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POINT OF VIEW

Scurvy: the return of an almost forgotten disease

Sheila Knupp Feitosa de Oliveira[1], Felipe Oliveira De Paula[2]

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Abstract

The last 20 years have shown an increpa-se in cases of scurvy in children complaining of pain in the limbs, back and refusal to walk. The article helps identify the causes and patients at risk, how to diagnose, treat and prevent.

[1] Universidade Federal do Rio de Janeiro, Pediatria - Rio De Janeiro - Rio de Janeiro - Brazil.
[2] Universidade Estácio de Sá, Curso de Medicina - Rio De Janeiro - Rio de Janeiro - Brazil.

Correspondence to:
Sheila Knupp Feitosa de Oliveira.
Universidade Federal do Rio de Janeiro. R. Bruno Lobo, 50 - Cidade Universitária, Rio de Janeiro, RJ, Brazil. CEP 21941-912 E-mail: sheila_knupp@hotmail.com

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For decades, we have not heard a thing about scurvy. It belonged to the past; a disease described a few centuries ago seen in sailors with minimal access to fresh fruit and vegetables and later in children living in areas where malnutrition was a major socioeconomic problem. This year, I was surprised to see four paper presentations delivered at scientific meetings held in Brazil, in which the cases of five children with scurvy were described. Their symptoms included arm and leg pain, and they were treated in pediatric rheumatology services in four Brazilian states (Rio de Janeiro, Rio Grande do Sul, Bahia, and Minas Gerais) [1-4]. Scurvy is coming back in other parts of the world, as shown in a review article published in 2022 featuring 166 similar cases reported in the last 20 years [5].

What links this disease to pediatric rheumatology? What distinguishes the children who contract the disease? How does the disease behave? How is it diagnosed? How should patients be treated? These are some of the questions pediatricians should answer to identify and treat patients with scurvy and prevent the complications resulting from delayed diagnosis.

Scurvy is caused by a lack of vitamin C (ascorbic acid), a nutrient found in fresh fruits and vegetables. It is involved in several organic functions, such as the synthesis of collagen, neurotransmitters, nitric oxide, and the transport of fatty acids. Unfortunately, the human body cannot store vitamin C, and the first signs of deficiency may appear only after one to three months [6]. The cases described in the last 20 years often involved children with neurological comorbid conditions or under voluntary dietary restriction (selective appetite). Sixteen percent of the individuals with scurvy had autism spectrum disorder, and 8% had cerebral palsy and developmental delays. Twenty-nine percent were malnourished, and 23% were in good health [5].

The first symptoms are nonspecific and include fatigue, pain, irritability, loss of appetite, and mood changes. The disease progresses with skin and bone manifestations due to reduced collagen synthesis, which weakens the capillaries and the bone structure [6]. Skin disorders include hyperkeratosis, petechiae, ecchymosis, and perifollicular hemorrhage. Hematological diseases such as von Willebrand disease, purpura, and leukemia are often considered in the differential diagnosis of scurvy. However, only 42% of pediatric patients become anemic, likely due to the role of vitamin C in iron absorption; none develop thrombocytopenia, and their coagulation tests are unremarkable [6]. Disordered osteoid matrix formation is more significant in children than adults and often causes severe skeletal alterations. Arthralgia, myalgia (mainly in the legs), back pain, acute limping, and refusal to walk are the most common symptoms (92%) and usually lead to a diagnosis of scurvy after a few weeks or months of investigation for other conditions, such as rheumatological and hematological diseases, infection, and amplified musculoskeletal pain syndrome. An essential element is the persistence of painful symptoms with immobilization. Hemarthrosis, bleeding within the muscles and periosteum, is rarely described. Gingival hypertrophy and gum bleeding, classic findings in cases of scurvy, usually manifest after patients have been diagnosed with the disease [5].

