

# Scurvy presenting as pseudo-scleroderma of the leg



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## INTRODUCTION

Scurvy is the result of vitamin C dietary deficiency. It is an old disease, brought to light during the Renaissance, affecting mainly sailors.<sup>1</sup> Today, scurvy is observed in developing countries touched by malnutrition. However, in industrialized countries, several cases have been reported in impoverished populations, isolated elderly individuals, and those addicted to drugs and alcohol.<sup>2,3</sup> Typical dermatologic signs include dry skin, follicular hyperkeratosis, perifollicular skin hemorrhage expanding into larger ecchymotic patches, corkscrew hairs, and hypertrophic hemorrhagic gingivitis. Here, we report 2 cases of chronic scurvy manifesting as a sclerotic ecchymotic edema of a lower limb.

## CASE REPORTS

### Case 1

A 42-year-old white man was referred to our hospital because of a swollen, ecchymotic, and indurated right leg that had developed over several months (Fig 1). The skin changes began in the right foot and extended to the knee. He also noted bruising of the left foot and lower portion of the leg, as well as decreased mobility of the knees. He smoked, consumed alcohol, and for several years ate only convenience foods, without any fruits or vegetables. Physical examination showed poor dentition but no gingivitis. He had ecchymotic legs bilaterally with induration on the right leg.

Blood tests showed systemic inflammation (C-reactive protein level, 77 mg/L; normal value, <5 mg/L) and anemia (hemoglobin level, 8.8 g/dL) without hyperleukocytosis. Doppler echography

and magnetic resonance imaging showed a nonspecific subcutaneous infiltration of the right thigh. Skin pathology was nonspecific. In this context of systemic inflammation and suspicion of cellulitis, amoxicillin/clavulanic acid 2 g 3 times/d was started. Despite the regression of local inflammation during antibiotic therapy, the sclerotic appearance of the limb persisted, and new purple infiltrated patches appeared on the left leg, where a few perifollicular purpuric lesions and twisted, coil-like hairs were successively observed (Fig 2).

As a result, scurvy was considered as a plausible diagnosis. Vitamin C levels were extremely low (<3  $\mu\text{mol/L}$ ; average range, 26-85  $\mu\text{mol/L}$ ), confirming our diagnostic hypothesis. Ascorbic acid supplementation was started orally (1 g/d) for a month. Within a few days, the disappearance of the newest lesions on the left leg was sudden. Nevertheless, the sclerotic texture of the right limb did not show any signs of improvement, despite its attenuation in color. The patient's mobility equally improved as a result of a quick but partial resolution of the swelling of the knees. One month later, a healthy left leg and a clear reduction of right leg edema were found during follow-up consultation. The discoloration and induration on the right leg did not seem to disappear. Oral supplementation was stopped, and the patient received advice about a proper diet.

### Case 2

A 57-year-old man was hospitalized in our university hospital with a purpuric eruption on the lower limbs and edema of the right calf that had

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**Fig 1.** Ecchymotic and edematous right leg.



**Fig 2.** A few perifollicular hemorrhages and corkscrew hairs.

evolved for several weeks. He presented with a large purple purpuric patch on the right leg associated with sclerotic edema and necrosis on the external side of the foot (Fig 3).

Similar to the first patient, multiple purpuric macules and corkscrew hairs could be noted after careful skin examination. This patient showed signs of gingivitis, and his oral hygiene was poor. He had a poor diet totally lacking in vegetables and fruits. Blood tests showed anemia (hemoglobin level, 8.2 g/dL), systemic inflammation (C-reactive protein level, 126 mg/L; normal value, <5 mg/L), and low ascorbic acid levels (12  $\mu\text{mol/L}$ ; average range, 26–85  $\mu\text{mol/L}$ ). Scurvy was diagnosed, and vitamin C supplementation started. The treatment allowed a rapid improvement of skin lesions despite the persistence of the sclerotic edematous texture on the right limb.



**Fig 3.** Ecchymotic, edematous right leg with necrosis on the lateral side of the foot.

## DISCUSSION

Ascorbic acid plays a role in iron absorption and the synthesis of collagen through hydroxylation of proline and lysine.<sup>4</sup> Scurvy will clinically manifest after 3 months of dietary deprivation.<sup>1</sup> Diagnostic confirmation is obtained by blood tests, which show decreased vitamin C levels (<10  $\mu\text{mol/L}$ ). Ascorbic acid deficiency is responsible for the fragility of the small vessels and wound healing disorders. These abnormalities account for classic scurvy signs such as follicular purpuric macules, ecchymotic purpura, and skin hemorrhages.<sup>5</sup>

Abnormalities on the hair follicle due to scurvy can be explained by the fragility of perifollicular collagen. The susceptibility of the lower limbs to hemorrhage is due to the increasing hydrostatic pressure on the small perifollicular vessels in the orthostatic position. Scurvy presenting with its typical skin changes is easy for dermatologists to diagnose. However, the presence of an atypical pseudo scleroderma-like appearance can be tricky and might persuade the dermatologist to consider other possible diagnoses such as linear scleroderma, eosinophilic fasciitis, and acrodermatitis chronica atrophicans. Considering the severity of the vitamin C deficiency of the patient in case 1, the absence of gingival manifestations was quite surprising. In general, the identification of gingival manifestations when scurvy is suspected is helpful in suggesting the right diagnosis.

In 1954, Grusin and Kincaid-Smith first described the scleroderma-like clinical presentation of chronic scurvy in 2 African patients among 30 persons affected by this disease.<sup>6</sup> They also described the persisting sclerotic aspect despite ascorbic acid supplementation. Similar to the cases reported here, the histology was not specific; it was characterized by perivascular inflammation with red blood cell extravasation and dermal hemosiderin deposits. A similar presentation was described in advanced forms of scurvy by Schulz and Swanepoel in a series of 3

patients.<sup>7</sup> The authors suggested that the scleroderma-like clinical presentation was a direct consequence of chronic blood extravasation into the dermis, hypodermis, and muscles. In their opinion, hemosiderin tissue deposits stimulate fibroblastic growth and trigger cutaneous sclerosis. Supporting this hypothesis are experiments by Golberg et al,<sup>8</sup> who studied histologic cutaneous damages induced by the subcutaneous injection of iron into mice. They witnessed siderophages infiltrating the dermis, generating a fibrous reaction. Chronic vitamin C deficiency may cause repeated minor cutaneous hemorrhages with hemosiderin extravasation, stimulating fibroblastic proliferation and, as a result, dermal sclerosis.

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