Vitamin C deficiency presenting as pseudoscleroderma in a pediatric patient with food aversion

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INTRODUCTION

Scurvy is a rare but potentially fatal nutritional disorder caused by a deficiency of vitamin C, an essential nutrient crucial to collagen synthesis, wound healing, and antioxidant defense.[1] The manifestations of scurvy can affect multiple organ systems, including the skin, and mimic a wide range of dermatologic, orthopedic, hematologic, and rheumatologic conditions.[2] Common cutaneous findings include perifollicular petechiae, purpura, and swelling.[3]

Historically, scurvy was an important health concern among sailors on long sea voyages with limited access to fresh fruits and vegetables. Today, scurvy remains a concern among individuals who face food insecurity because of old age, unstable housing, or substance use disorders.[2] Pediatric patients with autism are at particular risk of scurvy resulting from avoidant/restrictive food intake disorder.[4]

Here we present a case of chronic scurvy manifesting as sclerotic edema of the bilateral lower extremities in a pediatric patient with specific food aversions.

CASE REPORT

A 17-year-old male presented to the emergency department of our hospital with a 2-month history of fatigue and worsening pain, stiffness, swelling, and discoloration of the left lower extremity. More recently, the patient noted similar changes occurring in his right lower extremity, as well as an unintended weight loss of 19 lbs over a 2-month period. On examination, abnormal findings were limited to the bilateral legs. These findings included multiple indurated violaceous plaques involving the knees, shins, and ankles, with associated skin textural changes, purpura, and follicular prominence (Fig 1). Prior to admission, concern for vasculitis was raised and treatments included a short course of prednisone (40 mg daily for 7 days) and naproxen (500 mg twice daily for 3 weeks), both with minimal symptom relief.

Complete blood count was significant for microcytic anemia, with a hemoglobin of 7.0 g/dL and mean corpuscular volume of 78.2 fL. The reticulocyte index was 1.12 with a negative direct Coombs and a ferritin of 88 ng/mL. Platelets, eosinophils, and white blood cell were within normal limits. His c-reactive protein was 6.70 mg/dL (ref 0.0-0.99) and his erythrocyte sedimentation rate was 22 mm/hr (ref 0-15). He had a negative rheumatologic work-up, including a negative anti-nuclear, rheumatoid factor, anti-cyclic citrullinated peptide, anti-topoisomerase I, and anticentromere antibodies.

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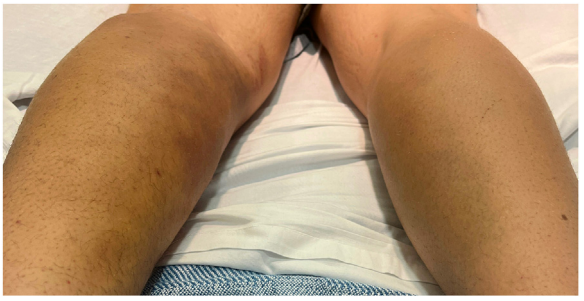

Fig 1. Indurated violaceous plaques of right and left posterior lower limbs with associated purpuric macules and patches most prominent along medial left lower limb. Well-defined violaceous indurated plaques were also noted on the anterior lower limbs and posterior thighs, with purpuric patches, follicular prominence, and textural changes.

Magnetic resonance imaging of the left lower extremity demonstrated moderate diffuse subcutaneous edema, most prominent along the fascial plane along the subcutaneous fat. Edema was also noted throughout fascial planes of the upper left leg extending to the knee. Skin punch biopsy of the indurated left posterior calf demonstrated an expanded dermis with thick collagen bundles, minimal superficial dermal mixed inflammation, and extravasated red blood cells (Fig 2). Fascial biopsy from the left lower limb demonstrated a dense histiocytic infiltrate with numerous hemosiderin-laden macrophages, and small pockets of lymphocytic inflammation with eosinophils (Fig 3).