The painful areas pinpointed during the interview and physical examination are the target areas of imaging exams performed to confirm scurvy and rule out other conditions. Trampani et al. noted the presence of the following radiological changes in 166 patients with scurvy: generalized osteopenia and cortical thinning (31%); Wimberger ring, an opaque circular calcification surrounding the osteoporotic epiphyseal center of ossification (41%); Frankel line, a dense zone of epiphyseal calcification (73%); Trümmerfeld zone, a lucent metaphyseal band underlying Frankel line (14%); Pelkan spur, or cupping of the metaphysis (18%); periosteal reaction due to subperiosteal hemorrhage; and hemarthrosis [5]. Although these signs are specific, they will not be adequately evaluated if a diagnosis of scurvy is not considered. Magnetic resonance imaging is rarely needed, and few cases in the literature have described it [7]. Symmetrical, diffuse, or multifocal involvement helps in the differential diagnosis of other diseases [5,7]. Symmetrical areas of hypointense T1 and hyperintense T2 images can be observed in the bone marrow along the metaphysis of long bones, soft tissue edema and sclerotic metaphyseal bands, subperiosteal fluid collections, and periosteal reaction [8]. The differential diagnosis includes high morbidity conditions such as leukemia, metastatic neuroblastoma, chronic multifocal osteomyelitis, and bacterial osteomyelitis [7,9].

The diagnosis is evident in patients with a dietary history consistent with vitamin C deficiency and clinical and radiological signs and symptoms. When in doubt, blood tests can confirm the diagnosis by showing very low or undetectable levels of ascorbic acid (typical values vary between 0.6 and 2 mg/dL) [5,7].

Children are treated with three daily doses of 100mg of ascorbic acid in the first week, followed by one daily dose for several weeks until full recovery [6]. Treatment is effective, and signs of response are seen within 48 hours. Complete recovery from symptoms occurs within two weeks [7].

This article is meant to warn pediatricians about the increase in cases of scurvy diagnosed in children with inadequate eating habits who do not eat vitamin C-rich foods[10]. Food selectivity, a rare condition in healthy children and a common one in individuals with autism spectrum disorder and developmental delay who often suffer from gait impairment, difficulty swallowing, and need a feeding tube to eat, increases the susceptibility to scurvy [8,11]. These patients may present other nutritional deficiencies that deserve closer investigation. Interestingly, although a pediatrician regularly followed many of the children described, little importance was given to their dietary history.

REFERENCES

1. Campos L, Oliveira C, Lima F, et al. Pediatric Scurvy Mimicking Spondyloarthritis. Apresentação de Cartaz no XXV Congresso Pan-Americano de Reumatologia – PANLAR; 2023 abr 26-29; Rio de Janeiro/RJ.
2. Soveral RT, Carlos LR, Gonçalves LMK, et al. Vitamin C deficiency mimicking artritis associated with enthesitis. Apresentação de Cartaz no XL Congresso Brasileiro de Reumatologia; 2023 set 4-7; Goiânia/GO.
3. França MS, Cunha ALG, Santos ITBM, et al. Scurvy as a differential diagnosis for lower limb pain: presentation of two cases. Apresentação de Cartaz no XL Congresso Brasileiro de Reumatologia; 2023 set 4-7; Goiânia/GO.
4. Camargo MB, Ourique JM, Scheid TDS, Machado SH. Escorbuto mimetizando doença reumatológica. Cartaz apresentado no XL Congresso Brasileiro de Reumatologia, 4 a 7 de setembro de 2023, Goiânia/GO.
5. Trapani, S, Rubino C, Indolfi G, Lionetti P. A narrative review on pediatric scurvy: the last twenty years. Nutrients. 2022;14(3):684-96.
6. Gandhi M, Elfeky O, Ertugrul H, Chela HK, Daglila E. Scurvy: Rediscovering a forgotten disease. Diseases. 2023;11(2):78-89.
7. Biswas S, Miller S, Cohen HL. Scurvy in a malnourished Child: Atypical imaging Findings. J Radiol Case Rep. 2022;16(9):11-5.
8. Alten ED, Chaturvedi A, Cullimore M, Fallon AA, Habben L, Hughes I, et al. No longer a historical ailment: two cases of childhood scurvy with recommendations for bone health providers. Osteoporos Int. 2020;31(5):1001-5.
9. Gulko E, Collins LK, Murphy RC, Thornhill BA, Taragin BH. MRI findings in pediatric patients with scurvy. Skeletal Radiol. 2015;44(2):291-7.
10. Hahn T, Adams W, Williams K. Is vitamin C enough? A case report of scurvy in a five- year-old girl and review of the literature. BMC Pediatrics. 2019;19(74):1-6.
11. Zessis NR, Peters SW, Samet JD, Parzen-Johnson S, Russo LT, Samady W, et al. Unusual Presentation of Pediatric Scurvy: A Necrotic Gastrostomy Tube Site in a 14-Year-Old Boy. Am J Case Rep. 2023;24:e940770-1– 8.