Scurvy: a 21st century disease. A serious complication of a modern lifestyle

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Summary

Scurvy is a diagnosis seldom considered in the context of modern society. However, some research suggests that its incidence is greater than perceived especially in high risk groups. When supported by an appropriate medical history the clinical manifestations are pathognomonic. We describe a case that presented to the emergency department at a major tertiary hospital.

KEY WORDS: scurvy; vitamin C; nutrition; deficiency; ascorbic acid; petechiae.

Introduction

Scurvy is an ancient disease which continues to this very day. First alluded to as far back as 1550 BC, the link between ascorbic acid and scurvy was only definitively described in the 1930's (1). Though James Cook, English navigator and explorer, is often credited with seeing the relationship between fresh fruit and scurvy, a Portuguese navigator, Vasco da Gama, three centuries earlier noticed that eating oranges improved scurvy symptoms. Despite this, scurvy continued to be a frequent scourge equally affecting civilians during times of famine. Especially affected were impoverished groups such as prisoners and the socially isolated with so called 'land scurvy'. Since then, modern medicine, improved living standards and community awareness has resulted in a marked decrease in the prevalence of scurvy. Despite this, scurvy continues to present in the clinical setting though its perceived rarity leads to delayed detection. Reports from the United Kingdom suggest the prevalence of vitamin C deficiency of up to 46% in men and 35% in women amongst low-income and materially deprived households (2). Smoking and poor vitamin C intake were identified as additional risk factors. Cross-sectional research from the US with a much larger sample size supported the idea of widespread vitamin C deficiency though to a lesser extent (3). The authors report an overall deficiency prevalence of 7.1% but as high as 17.4% in low income subgroups.

Case report

A 32-year-old male smoker on a disability support pension for chronic back pain presented to the emergency department with a history of progressive malaise, lethargy and shortness of breath. He was primarily concerned about a sudden onset of bruising on his thighs in the absence of trauma. Ten days earlier he noted a rapidly progressing non-pruritic rash on his legs associated with limb swelling. He also complained of oral ulcers. On examination there was an extensive non-blanching non-palpable purpuric rash over the lower limbs and arms in a diffuse perifollicular distribution (Figure 1). There were significant ecchymoses bilaterally on the lateral aspects of the thighs that were tender to palpation and accompanied by localised oedema (Figure 2). There was evidence of 'cork screw' coiled body hair. Friable gums and haemorrhagic gingivitis with easy bleeding was noted on oral examination. The initial diagnosis by emergency staff was of vasculitis but later dermatologic review proposed a diagnosis of scurvy based on clinical examination. Further history revealed a diet comprised mostly of flavoured milk and absent of fruit or vegetables.

Blood examination revealed a significant microcytic anemia, Hb 59g/L (Reference range 135-175) with a congruent iron deficiency with a level of 2 umol/L (Ref. 8-30). Serum ascorbic acid was 22umol/L (Ref. 50-90). Histopathology of the perifollicular petechiae demonstrated a non-specific superficial and sparse lymphocytic dermal inflammatory infiltrate and no evidence of vasculitis (Figure 3).

The patient was transfused with 3 units of packed red blood cells and commenced on multivitamin replacement including thiamine and iron. For the first three days he was given a total of 3000mg of ascorbic acid daily over three divided doses, later titrated to 1000mg daily for two weeks. He was later seen as an outpatient that confirmed full resolution of his signs and symptoms with a corrected nutritional state.
Discussion

The role of vitamin C in collagen biosynthesis has been extensively investigated and explains the basis for the clinical manifestation of scurvy. It cofactors the hydroxylation of two enzymes that are integral to the assembly of the trihelical molecular arrangement of mature collagen (4). Deficiency produces derangement in collagen synthesis leading to soft tissue compromise. Blood vessel fragility due to defective collagen seen in scurvy leads to the perifollicular hemorhages, petechiae, ecchymosis and oedema most predominant in the dependent areas (5).

