Early scurvy in the modern era: A case series



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Key words: ascorbic acid; corkscrew hairs; nutritional deficiency; perifollicular hemorrhage; scurvy; water-soluble vitamins.

INTRODUCTION

Scurvy, or vitamin C deficiency, historically presented in those whose diet lacked fresh fruits and vegetables, classically including sailors and explorers. In modern times, scurvy is viewed as a rare condition; however, it continues to present even within highly developed nations due to malnutrition and unbalanced diets. According to trials by the National Health and Nutrition Examination Survey, the prevalence of vitamin C deficiency within the United States has reduced from 13% in 1988-1994 to 7.1% in 2003-2004. Ongoing cases of scurvy may be explained by the ongoing impact of the obesity epidemic. Additionally, alcohol and tobacco intake both impair ascorbic acid absorption, leading to a higher risk of scurvy.

Vitamin C is implicated in fatty acid regulation, immune function, neurotransmitter generation, prostaglandin metabolism, and nitric oxide synthesis. 4,5 Therefore, severe cases of scurvy may result in serious systemic functional deficits, and in untreated cases, even sudden death. Dermatologic findings associated with clinical scurvy include follicular hyperkeratosis, perifollicular hemorrhage, hair coiling (corkscrew hairs [CH]), and petechia. Mucosal findings include ecchymosis and gingivitis with recidivism and atrophy. 6 A high index of suspicion is necessary, as clinical recognition is often the first line in diagnosis and timely treatment of scurvy. Here we present 5 cases of patients presenting to a dermatology clinic for routine skin concerns, incidentally found to have the lower limit of normal or deficient vitamin C levels.

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Abbreviations used:

CH: corkscrew hairs
PE: perifollicular erythema

MATERIALS AND METHODS

Identified cases included patients presenting with dermatologic findings consistent with scurvy who were subsequently diagnosed with low to deficient Vitamin C levels. All cases were retrieved from patients seen by one of the authors (Dr Carly Dunn) at one of 2 clinical sites. In each case, dermatologic findings on exam were consistent with those of scurvy; therefore, serum ascorbic acid levels were collected. All patients with the lowest limit of normal or deficient vitamin C levels were included. Electronic medical records were reviewed to assess for patient and clinical characteristics including age, sex, dermatologic findings, affected sites, and treatment. Risk factors including dietary habits, tobacco, and alcohol use were assessed. Available skin biopsy results were reviewed in one case.

RESULTS

Five patients were identified based on our inclusion criteria. A summary of the clinical characteristics and risk factors of these patients is provided in Table I. All 5 patients were male with a median age of 67 (range, 61-74). Three patients presented to dermatology clinic for a periodic skin check, 1 patient for follow up of his acne, and 1 patient for a rash. Below, we present a summary of each case.

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Table I. Clinical characteristics

Case	Sex	Age (y)	Reason for dermatology visit	Skin findings (site)	History of easy bruising (Y/N)	Dietary restrictions	Other risk factors	Exogenous vitamin intake	Serum ascorbic acid level (mg/dL)	Clinical outcome
1	М	73	Periodic skin check	PE, CH (abdomen); E (forearms)	Υ	No fruit or vegetable intake	Smoking (81-pack years)	Vitamin D; folic acid	<0.1	Lost to follow up
2	M	62	Periodic skin check	PE, CH (upper back, outer thighs)	Υ	Fruit intake once weekly	-	Multivitamin; folic acid; B12	0.3	Skin findings present at 6 mo follow up visit
3	М	74	Rash	PE (Thighs, arms, abdomen, buttocks, mid-back)	Υ	Fruit intake once weekly	-	Multivitamin	0.4 (LLN)	Rash resolved within 2 wk prior to starting Vitamin C supplementation
4	М	67	Periodic skin check	PE, CH (back, chest)	Υ	Infrequent vegetable intake (once monthly)	Prior smoker (90-pack-years)	-	0.4 (LLN)	Skin findings present at 9-mo follow up visit. Patient did not start supplementation
5	M	61	Follow up for acne	PH	N	Infrequent citrus fruit intake	Periodic cigar smoking	Vitamin D	<0.1	Skin findings present at 10-mo follow up visit while still on supplementation

CH, Corkscrew hairs; E, ecchymosis; LLN, lower limit of normal; M, male; PE, perifollicular erythema; PH, perifollicular hyperpigmentation.



Fig 1. A, Perifollicular erythema and corkscrew hairs on the *upper* back. **B,** Corkscrew hairs (*white arrow*).

