trunk and lower extremities. After blood drawing, there was a marked increase in the petechiae on his arms distal to the sites of tourniquet placement, with sharp circumferential demarcation (**Figure 1**). He had normal abdominal examination results, without organomegaly. He had no apparent joint swelling, but there was pain with motion of his knees and hips. No muscle weakness was noted, although he displayed a wide-based, waddling gait and required assistance to ambulate. The remainder of his neurologic examination results were normal. Intraoral examination showed swollen gingiva with an ecchymotic, spongy appearance of several interdental papillae (**Figure 2**).

The differential diagnosis included vasculitis, coagulopathy, idiopathic thrombocytopenic purpura, and malignant neoplasm. Evaluation included a white blood cell count of $7.2 \times 10^9/L$ with a normal differential count, a platelet count of $238 \times 10^9/L$, and erythrocyte sedimentation rate of 17 mm/h. He had a micro-

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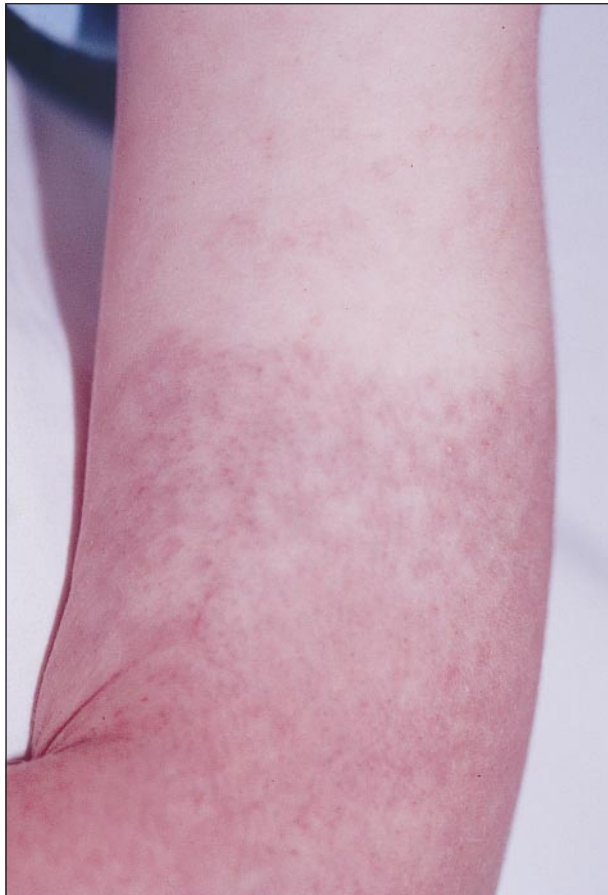


Figure 1. Well-demarcated petechiae on the upper arm distal to the site of tourniquet placement (Rumpel-Leede sign).

cytic anemia, with a hematocrit level of 0.30 and mean corpuscular volume of 77 fL. Coagulation times and blood chemistry results were normal, except for a lactate dehydrogenase level of 804 U/L (reference range, 372-744 U/L). Bone scan showed minor uptake in the sacroiliac regions. Knee radiographic scans showed dense lines at the ends of the metaphyses, with adjacent lucent bands (**Figure 3**).

Because his normal platelet count was believed to be incompatible with leukemia as a cause of diffuse intradermal bleeding, the alternative diagnosis of scurvy was considered. A detailed review of the boy's dietary history revealed severely limited variety: in the most recent 5 months, his diet consisted of biscuits, Pop-Tarts® (Kellogg's C, Battle Creek, Mich), cheese pizza, and water. He refused fruits, vegetables, and juices, and his mother had been unsuccessful in administering chewable vitamin supplements. Serum ascorbic acid level was lower than 5.68 µmol/L (reference range, 11.4-114 µmol/L). After an intravenous infusion of 100 mg of ascorbic acid, a 24-hour urine ascorbic acid level was low at 11 mg (15-30 mg/24 hours), which was consistent with depleted ascorbic acid stores. Other nutritional laboratory study results were normal, except for a low zinc level of 8.4 µmol/L (reference range, 9.2-18.4 µmol/L).