Nutritional work-up was significant for vitamin C less than 0.1 mg/dL (ref 0.2-2.1). Vitamins D and E, as well as thiamine, copper, and zinc were all within normal limits. Further history from the patient's parents confirmed the patient's diet included primarily peanut butter sandwiches, pizza, chicken nuggets, candy, and popcorn. He did not eat red meats, green vegetables, or fruit, other than bananas.

The patient was ultimately diagnosed with fasciitis and pseudoscleroderma secondary to vitamin C deficiency and was initiated on ascorbic acid supplementation, 500 mg twice daily for 7 days. At 6 weeks follow-up he had full laboratory correction of anemia and vitamin C values. At 10 weeks he had near complete resolution of his symptoms and skin changes, with only residual hyperpigmentation.

DISCUSSION

Our case highlights pseudoscleroderma with fasciitis as an uncommon presentation of scurvy in an adolescent patient. In the United States, pediatric patients with food aversions may still be at risk for scurvy despite access to foods that contain vitamin C.

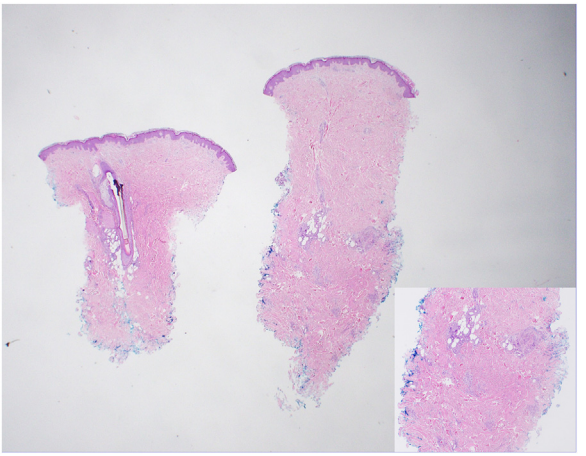

Fig 2. Skin punch biopsy of the left thigh demonstrated an expanded dermis with thick collagen bundles, as well as minimal superficial dermal mixed inflammation and hemosiderin-laden macrophages (high power insert).

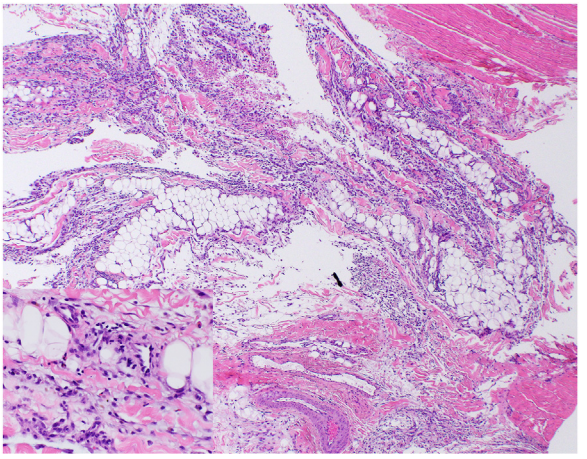

Fig 3. Fascial biopsy of the left thigh demonstrated a dense histiocytic infiltrate with numerous hemosiderin-laden macrophages and small pockets of lymphocytic inflammation with eosinophils (high power insert).

Avoidant/restrictive food intake disorder is often seen in individuals with developmental disabilities such as autism spectrum disorder, which can result in deficiencies in vitamin C and other essential nutrients.[4] Cases of scurvy among pediatric patients with autism have been documented,[5-7] with diagnoses often made following dramatic signs of deficiency such as refusal to walk, bone pain, lower extremity swelling, and gingival bleeding. Our case highlights the variety of skin findings which can present in this disease.

