

Pain, purpura and curly hairs

J. D. Fleming,¹ B. Martin,² D. J. Card³ and J. E. Mellerio¹

Departments of ¹Dermatology and ²Dermatopathology, St John's Institute of Dermatology, Guy's and St Thomas' NHS Foundation Trust, London, UK; and ³Nutristasis Unit, GSTS Pathology, St Thomas' Hospital, London, UK

doi: 10.1111/ced.12118

Clinical findings

A 43-year-old man was referred to dermatology after presenting to the accident and emergency department with leg pain, swelling and rash. The purpuric rash had developed over the preceding 4 weeks. He had no history of trauma or recent illness, and was otherwise well at presentation. He had a history of schizoaffective disorder that was being treated with olanzapine, and he lived alone in sheltered accommodation. On direct questioning, he also complained about oral discomfort and tooth loosening over the previous few weeks, as well as leg swelling and bone pain.

On physical examination, there were large areas of purpura seen on the anteromedial thighs and pitting oedema to the mid calves bilaterally (Fig. 1). There was widespread perifollicular haemorrhage, and the hairs on the patient's legs were corkscrew in shape (Fig. 2). The skin was otherwise dry, and there was diffuse hyperkeratosis affecting the dorsa of the feet. In the mouth, there was redness of the interdental papillae.

Blood tests showed: haemoglobin 11.4 g/dL (normal range 13.0–18.0 g/dL), white blood cell count 14 ($4-11 \times 10^9/L$), neutrophils 11.5 ($1.5-7 \times 10^9/L$), sodium 128 mmol/L; (135–144 $\mu\text{mol/L}$), creatinine 102 $\mu\text{mol/L}$; 90–120 mmol/L and bilirubin 30 (1–22 $\mu\text{mol/L}$), and normal results for platelets $326 (150-400 \times 10^9/L)$ and potassium 4.8 (3.5–4.9 mmol/L). Liver function tests, coagulation parameters, autoimmune profile, vasculitic screen, urinalysis and chest radiography were normal.

Histological findings

On histological examination of a 4 mm punch biopsy taken from the patient's thigh, a dysmorphic hair follicle containing a corkscrew hair was seen, along with follicular hyperkeratosis and a perifollicular infiltrate consisting of lymphocytes only (Fig. 3). There was red blood cell extravasation in the superficial dermis, but no evidence of vasculitis.

What is your diagnosis?

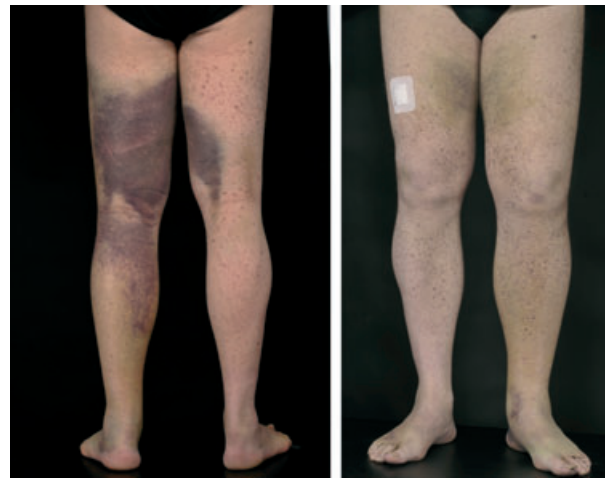


Figure 1 Leg purpura and left leg oedema.

Correspondence: Dr John D. Fleming, Dermatology Department, King's College London, Denmark Hill, London, SE5 9RS, UK
E-mail: john.fleming@gstt.nhs.uk

Conflict of interest: none declared.

Accepted for publication 20 November 2012

Diagnosis

Vitamin C deficiency (scurvy).