These laboratory findings were diagnostic of scurvy. After administration of ascorbic acid, his pain improved rapidly, and he again began to walk. After discharge from

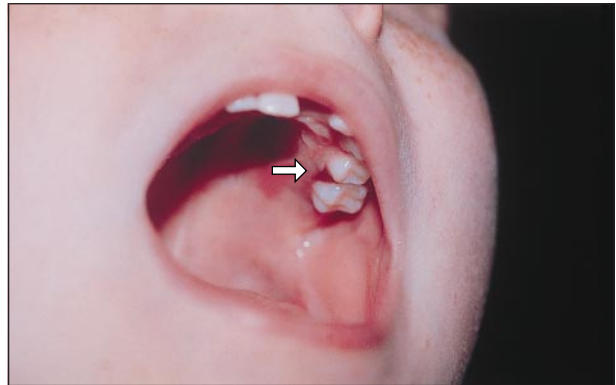


Figure 2. Interdentary papillae with an ecchymotic and spongy appearance (arrow).

the hospital, he took 100 mg of ascorbic acid twice daily, 2.5 mg of zinc sulfate twice daily, 1 multivitamin with iron daily, and 224 g of a pediatric nutritional supplement with each meal. Complete resolution of his pain occurred within 1 week, and his skin lesions faded simultaneously. Follow-up serum ascorbic acid level at 6 weeks was 90.8 µmol/L. With behavioral modification, his dietary intake improved, and he remained well.

COMMENT

Ascorbic acid is a potent reducing agent and a cofactor for numerous enzymes that affects a variety of body functions. Humans cannot synthesize ascorbic acid and are dependent on intestinal absorption of this essential nutrient through an active sodium-dependent transporter protein. Absorbed ascorbic acid is transported to tissues in water-soluble form.^{3,4} It is estimated that evidence of scurvy may appear within 2 to 4 months of inadequate ascorbic acid intake.⁵

Ascorbic acid plays an essential role as a cofactor for procollagen proline/lysine hydroxylase. The most common signs and symptoms of scurvy are manifestations of decreased production and increased fragility of collagen, because of faulty hydroxylation of proline and lysine residues critical in procollagen triple helix formation.^{6,7} Signs of scurvy are secondary to capillary fragility, and include intradermal and gingival hemorrhage, bone pain from subperiosteal hemorrhage, and loosening of the teeth secondary to decreased dentin.

Infants may present with irritability and "frog-legged" pseudoparalysis, with external rotation and flexion of their lower extremities from the bone pain.^{8,9} "Scorbutic beads," resembling the rib changes associated with rickets, may develop at the costochondral junctions because of subluxation of the sternum and associated hemorrhage.⁹ Older children present with bone pain or inability to walk¹⁰ and bleeding, mimicking leukemia or a diffuse bleeding disorder. Severe scurvy manifests as skeletal muscle degeneration, cardiac hypertrophy, diminished bone marrow function, and adrenal atrophy.⁸ In contrast, adults with scurvy initially present with fatigue, lethargy, and, in addition to bone pain, may develop sicca, anemia, ocular hemorrhage, psychological changes, and, rarely, femoral neuropathy, alopecia, and

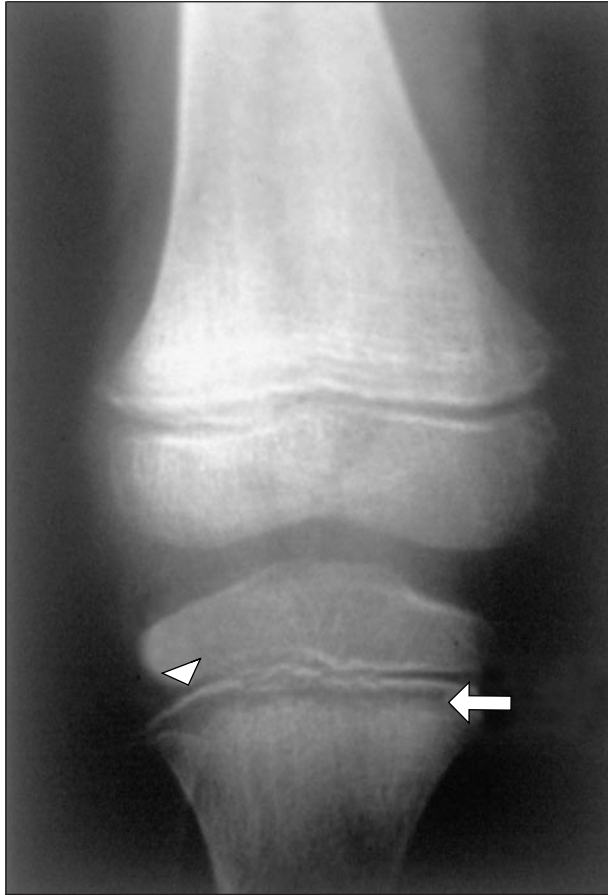


Figure 3. Knee radiographic scan shows a dense white line at the proximal end of the tibial metaphysis (arrowhead), with adjacent lucent band (arrow).

diffuse edema.³ If left untreated, death may ensue from cerebral hemorrhage or hemopericardium.¹¹

Dermatologic findings appear early in the disease and may be helpful clues to the diagnosis. These include petechiae, ecchymoses, corkscrew or swan-neck hairs, follicular hyperkeratosis, and perifollicular hemorrhage. Perifollicular hemorrhage occurs secondary to plugging of hair follicles with keratin and associated proliferation and bleeding of fragile blood vessels.¹² A unique finding associated with the vascular fragility is called the Rumpel-Leede sign. This is a distal shower of petechiae with a sharp proximal line of demarcation, occurring immediately after release of venous occlusion of the extremity.¹³ Although this phenomenon is sometimes seen in association with sepsis or thrombocytopenia, it is also a helpful sign of capillary fragility. Our patient developed petechiae distal to tourniquet placement.