Case 1

A 73-year-old man with history of basal cell carcinoma presented to dermatology clinic for a periodic skin check. On physical exam, he was noted to have asymptomatic perifollicular erythema (PE) with CH on the lower abdomen and few ecchymoses of bilateral forearms. He reported easy bruising for many years but had been taking aspirin daily. He denied gum bleeding or swelling. He had an 81-pack year history of cigarette smoking but denied alcohol use. Dietary intake did not include any fruit or vegetables, with meals consisting of only meat and potatoes. Exogenous vitamin intake included solely vitamin D and folic acid.

A vitamin C level was collected and resulted as <0.1 mg/dL consistent with severe deficiency. The patient was started on ascorbic acid 500 mg daily for 6 months with recommendation to increase dietary intake of foods rich in vitamin C.

Case 2

A 62-year-old man with a history of cutaneous squamous cell carcinoma and a prior kidney transplant on immunosuppression presented for a periodic skin check. He reported dark spots under his toenails which he believed may have resulted after trauma. On exam, he was found to have PE and CH on the upper back bilaterally as well as the outer thighs (Fig 1). Blue to purple discoloration was present under the left fourth and fifth toenails consistent with subungual hematomas.

He denied gum swelling or bleeding and saw a dentist regularly. For the past few years, he experienced easy bruising of the arms and legs. Medications included aspirin and an immunosuppression regimen consisting of tacrolimus, mycophenolate mofetil, and prednisone. He denied any alcohol, tobacco, or drug use. He had been taking a



 $\textbf{Fig 2.} \ \ \text{Perifollicular erythema of the bilateral outer thigh}.$

multivitamin for the past 11 years in addition to vitamin B12 and folic acid. He denied any tobacco or alcohol use. His diet included vegetable intake multiple times per week but limited fruit intake, roughly once weekly.

An ascorbic acid level was ordered, returning in the deficient range at 0.3 mg/dL. The patient was started on ascorbic acid supplementation at 500 mg daily for 6 months in addition to dietary counseling.

Case 3

A 74-year-old man presented to dermatology clinic for an itchy rash on the lateral thighs, waist, and buttocks present for 2 months. He had been using triamcinolone 0.1% ointment and camphor-menthol lotion which reduced the itching but had not eliminated the eruption. Physical exam demonstrated folliculocentric erythematous macules and papules with central keratin plugging and excoriations on the bilateral outer thighs, bilateral arms, abdomen, outer buttocks, and mid-back (Fig 2). No CH were evident. The patient denied gum bleeding/swelling. He reported easy bruising, but his medications included

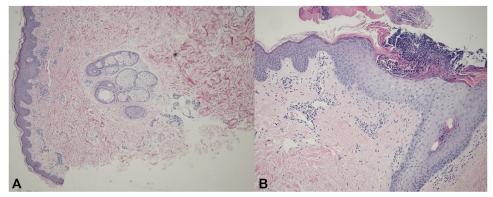


Fig 3. A, Case 3 punch biopsy showing superficial perivascular lymphocytic and neutrophilic infiltrates admixed with eosinophils. B, Epidermis with excoriated changes.

aspirin and clopidogrel. He took a multivitamin daily and denied tobacco or heavy alcohol use. He described daily vegetable intake and fruit intake once weekly.

Diagnoses of keratosis pilaris, scurvy, and folliculocentric mycosis fungoides were considered. A punch biopsy of the right abdomen demonstrated mild superficial perivascular lymphocytic and neutrophilic infiltrates admixed with eosinophils in the dermis and epidermal changes consistent with excoriation (Fig 3). Periodic acid-Schiff stain was negative for fungal organisms. The patient started ammonium lactate cream twice daily, and hydrocortisone 2.5% cream. At follow up 2 weeks later, he reported resolution of his pruritus with near resolution of his rash. At this time, his vitamin C level returned at 0.4 mg/dL, representing the absolute lower limit of normal. He was started on vitamin C supplementation at 500 mg daily for 4 weeks and advised to increase intake of vitamin C rich foods.

Case 4

A 67-year-old man presented to dermatology clinic for a skin check. On exam, he was found to have PE of the back and chest in addition to curly, cork-screw hairs. He denied any gum bleeding or swelling but endorsed easy bruising of arms and legs for the past few years. He denied alcohol use and reported a prior 90-pack year history of smoking, having quit 24 years ago. His diet included fruit intake 3 to 4 times weekly and infrequent vegetable intake, once monthly. He did not take any vitamin supplements.

A vitamin C level was ordered resulting at 0.4 mg/ dL, the absolute lower limit of normal. The patient was advised to avoid alcohol, incorporate more citrus fruit into his diet, and begin ascorbic acid 100 mg or 250 mg daily for a few months.



Fig 4. Hyperpigmented follicular-based papules on the forearm.