Discussion

Ascorbic acid (vitamin C) is an essential cofactor in the enzymatic step forming the collagen triple helix via hydroxylation of collagen peptides. Scurvy does not occur in most animals because they can synthesize their own vitamin C. However, humans lack the enzyme (L-gulonolactone oxidase) necessary for such synthesis, and must obtain vitamin C through their diet, with particularly high concentrations occurring in citrus fruits, tomatoes, potatoes, cabbages and green peppers.¹ In scurvy, the lack of hydroxylation of prolines and lysines causes a looser triple helix of procollagen, resulting in weaker tropocollagen and subsequent collagen fibrils. This in turn causes weaker capillaries, leading to erythrocyte leakage and bleeding.²

Body stores of vitamin C last for between 1 and 3 months. Symptoms of deficiency will therefore present after 3 months of dietary exclusion.³ Owing to financial constraints, our patient had limited his diet to biscuits and bread for the preceding 3 months, and his serum ascorbic acid level at presentation was undetectable. The purpura, oedema, leg pain and corkscrew hairs all returned to normal

after 2 weeks of oral ascorbic acid 500 mg four times daily.

The initial symptoms of scurvy are nonspecific, and include malaise, lethargy and loss of appetite, while the hallmark cutaneous signs comprise perifollicular hyperkeratosis and haemorrhage, ecchymoses, purpura, xerosis, leg oedema, poor wound healing, and corkscrew hairs.⁴ The ecchymoses and purpura seen in scurvy may lead physicians to suspect a cutaneous vasculitis. Examination of the mouth may identify gum abnormalities (occurring only in patients with teeth) which include gingival swelling, purplish discoloration, erythema and haemorrhage of the interdental papillae, collectively described as 'scorbutic gums'.⁵

Patients can present with painful joints, caused by haemarthrosis. Bleeding from the mucous membranes and resultant iron-deficiency anaemia is also seen.⁶ Myalgias may occur because of reduced carnitine production, and bone pain may be present because of reduced osteoid bone formation and subperiosteal haemorrhage.⁷ Psychosis, pseudoparalysis,^{8,9} dyspnoea and syncope can occur, and sudden death has been described from cerebral haemorrhage or haemopericardium,¹⁰ highlighting the importance of recognizing the hallmark cutaneous signs early, and starting vitamin C replacement. Scurvy, although a rare modern disease, seems to be becoming more prevalent in the developed world¹¹ and can present clinically with signs suggestive of a vasculitis.

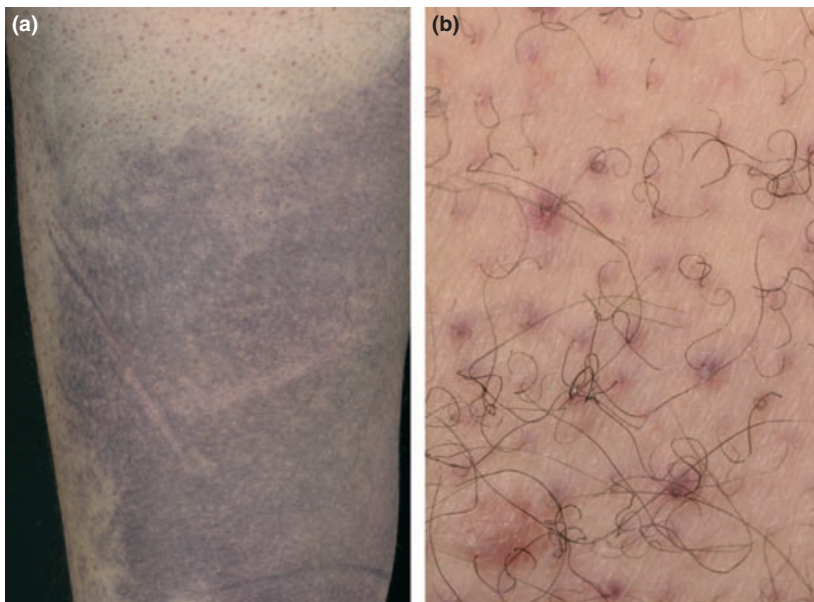


Figure 2 (a) Thigh purpura and (b) perifollicular haemorrhage and corkscrew hairs.