Anemia is the most common hematologic manifestation of scurvy, present in up to 80% of cases, and was seen in our patient. The etiology of anemia is believed to be multifactorial, including blood loss, concurrent vitamin deficiencies (especially folate), and decreased iron absorption.¹⁴ Ascorbic acid increases iron absorption by reducing it from the ferric (Fe+3) to the more absorbable ferrous (Fe+2) state.³

The most common radiographic finding in scurvy is bony demineralization. There is preservation of the zones of provisional calcification at the ends of the me-

taphyses (the white lines of Frankel). The lucent line adjacent to this is the scurvy zone, an area through which fractures may occur. The classic Pelkan spurs at the periphery of the zone of calcification are the result of healing of these fractures. Another classic finding is the fine white line (Wimberger ring) outlining the epiphyses. If subperiosteal bleeding occurs, extensive periosteal new bone is seen along the shafts of the long bones.^{15,16} Our patient's x-ray films demonstrated the white lines of Frankel and the scurvy zones. Because he did not demonstrate other classic radiographic findings of scurvy, the differential diagnosis of his symptoms included leukemia, since lucent metaphyseal bands occur in this condition also.

The diagnosis of scurvy is largely made on clinical grounds,⁸ and may be supported by serum ascorbate levels of less than 11.4 $\mu\text{mol/L}$, or buffy-coat levels of lower than 1700 $\mu\text{mol/L}$. The serum values reflect metabolic turnover, while buffy-coat values represent tissue stores.¹⁷ In practice, serum values are simpler to obtain and more universally available.

Serum ascorbate levels are specific but not sensitive. They may be normal, even in states of severe depletion. A better indicator of body stores is the measure of urinary excretion after parenteral ascorbic acid challenge. Eighty percent of a given parenteral dose should be excreted within 5 hours if body stores are adequate.^{8,18} Our patient had both low serum values and reduced excretion after intravenous administration, supporting the diagnosis of scurvy. The strongest indicator of disease is the resolution of manifestations after the administration of ascorbic acid. For mild disease, 100 mg daily, and for severe disease, 500 to 1000 mg daily, may be administered for 1 week, with reduced doses until full recovery.^{8,12,19-22} Maintenance is achieved with doses of 30 to 60 mg daily. Oral and constitutional symptoms will usually resolve in 2 to 3 days,²³ with ecchymoses, bone abnormalities, and anemia usually resolving within 2 weeks.^{12,19,21}

Because foods are increasingly supplemented with ascorbic acid, concern for scurvy has diminished in the developed world. However, individuals who remain at risk for scurvy include the elderly, those who infrequently prepare balanced meals,²² psychiatric patients,²⁴ the homeless, food faddists,²⁰ alcoholics,²¹ and dyspeptics who avoid acid-containing foods.²² Also at risk are dialysis patients²⁵ and infants who are fed evaporated or boiled milk in which the ascorbic acid has been destroyed by heat.^{8,26}

The development of scurvy in an otherwise healthy child is remarkable, contributing to the initial confusion at our patient's presentation. While his mother described his diet as "normal for his age" during the admission history, the child exhibited severe dietary restriction, which seemed to be behavioral in nature. Despite repeated attempts by his mother to modify his diet, he had steadfastly refused to alter dietary intake. Eating behavior severe enough to result in malnutrition and scurvy should alert the practitioner to possible underlying psychiatric disorders in the patient or in the family. Psychiatric disorders to consider include mood disorders, obsessive-compulsive disorder, eating disorders, or thought disorders.²⁷ Behavioral modification resulted in

improved dietary intake and resolution of symptoms for our patient, but he should continue to be observed for future development of psychiatric and/or behavioral problems.

