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A Case Report of a Modern-Day Scurvy

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INTRODUCTION

Scurvy is a nutritional deficiency due to lack of vitamin C that can result in serious consequences.¹ The first written description of scurvy can be traced back to as early as 1500 BC by the Egyptians, and it mostly has disappeared in developed countries due to advances in nutrition and food supplements.²-⁵ Due to the rarity of scurvy, diagnosis often is delayed or missed completely, resulting in potentially serious complications and unnecessary workups.⁶ Recently, however, there has been a slight increase in the diagnoses of scurvy in hematology clinics around the country. Certain factors can predispose patients to develop this ancient disease. Scurvy is seen more often among people who live alone and prepare their own food, persons with alcohol use disorder, the elderly, and indigent persons.⁶ If sufficient vitamin C is not ingested through diet, the body's stores can deplete in as little as one to three months.⁶ Therefore, it is essential to have a steady dietary supply of vitamin C.

Vitamin C is essential for many basic functions. Vitamin C aids in collagen synthesis and helps with wound healing and bone formation, plays a role in the synthesis of the neurotransmitter norepinephrine, and increases iron absorption in the stomach.⁸⁻¹¹ Vitamin C also plays a role in the metabolism of prostaglandins, which in turn aids in the management of the inflammatory response by preventing the inhibition of the innate immune response.¹²⁻¹⁴ Furthermore, vitamin C has significant antioxidant properties, helping to protect against cellular damage.^{15,16}

The most common manifestations of vitamin C deficiency often present as hematologic, skin, and immunologic dysfunction. Bleeding gums, perifollicular hemorrhages, corkscrew hairs, and poor wound healing characterize some of the signs of a classic presentation of scurvy. Clinically, perifollicular hemorrhages are identified by small petechiae surrounding the base of the hair follicles and corkscrew hairs are thin wiry strands that can become kinked (Figure 1). Lack of vitamin C replacement can lead to disastrous consequences including hemolysis, seizures, and death. Prompt recognition and treatment is key as scurvy is an easily treatable disease. We report a case of scurvy in an 18-year-old male.

CASE REPORT

An 18-year-old male with a history of cerebral palsy, autism, and spina bifida presented to the emergency room with ankle pain and fever. The patient was nonverbal and seemed agitated at baseline, so the history was taken from the parents, who noted the young man fell three weeks prior and hurt his ankle. At the time, he was evaluated by his primary care physician and diagnosed with a mild ankle sprain. His ankle pain initially improved but swelling worsened during the past week. On the day of presentation, he developed fever, swelling in his ipsilateral knee, bleeding gums, and diffuse perifollicular hemorrhages across his lower and upper extremities bilaterally. Family history was pertinent for an unspecified bleeding disorder on his maternal side.

On general exam, he was not toxic appearing, but had a fever of 38.9°C and was tachycardic with a heart rate of 125 beats per minute. His blood pressure and oxygen saturation were within normal limits. There was mild clubbing of the nails of his lower extremities and splinter hemorrhages were present on the nails of his upper extremities bilaterally. Other pertinent aspects of the physical exam included tacky mucous membranes and evidence of dried blood on his gums. His right knee and ankle were mildly swollen and warm to the touch with pain on passive range of motion. The dermatologic exam revealed extensive perifollicular hemorrhages on the lower extremities bilaterally (Figure 1), ecchymoses were present on the plantar aspect of both feet, and the presence of petechiae on the back. Furthermore, corkscrew hairs were identified diffusely across the upper and lower extremities (Figure 1).



Figure 1. Photo shows perifollicular hemorrhages and corkscrew hairs (circled). [Reprinted with permission from Vogt KA, Lehman JS. Corkscrew hairs. Cleve Clin J Med 2015; 82(4):216. doi:10.3949/ccjm.82a.14135. Copyright © 2015 Cleveland Clinic Foundation. All rights reserved.]

Pertinent admission labs revealed a moderate iron deficiency anemia, a normal white blood cell count, and an elevated lactic acid of 3.2 mmol/L (reference range, 0.5 - 2.1 mmol/L). Pending labs at the time of admission included blood cultures, vitamin C level, vitamin D level, Factor 8, Factor 9, DIC panel, LDH, and haptoglobin. Plain films of the ankle and knee were unremarkable.

Prior to transfer to the medical floor, the patient received an IV bolus of normal saline, empiric antibiotics of vancomycin and cefepime, and he was started on high-dose vitamin C $(1,000 \, \text{mg} \, \text{daily})$ by the emergency room physician. This treatment regimen subsequently was continued by the admitting team. An orthopedic surgeon

was consulted to rule out a septic joint and discerned that no drainable effusion was present in either joint. The intravenous fluids, antibiotics, and daily vitamin C were continued upon admission. On hospital day two, a lower extremity venous ultrasound revealed no evidence of deep vein thrombosis. The patient did not have any further bleeding from the gums, his joint swelling was stable, and clinically he was improving. On hospital day three, the vitamin C level was < 0.01 mg/dL (reference range, 0.6 - 2.0 mg/dL) and the vitamin D level was < 3 mg/mL (reference range, 20 - 50 ng/mL). Lactic acid dehydrogenase, haptoglobin, DIC panel, Factor 8, and Factor 9 levels were normal. He continued to improve clinically and remained afebrile for more than 48 hours with no growth on blood or urine cultures, and antibiotics were discontinued. He was discharged home on oral vitamin C 250 mg daily and vitamin D 1,000 IU daily with follow-up scheduled.

DISCUSSION

This case was unique due to the combination of pathognomonic signs and the patient's atypical presenting symptoms. Certainly, the presence of bleeding gums, perifollicular hemorrhages, and corkscrew hairs increased the suspicion for vitamin C deficiency. However, the patient's fever and tachycardia were a major focus upon initial presentation. Treatment for suspected sepsis with antibiotics was initiated, but no infectious etiology for the fever was identified and antibiotics were discontinued prior to discharge. Fever and tachycardia do not appear to be a common presentation of scurvy in reported case studies. ^{6,7,17,18}

This case demonstrated the importance of a thorough history and physical examination. Though the fever and tachycardia were concerning, the emergency room physician's recognition of the constellation of symptoms typically associated with scurvy allowed for prompt intervention to be initiated without laboratory confirmation. This recognition and treatment were imperative for the patient's quick clinical improvement. Further, the patient's history helped pinpoint the diagnosis. The patient's nonverbal status complicated the history, but caregivers were able to provide important details. Importantly, the parents reported at baseline that the patient had a severe aversion to nearly all foods; his diet consisted almost exclusively of Pop-Tarts* which contain 0 mg of vitamin C.

Another complicating factor to this case was the ankle injury that occurred weeks prior. Symptoms had not resolved from that injury, so much of the history focused on the injury. Vitamin C is important for skin, hematologic, and immune function so it is reasonable to conclude that the deficiency may have led to longer wound healing. Was this a coincidence that the signs of scurvy happened to manifest during his recovery from this injury? It is possible that the patient barely was getting enough vitamin C to not manifest symptoms of scurvy, but the injury increased vitamin C demand beyond that minimal level of supply and tipped him over the edge into symptomatic scurvy.

CONCLUSIONS

Scurvy is an ancient disease that has been mostly eradicated. However, as this case demonstrated, certain lifestyle factors can predispose one to be susceptible, even in the 21st century. It is imperative to approach each patient with a broad differential and let the history and physical exam guide clinical decision making. Even 21st century physicians should be alert to the risk factors, signs, and symptoms of this ancient disease.

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scurvy continued.

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