Denouement and Comment

Scurvy

The child's presentation was suggestive of scurvy. Testing of the serum ascorbic acid level was ordered, and the patient was discharged on 300 mg of ascorbic acid daily and a multivitamin. One week after discharge, his gingival swelling and petechiae resolved. By 1 month, he was ambulating and his hemoglobin level was 12.2 g/dL. The ascorbic acid level on the initial testing was 1.057×10^{-4} mg/dL (reference range, $>4.227 \times 10^{-4}$ mg/dL; to convert to micromoles per liter, multiply by 56.78).

Scurvy is thought to linger among alcoholic and disadvantaged individuals. However, vitamin C deficiency in children with developmental disorders who consume restricted diets may be more common than thought. Noble et al[1] reported 23 case studies of scurvy in children with restricted diets, including children with autism, developmental delay, and cerebral palsy.

Humans require exogenous vitamin C as they lack the innate ability to convert glucose to ascorbic acid. It is an important cofactor for collagen hydroxylation and protein biosynthesis.[2] Common sources include fresh fruits, vegetables, and supplements. The clinical presentations of vitamin C deficiency are directly linked to its various uses by the body.

Signs generally develop after 1 to 3 months of inadequate vitamin C intake. The earliest signs are typically perifollicular petechiae, ecchymoses, hyperkeratosis, and corkscrew hairs attributed to capillary fragility due to diminished collagen levels. Gingival disease characterized by swelling, ecchymoses, bleeding, and loosening of the teeth secondary to vessel instability often follows.[3]

Children with scurvy most frequently present with bone disease[4] related to a defect of osteoid matrix formation and cartilage resorption leading to disordered bone structure and subsequent fractures around the growth plate.[5] Other clinical signs include skeletal muscle degeneration, cardiac hypertrophy, diminished adrenal and bone marrow function, arthritis, poor wound healing, edema, and alopecia.[6] Concurrent nutritional deficiencies may play a role in the presentation.

Anemia is a hallmark of scurvy, as ascorbic acid improves iron absorption by reducing it to the ferrous state.[3] Our patient, despite taking iron supplements, remained anemic until vitamin C supplementation. Bleeding and other dietary deficiencies may contribute to the iron deficiency anemia. An elevated erythrocyte sedimentation rate and C-reactive protein level can be seen.[5,7]

Radiological imaging can assist in diagnosis. The most common radiographic finding is nonspecific osteopenia. A high index of suspicion for scurvy is required to be alert to more specific findings including preservation of the zones of calcification at the distal metaphyses with an adjacent lucency (the scurvy zone), Pelkan spurs, and an increased density outlining the epiphyses (Wimberger ring).[8] Periosteal bone may be seen along the shafts of long bones, where subperiosteal bleeding can occur. Magnetic resonance imaging findings consistent with scurvy include subperiosteal hemorrhage or hemorrhage at fracture sites on T1- and T2-weighted images.[9]

The diagnosis of vitamin C deficiency is confirmed with a low serum ascorbic acid level. The best evidence of scurvy is resolution of symptoms with ascorbic acid supplementation. A dosage of 100 to 300 mg of vitamin C daily for infants and children and 500 to 1000 mg daily for adults is required for 1 month or until symptoms completely resolve. Evidence suggests that supplementation as low as 10 mg/d can cure scurvy in human volunteers.[10] Spontaneous bleeding, oral symptoms, and constitutional symptoms begin to improve within days, while bony changes and ecchymoses may take several weeks to resolve.[11] Untreated scurvy can result in severe morbidity, and death may occur from secondary infection, cerebral hemorrhage, or hemopericardium.[10]

This case highlights the importance of obtaining a dietary history and maintaining a high index of suspicion for restricted diets in those with developmental or behavioral disorders. When a child with developmental disability presents with musculoskeletal pain, clinicians must consider vitamin C deficiency. Increased awareness of scurvy among this population may prevent children from undergoing invasive and unnecessary investigations.

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