Case 5

A 61-year-old African American man presented to dermatology clinic for an acne follow up. He noted intermittent blackhead-like lesions on his arms. On physical exam, hyperpigmented, erythematous follicular-based papules were appreciated on bilateral forearms (Fig 4). Few compound hair follicles were present without evidence of any hair shaft changes. He denied gum bleeding/swelling or easy bruising. He took a baby aspirin and vitamin D daily. He has been drinking 2 ounces of hard liquor 3 times monthly for the last 2 years with periodic cigar smoking. His diet consisted of vegetable and fruit intake a few times weekly.

Differential diagnosis included keratosis pilaris versus vitamin C deficiency. The patient was initiated on topical amlactin twice daily. His vitamin C level returned as severely deficient at <0.1 mg/dL. He then began supplementation with vitamin C 500 mg daily for 12 months.

DISCUSSION

Scurvy is a diagnosis that is not commonly considered, particularly in industrialized countries. The source of nutritional deficiencies in highly developed nations may be a result of unbalanced and restricted diets. As our patients clearly demonstrate, the typical "malnourished and starving" presentation need not be present for clinical progression to scurvy, as manifest by somewhat subtle cutaneous signs.

Within our cohort of patients with very low to frankly deficient vitamin C levels, dietary habits varied greatly. Case 1 was the only patient to report no fruit or vegetable intake whatsoever. Cases 2 and 3 reported fruit intake once weekly, while case 4 had infrequent vegetable intake. While case 5 reported frequent vegetable and fruit intake, he did not eat citrus fruits and mainly ate cooked vegetables. Fruits and vegetables high in vitamin C include citrus fruits, strawberries, broccoli, and sweet peppers. Another factor to consider is the method of cooking/heating produce, as exposure to water and/or heat reduces levels of the water-soluble vitamin C.⁷ These cases demonstrate the importance of dietary counseling as a clinical tool in the prevention and treatment of scurvy, particularly in the extremely early stages of the disease.

The use of dietary supplements has continued to increase in favor within the United States. Within our cohort, 2 of the 5 (40%) patients reported use of a multivitamin daily without interruption. The presence of low vitamin C levels among our patients on a daily multivitamin calls into question levels of compliance or potential issues with absorption, and further emphasizes the need for regular dietary intake in addition to supplements alone.

In addition, rather than the mucosal findings and petechial bruising that are commonly associated with scurvy, the most pertinent presentation for our patients included perifollicular hyperpigmentation and CH. None of our patients reported mucosal symptoms of gum bleeding/swelling. Interestingly, case 2 presented with the most evident CH on exam; however, he was found to be just above the level of deficiency (0.4 mg/dL) rather than severely deficient (<0.1 mg/dL) as in cases 1 and 5. Furthermore, PE or hyperpigmentation was the only clinical sign present among all 5 patients and therefore may represent a particularly useful albeit overlooked diagnostic sign. Easy bruising was also reported by 80% of our

patients; however, this symptom was difficult to interpret due to concurrent use of blood-thinners. Clinical exam findings at last available follow up (Table I) demonstrated some ongoing skin findings in 3 patients. Therefore, the skin findings may not have been related to scurvy in all cases; however, it remains worthwhile to check vitamin C levels when there is concern as these patients were found to have low to deficient levels.

The importance of vitamin C in immune function has been well studied. Vitamin C has been implicated in leukocytic production, regulation, and differentiation, increase in complement protein levels, and apoptosis and autoregulation of spent immune cells. In older patients, vitamin C supplementation has been shown to protect against systemic infection and respiratory infection. Supplementation of antioxidant vitamins A, C, and E led to higher CD4 and CD8 T-cell counts as well as improved immune response to mitogen stimulation in elderly patients. Therefore, immunosuppressed patients, such as case 2, require timely diagnosis of vitamin C deficiency to support immune function and prevent increased infection risk.

Given vitamin C's antioxidant role within the body, inflammation and oxidative stressors including smoking, alcohol, and chronic diseases may lead to deficiency. ^{1,3} Older age may represent an additional risk factor for scurvy in our cohort as all patients were over the age of 60. This finding is thought to be secondary to greater oxidative demands as well as decreased gut absorption with older age. ^{12,13}

CONCLUSION

With the litany of dermatologic differentials that must be kept in mind, it is important to remember that scurvy is not a disease of the ancient past. Our patients emphasize the utility of a thorough skin exam, as the subtle dermatologic findings of PE and CH may present earlier than severe mucosal and systemic findings. The importance of speedy diagnosis is demonstrated by the rapid recovery and relative ease by which patients can be brought back to baseline homeostatic health. In addition, recognition of scurvy can allow treatment of co-occurring nutritional deficiencies and poor dietary habits. Therefore, we must remain vigilant and cautious to recognize scurvy during its early stages before progression to advanced disease.

Conflicts of interest

None disclosed.

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