A 15-MONTH-OLD boy had a history of unexplained bleeding from his gums for several weeks and fever for 2 days. He had been fed only cow’s milk and oatmeal since age 4 months. On physical examination he had almost no spontaneous movement. His legs were held in a “frog leg” position (Figure 1), were swollen along the long bones, and were tender to palpation. His skin was dry and pale. Hemorrhages of the gingiva were obvious as were 2 blood-filled cysts of the lower canine teeth (Figure 2). The tympanic membranes were hyperemic, and evidence of middle-ear fluid was present. Palpable prominence of the costochondral junctions of the chest wall was noted.

The results of laboratory examinations revealed a normal white blood cell count, a hemoglobin level of 76 g/L, and a platelet count of 334 $\times 10^9$/L. The serum calcium, phosphorous, copper, and alkaline phosphatase levels were normal. Thyroid stimulating hormone, triiodothyronine, and thyroxine levels were also normal. The serum level of vitamin D was normal, but vitamin C levels were low, 28 µmol/L (reference range, 45-108 µmol/L). Chest x-ray film showed a scorbutic rosary at the costochondral junctions with a “corner” sign noted in the proximal metaphysis of the humerus. Lower extremity radiographs demonstrated abnormalities (Figure 3).

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Infantile Scurvy

Infantile scurvy (Moeller-Barlow disease) is rarely reported in developed countries except in association with global malnutrition. The rarity of occurrence is responsible for the frequent delayed recognition of this disorder. A deficiency of vitamin C (ascorbic acid) is responsible for the manifestations of scurvy. Vitamin C is a cofactor for numerous enzymes that are critical to several body functions. The most common manifestations of scurvy are due to the decreased production and increased fragility of collagen, a result of faulty hydroxylation of proline and lysine in forming a precise triple-helical collagen structure. Vitamin C also plays a number of roles in hematopoiesis, including the promotion of iron absorption and as a cofactor for the conversion of folic acid to folinic acid. Humans cannot synthesize vitamin C and must rely on intestinal absorption from ingested nutrients.

Clinical Manifestations

Most cases of scurvy are seen between ages 6 and 24 months. Neonatal scurvy is rare, except in infants of mothers with extreme hypovitaminosis C. Breast milk contains sufficient amounts of vitamin C unless the mother's diet is deficient in this vitamin. The first clinical manifestations of scurvy are often associated with acute febrile illnesses that seem to increase the need for vitamin C. Initial manifestations of scurvy are vague and include irritability, decreased appetite, and delayed development. As effects of vitamin C deficiency progress, affected children lie still with little movement because of generalized tenderness, most apparent in bones as a result of subperiosteal hemorrhages. Swelling may be noted along the shafts of long bones. Pseudoparalysis may be apparent as a result of the bone pain. Infants often hold their legs in a “frog leg” position and dislike being handled, often refusing to walk.

Gingival hemorhages may occur along with spongy, hemorrhagic swellings of the mucous membranes overlying teeth. Petechial hemorhages may occur in the skin spontaneously or, more commonly, below the site of tourniquet application, the Rumpel-Leede sign, as a manifestation of capillary fragility. Swelling may be palpated along the costochondral junctions of the rib cage, resembling the rosary seen in rickets.

Radiographic Findings

Changes in the long bones, particularly around the knee, are most diagnostic of scurvy. The earliest finding is generalized demineralization with a ground-glass appearance to the bones. The cortex is thinned. A white line, so-called Frankel sign, is apparent at the ends of metaphyses, representing widening of the provisional zones of calcification and its increased density. The epiphyseal centers show central rarefaction and are also surrounded by a white line of calcification, referred to as the Wimberger ring or halo-ossification center. Transverse bands of diminished density adjacent to the Frankel sign are known as scurvy lines. Lateral metaphyseal spurs (corner sign) between the provisional zone of calcification and the cortex are the result of metaphyseal infarctions. The corner sign is more pathognomonic of scurvy than the other bony findings. Irregular calcification and widening of the costochondral junctions results in the scurbutic rosary. Periosteal hemorrhages cannot be detected on radiography in scurvy until they become calcified during healing.

Differential Diagnosis, Diagnosis, and Treatment

Bleeding manifestations and bone pain may suggest acute leukemia. Bone pain and refusal to walk may lead to consideration of osteomyelitis, septic arthritis, and rheumatic disorders.

The diagnosis of scurvy is based on a combination of clinical and radiographic findings. A dietary history compatible with insufficient intake of vitamin C should be present. Accurate laboratory measurement of vitamin C levels is difficult because of the instability of vitamin C.

Healing occurs rapidly with the oral administration of 100 to 200 mg/d of vitamin C. As healing occurs, the intake of vitamin C may be reduced to 50 mg/d until complete clinical and radiologic resolution has taken place.

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References

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