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Case Report

Scurvy: Cases of an Old Disease in Two Neurotypical Pediatric Patients in a Children's Hospital

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Abstract

Scurvy, caused by the dietary deficiency of vitamin C, is an uncommon disease among the pediatric population. In this report, we discuss two cases of previously healthy pediatric patients, who initially presented for evaluation of severe muscle and bone pain, with the concern of malignancy or vasculitis. Both patients showed similar physical symptoms - gingival hyperplasia with associated bleeding, petechial rash, bone tenderness and decreased mobility. Only after extensive workup and images, the diagnosis of scurvy was made after evidence of undetectable levels of vitamin C. Patients were initially managed with adequate supplementation, showed improvement, and thereafter experienced resolution of the symptoms. This ancient diagnosis, along with other nutritional deficiencies, should be considered by general pediatricians while evaluating limping, bone pain and the associated findings on the oral mucosa, especially when treating patients with restricted diets.

Keywords: Vitamin C; Ascorbic acid; Scurvy; Hypovitaminosis; Neurotypical; Autism spectrum disorder

Introduction

Scurvy is a consequence of a severe dietary deficiency of vitamin C. Modern physicians primarily associate this disease with history books that describe naval medicine, where sailors used to present with the most severe forms of scurvy due to the absence of fresh fruits and vegetables in their diet [1]. Vitamin C, or ascorbic acid, is a water-soluble, essential vitamin that cannot be stored in the body for prolonged periods of time. For this reason, it must be ingested on a regular basis in order to maintain adequate levels of vitamin C in blood. Signs of deficiency occurs 1-3 months after prolonged poor vitamin ingestion, registering below 10 mg/dl. Patients with scurvy present with a variety of symptoms that include dermatological, dental, bone and systemic findings. This vitamin plays an important role in the formation of tendons, ligaments, blood vessels and skin, as it has a major contribution in collagen formation. Low intake of this vitamin translates into fragility of the small blood vessels and capillaries and can present upon physical exam as petechial rash, ecchymosis and gingival hypertrophy [2-4]. Currently, among the pediatric population, this diagnosis is mostly associated with autism spectrum disorder or with a neurological disadvantage diagnosis such as cerebral palsy. We present two cases of unsuspected scurvy in two neurotypical patients who presented to Nicklaus Children's Hospital located in Miami, Florida, former Miami Children's, with nonspecific symptoms of fatigue, petechial rash, bleeding gums, follicular hemorrhages and bone pain suggestive of osteopenia. In both cases, the initial suspected diagnosis ranged from vasculitis to malignancy, with vitamin C deficiency as a less likely diagnosis, given its uncommon prevalence.

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Case Presentation

Case 1: Hypovitaminosis related to goat's milk intake in a 3-year-old

A 3-year-old female presented to the hospital with lower extremity pain and limping, a refusal to bear weight, as well as petechial rash and abdominal pain. The patient had a history of poor nutritional intake, described as a selective food eater. In addition, she was found to have several nutritional allergies, such as nut allergy and lactose intolerance, restricting her diet to goat's milk; she refused to eat or drink any solid foods, fruits or vegetables. She was initially evaluated in the emergency department, where she was found to have macrocytic anemia and neutropenia. Her physical exam was positive for gingival hyperplasia with associated bleeding, petechial rash involving the face and lower extremities, follicular hyperkeratosis with associated perifollicular hemorrhage, refusal to walk and an inability to bear weight. Her anthropometric evaluation showed moderate malnutrition, evidenced by a BMI z-score of -2.25. Hypovitaminosis was highly suspected, due to a severely limited diet and supported by undetectable vitamin C levels, in addition to low vitamin B12 and folate levels. She was provided with supplemented vitamin C, 100 mg

daily for one month, and showed improvement of symptoms by day four of hospital admission.

Case 2: Hypovitaminosis related to poor diet in a 16-year-old male

A 16-year-old male with a history of gastritis, leading to poor nutritional intake, presented for evaluation of leg pain, leg swelling, petechial rash and gum bleeding. He attested to pain in the ribs that began four weeks prior, that then progressed to leg pain and associated limping, impairing his ambulation. A week prior to his admission, he developed a petechial rash in the legs, which progressively worsened, and gum bleeding. He also complained of fatigue and easy bruising. On the day of admission, the pediatrician evaluated him and he was referred to the ED for further evaluation of hepatomegaly. the physical exam was positive for gum bleeding, petechial rash involving upper and lower extremities, follicular hyperkeratosis with associated perifollicular hemorrhage, refusal to walk and inability to bear weight on the left leg, significant rib tenderness, and hepatomegaly 2 cm below the rib cage. Hypovitaminosis was suspected, given poor dietary intake of vegetables and fruits. Levels of vitamin C were undetectable and skin biopsy was consistent with scurvy (Figure 1). The patient was started on Vitamin C therapy with 500 mg daily, showing improvement by day three of admission, and resolution of symptoms by two weeks; treatment continued for one month.