Early reports of scleroderma-like skin changes were documented in a cohort of 40 scurvy patients admitted to a Johannesburg hospital between 1952 and 1953.[8] Similar to our case, histopathology demonstrated red blood cell extravasation,

hemosiderin deposition, and perivascular inflammation. Schulz and colleagues presented 3 cases of scleroderma-like skin and subcutaneous changes related to vitamin C deficiency in the 1960s. Histologic examination in these patients demonstrated dermal fibrosis, mixed perivascular inflammation, and hemosiderin deposition. The authors proposed that these sclerotic changes result from a cascade of events, including hemorrhage and hemosiderin deposition within the affected areas that subsequently stimulates tissue fibroblast activity.[9]

More recently, Ahangari et al presented 2 cases in which patients developed swollen, indurated lower limbs with associated purpuric skin changes and were found to have anemia, elevated inflammatory markers, and low vitamin C. Ascorbic acid supplementation led to improvement in skin findings but did not completely resolve the underlying sclerotic changes.[10]

In our case, fascial biopsies demonstrated an inflammatory infiltrate and hemosiderin deposition along deeper tissue planes, which could support prior considerations that pseudoscleredema may be in response to chronic hemorrhage and iron deposition within the tissue. In contrast to prior case reports, our patient's symptoms and cutaneous findings responded nearly completely to vitamin C supplementation. The reason for this discrepancy remains unclear, but may pertain to the chronicity of findings, patient's age, or alternative factors. Clinicians should be aware of this uncommon presentation of vitamin C deficiency and have a low threshold for nutritional deficiency work-up when considering a differential that includes morphea profunda or eosinophilic fasciitis. Additionally, increased recognition of the higher risk of scurvy among patients who exhibit selective eating behaviors is essential, as early recognition and treatment can effectively manage this condition and prevent further complications.

Conflicts of interest

None disclosed.

REFERENCES

1. Carr AC, Maggini S. Vitamin C and immune function. *Nutrients*. 2017;9(11):1211. https://doi.org/10.3390/nu9111211
2. Pimentel L. Scurvy: historical review and current diagnostic approach. *Am J Emerg Med*. 2003;21(4):328-332. https://doi.org/10.1016/s0735-6757(03)00083-4
3. Hirschmann JV, Raugi GJ. Adult scurvy. *J Am Acad Dermatol*. 1999;41(6):895-910. https://doi.org/10.1016/s0190-9622(99)70244-6
4. Yule S, Wanik J, Holm EM, et al. Nutritional deficiency disease secondary to ARFID symptoms associated with autism and the broad autism phenotype: a qualitative systematic review of case reports and case series. *J Acad Nutr Diet*. 2021;121(3):467-492. https://doi.org/10.1016/j.jand.2020.10.017
5. Sharp WG, Berry RC, Burrell L, Scahill L, McElhanon BO. Scurvy as a sequela of avoidant-restrictive food intake disorder in autism: a systematic review. *J Dev Behav Pediatr*. 2020;41(5):397-405. https://doi.org/10.1097/DBP.0000000000000782
6. Swed-Tobia R, Haj A, Militianu D, et al. Highly selective eating in autism spectrum disorder leading to scurvy: a series of three patients. *Pediatr Neurol*. 2019;94:61-63. https://doi.org/10.1016/j.pediatrneurol.2018.12.011
7. Pan T, Hennrikus EF, Hennrikus WL. Modern day scurvy in pediatric orthopaedics: a forgotten illness. *J Pediatr Orthop*. 2021;41(3):e279-e284. https://doi.org/10.1097/BPO.0000000000001731
8. Grusin H, Kincaid-Smith PS. Scurvy in adult Africans. *Am J Clin Nutr*. 1954;2(5):323-335.
9. Schulz EJ, Swanepoel H. Scorbutic pseudoscleroderma: an aspect of Bantu siderosis. *S Afr Med J*. 1962;36:367-372.
10. Ahangari D, Gillard M, Certad G, Bataille M, Lasek A, Modiano P. Scurvy presenting as pseudo-scleroderma of the leg. *JAAD Case Rep*. 2019;5(9):831-833. https://doi.org/10.1016/j.jdcr.2019.07.019