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REFERENCES

1. Pauling L. *Vitamin C, the Common Cold, and the Flu*. San Francisco, Calif: WH Freeman & Co; 1976:23.
2. Lind JA. *A Treatise on the Scurvy*. Edinburgh, Scotland: Edinburgh University Press; 1753.
3. Levine M, Rumsey S, Wang Y, et al. Vitamin C. In: Ziegler EE, Filer JL, eds. *Present Knowledge in Nutrition*. Washington, DC: ILSI Press; 1996:146-159.
4. Subcommittee on the Tenth Edition of the Recommended Dietary Allowances, National Research Council. *Recommended Dietary Allowances, 10th Edition*. Washington, DC: National Academy Press; 1989.
5. Hodges RE, Baker EM, Hood J, Sauberlich HE, March SC. Experimental scurvy in man. *Am J Clin Nutr*. 1969;22:535-548.
6. Minor RR. Collagen metabolism: a comparison of diseases of collagen and diseases affecting collagen. *Am J Pathol*. 1980;98:227-279.
7. Peterkofsky B. Ascorbate requirement for hydroxylation and secretion of pro-collagen: relationship to inhibition of collagen synthesis in scurvy. *Am J Clin Nutr*. 1991;54(suppl 6):1135S-1140S.
8. Behrman RE, Kliegman RM, Arvin AM, Nelson WE, eds. *Nutritional disorders*. In: *Nelson Textbook of Pediatrics*. Philadelphia, Pa: WB Saunders Co; 1996:178-179.
9. Cassidy JT, Petty RE, eds. *Musculoskeletal manifestations of systemic disease*. In: *Textbook of Pediatric Rheumatology*. Philadelphia, Pa: WB Saunders Co; 1995:569-570.
10. Ramar S, Sivaramakrishnan V, Manoharan K. Scurvy: a forgotten disease. *Arch Phys Med Rehabil*. 1993;74:92-95.
11. Adelman HM, Wallach PM, Gutierrez F, et al. Scurvy resembling cutaneous vasculitis. *Cutis*. 1994;54:111-114.
12. Ghorbani AJ, Eichler C. Scurvy. *J Am Acad Dermatol*. 1994;30:881-883.
13. Gross AS, Holloway RJ, Fine RM. The Rumpel-Leede sign associated with drug-induced erythema multiforme. *J Am Acad Dermatol*. 1992;27:781-782.
14. Oski FA. Anemia related to nutritional deficiencies other than vitamin B12 and folic acid. In: Williams WJ, Beutler E, Erslev AJ, Lichtman MA, eds. *Hematology*. New York, NY: McGraw-Hill Co; 1990:549-550.
15. Laor T, Jaramillo D, Oestreich AE. Musculoskeletal system. In: Kirks DR, Griscom NT, eds. *Practical Pediatric Imaging: Diagnostic Radiology of Infants and Children*. Philadelphia, Pa: Lippincott-Raven Publishers; 1998:452-453.
16. Swischuk LE, ed. *Imaging of the Newborn, Infant, and Young Child*. Baltimore, Md: Williams & Wilkins; 1989:791-792.
17. Wilson CW. Clinical pharmacological aspects of ascorbic acid. *Ann NY Acad Sci*. 1975;258:355-376.
18. Barness LA. Nutrition. In: Kelley VC, ed. *Practice of Pediatrics*. Philadelphia, Pa: Harper & Row; 1987:21.
19. Baumbach J. Scurvy by another name: a case report. *R I Med*. 1994;77:24-25.
20. Ellis CN, Vanderveen EE, Rasmussen JE. Scurvy: a case caused by peculiar dietary habits. *Arch Dermatol*. 1984;120:1212-1214.
21. Onorato J, Lynfield Y. Scurvy. *Cutis*. 1992;49:321-322.
22. Reuler JB, Broudy VC, Cooney TG. Adult scurvy. *JAMA*. 1985;253:805-807.
23. Yalcin A, Ural AU, Beyan C, Tastan B, Demiriz M, Cetin T. Scurvy presenting with cutaneous and articular signs and decrease in red and white blood cells. *Int J Dermatol*. 1996;35:879-881.
24. Mowad CM, Howe KL, Guzzo CA. Unexplained hemothorax, hemothrosis, and palpable purpura. *Hosp Pract (Off Ed)*. 1995;30:55-56.
25. Sullivan JF, Eisenstein AB. Ascorbic acid depletion in patients undergoing chronic hemodialysis. *Am J Clin Nutr*. 1970;23:1339-1346.
26. Silverman FN, Kuhn JP, eds. *Dysostoses, miscellaneous syndromes, and metabolic abnormalities*. In: *Caffey's Pediatric X-ray Diagnosis: An Integrated Imaging Approach*. St Louis, Mo: Mosby-Year Book Inc; 1993:1754-1759.
27. Lucas AR. Eating disorders. In: Lewis M, ed. *Child and Adolescent Psychiatry*. Baltimore, Md: Williams & Wilkins; 1991:573-582.