Vitamin C plays several roles in haemopoiesis, gastrointestinal absorption of iron and neurotransmitter synthesis (1). It is these roles that explain the anemia present in up to 75% of patients with scurvy and the non-specific symptoms of malaise, myalgia and fatigue often described (6). The anemia is typically normocytic but macrocytosis may occur in the context of concomitant folate deficiency (6). Our patient’s microcytic anemia is likely the result of the proven concomitant iron deficiency.
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Diagnosis of scurvy is largely clinical, based on dietary history and physical examination. Dermoscopy is a useful tool and findings include follicular hyperkeratosis, hemorrhages and ‘corkscrew’ hairs. Less frequent findings include an arrangement of orange and violaceous perifollicular halos (7). In addition, nails may reveal splinter hemorrhages and there may be evidence of alopecia and poor wound healing (8). Laboratory investigations may not be necessary to diagnose scurvy but can be useful to include or exclude the diagnosis. The findings are usually non-specific except for low serum vitamin C levels. A biopsy is not indicated unless history and examination are inconclusive. An extended nutritional screen including folate, B₁₂, zinc and thiamine is often warranted. Diagnosis of scurvy is greatly assisted by mindfulness of the risk factors that raise the pretest probability of the diagnosis. Groups at greatest risk include adults on low incomes with poor self-care, those with poor dentition or with a chronic or psychiatric diagnosis. Chronic alcoholism, drug abuse and smoking are also commonly associated with vitamin C deficiency. Smokers are at risk of deficiency >3 times as often as nonsmokers (3). Increased oxidative stress is suggested to increase vitamin C demand amongst smokers and also the elderly (9).

Modern social revolutions have resulted in lifestyle changes that have exposed wider groups to nutrition deficiencies. Many people opt for isolative indoor lifestyles composed of minimal exercise, solitary and sedentary entertainment and eating high caloric nutrient deficient convenience food. As in the case presented and others reported it is not uncommon to find adults consuming nutritionally deprived diets composed of flavoured milk and diet soda and devoid of nutrient rich food (10). Men seem to be the worst culprits. A recent summary from the Australian Institute for Health and Welfare (AIHW) noted that only 1 out of 20 (5%) Australian men eat sufficient fruit and vegetables with 54% not eating enough fruit and 92% not eating enough vegetables (11). Fruit and vegetable intake was highest amongst males aged over 65 years but lowest amongst those aged 25-34 years. Overall, adult males consumed less fruit and vegetables than women. Not surprisingly, men are more likely to be vitamin C deficient than women (2).

The goal of treatment is to replenish and maintain vitamin C body stores. Vitamin C is well absorbed at low doses and percentage of absorption decreases with rise in dose. It is not protein bound and is excreted by the kidneys. There is varying opinion on the dosing for acute replacement of vitamin C ranging from 300mg daily to 2000mg daily and no fixed opinion on duration (7, 12). There is no documented short-term risk of high doses of vitamin C though long term use has been shown to be associated with an increased risk of nephrolithiasis (13). Given that most cases of scurvy are associated with poverty, chronic alcohol use, unconventional dietary habits or social isolation a multidisciplinary approach needs to be employed both in an acute setting and in the community. Patient education is paramount and referral to a dietician may be useful. Community support services or even psychiatric referral may be required to assist adequate oral intake.

Conclusion

Scurvy is an old disease presenting in the present and needs to be considered in the differential diagnosis of ecchymoses and purpuric rashes. Awareness of its clinical manifestations and attention to ‘risk groups’ will assist prompt recognition and implementation of simple and definitive treatment. Today’s modern life-
style is leaving many exposed to vitamin deficiencies which have previously been overlooked. Patient education is paramount. Supplementation acutely and a modified diet is the gold standard treatment but a multidisciplinary approach may be required in ensure long-term treatment success.

References