Discussion

Scurvy is a disease that is primarily seen in developing countries, however it has been more frequently diagnosed in patients with poor nutritional intake, secondary to developmental impairment or food aversion. It is caused by a poor or absent intake of vitamin C, for a prolonged period of time, usually 1-3 months. Vitamin C acts as a major contributor in collagen formation, by hydroxylating proline and lysine. It also acts as an antioxidant and reduces iron to its ferrous form so it can be absorbed easily. Due to small blood vessels and capillary fragility, petechiae and follicular hemorrhages are commonly seen. Bone pain, due to osteopenia, leads to increased risk of micro fractures and subperiosteal hemorrhages [5]. Patients with restricted diets, such as in our two cases, have an increased risk of developing hypovitaminosis. Our first case described a diet consisting mainly of goat's milk. Goat's milk formula has been commercially available since the late 1980's. In terms of minerals and vitamins, compared to cow's milk, goat's milk has lower concentrations of riboflavin, vitamin B12, folate and pantothenate. The concentration of ascorbic acid, however, is similar in both formulas. The risk of vitamin C deficiency is greater, not due to a preference of drinking cow's versus goat's milk, but due to the restriction of other sources of nutrients such as vegetables and fruits (Table 1) [6,7]. Scurvy is uncommon in industrialized nations, and is particularly rare in children. Most documented pediatric cases of scurvy involve patients with cerebral palsy, autism spectrum disorder or some other neuropsychiatric disorder. Self-restrictive diets due to "picky eating", food aversion or avoidant restrictive food intake disorder, could lead to nutritional deficiencies in otherwise developmentally appropriate children. Given its rarity compared to other hypovitaminosis, is not usually suspected and frequently leads to a late diagnosis and treatment [8,9]. Vitamin C deficiency can lead to poor bone formation; early findings are swelling and tenderness of the joints, most commonly the knees and ankles, causing pseudo paralysis due to pain. This can progress to osteopenia and malformation of the bones, restricting bone growth [10]. Rarely can ascorbic acid deficiency progress to skeletal muscle degeneration, pulmonary hypertension, diminished adrenal and bone marrow function, cardiac hypertrophy, psychological changes or edema, but ultimately it can lead to disruption of metabolic processes of the body and can be fatal, with deaths reported as a result of infection, cerebral hemorrhage or hemopericardium [11].

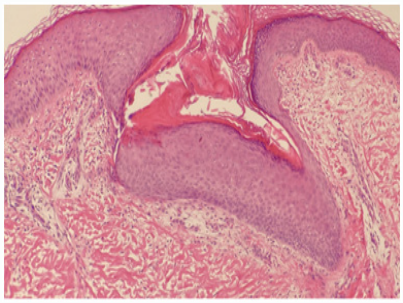

Figure 1: Skin Biopsy, Case 2. Follicular hyperkeratosis and plugging with sparse superficial perivascular lymphocytic infiltrate, along with perivascular periadnexal and interstitial. (Hematoxylin and eosin, original magnifications x100).

Table 1: Average vitamin content of goat's *vs.* cow's milk, expressed in mg/dL. Numbers from USDA Handbook 8-1 (1976) [11].

Vitamin	Cow	Goat
Ascorbic Acid	21.1	15
Vitamin D	33	23.7
Vitamin A	1560 IU/liter	2074 IU/liter
Thiamine	0.44	0.4
Riboflavin	1.75	1.84
Pyridoxine	0.64	0.07
Biotin	0.031	0.039
Folic Acid	0.0028	0.0024
Cobalamin	0.0043	0.0006

Conclusion

Vitamin C deficiency is a disease that can affect developmentally appropriate children in industrialized countries. This disease, although preventable and mainly seen in low-income countries, continues to be a cause of bone pain in the pediatric population of modern civilization. As pediatricians, a high index of suspicion for scurvy should be kept as a cause of unexplained bone pain accompanied by skin and mucosal findings. A good dietary history and pertinent vitamin supplementation can prevent hospitalization and extensive, high-cost workups.

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