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.

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 scurbutic 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 × 10^9/L with a normal differential count, a platelet count of 238 × 10^9/L, and erythrocyte sedimentation rate of 17 mm/h. He had a micro-
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 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...
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absorption.14 Ascorbic acid increases iron absorption by tamin deficiencies (especially folate), and decreased iron to be multifactorial, including blood loss, concurrent vi-
seen in our patient. The etiology of anemia is believed helpf ul sign of capillary fragility. Our patient developed association with sepsis or thrombocytopenia, it is also a petechiae distal to tourniquet placement.

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 spur s 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.5,10 Our patient’s x-ray films demonstrated the white lines of Frankel and the scurvy zones. Because he did not demonstra te 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 µmol/L, or buffy-coat levels of lower than 1700 µmol/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 intrave-

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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 infre-
dently prepare balanced meals,22 psychiatric patients,24 the homeless, food faddists,26 alcoholics,27 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 confu sion at our patient’s presentation. While his mother de scribed his diet as “normal for his age” during the adm ission 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