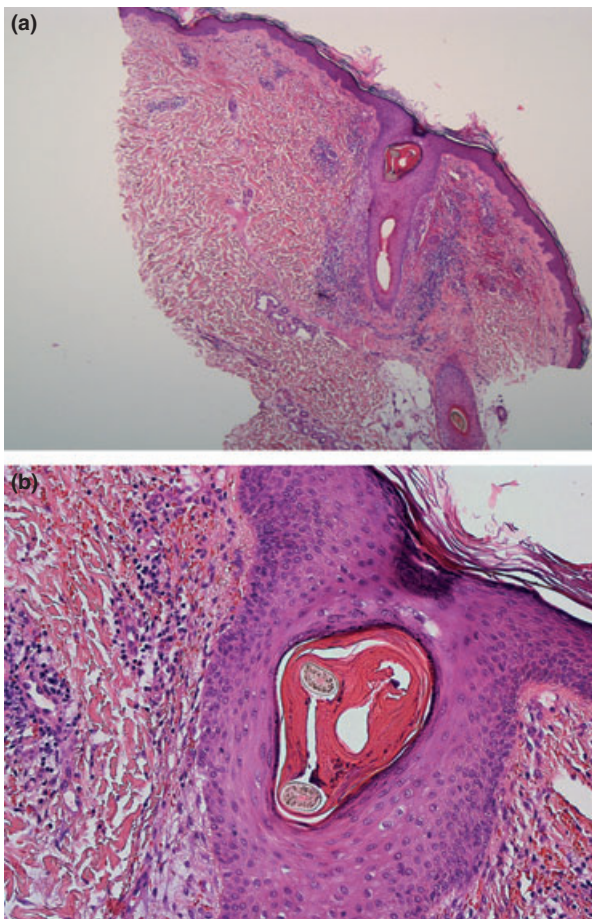


Figure 3 (a) A 4 mm punch biopsy of skin from the leg, showing a hair follicle in the centre with overlying follicular hyperkeratosis, and there is also a perifollicular infiltrate; (b) dysmorphic hair follicle with corkscrew morphology, perifollicular fibrosis, and an infiltrate comprising lymphocytes and extravasated red blood cells. Haematoxylin and eosin, original magnification (a) $\times 10$; (b) $\times 20$.

In summary, we describe a case of scurvy, presenting with all the hallmark cutaneous signs in a man with self-neglect secondary to a schizoaffective disorder and financial hardship. Dermatologists may be called upon to see patients with scurvy in the acute setting first. Rapid recognition of the clinical signs and instigation of vitamin C replacement can prevent life-threatening complications.

Learning points

- Leucocyte ascorbic acid levels have been replaced with serum levels in the UK.
- Scurvy, although a rare modern disease, seems to be becoming more prevalent in the developed world and can present clinically with signs suggestive of a vasculitis.
- Bone pain caused by subperiosteal haemorrhage may also be a presenting feature.
- Complications of untreated scurvy include haemarthrosis, haemopericardium and cerebral haemorrhage.
- Risk factors for scurvy include male gender, alcohol dependency, social isolation and financial hardship.

References

- 1 Gropper SS, Smith JL, Grodd JL. *Advanced Nutrition and Human Metabolism*, 4th edn. Belmont: Thomson Wadsworth, 2004; 260–75.
- 2 Hulmes DJ. Building collagen molecules, fibrils, and suprafibrillar structures. *J Struct Biol* 2002; **137**: 2–10.
- 3 Food Standards Agency. Food Standards Agency based guidelines for UK institutions. www.food.gov.uk (accessed January 2013).
- 4 Olmedo JM, Yiannias JA, Windgassen EB, Gornet MK. Scurvy: a disease almost forgotten. *Int J Dermatol* 2006; **45**: 909–13.
- 5 Masferrer E, Canal L, Alvarez A, Jucglà A. Gingival hypertrophy and anemia. *Arch Dermatol* 2009; **145**: 195–200.
- 6 Ho V, Prinsloo P, Ombiga J. Persistent anaemia due to scurvy. *N Z Med J* 2007; **120**: U2729.
- 7 Fain O. Musculoskeletal manifestations of scurvy. *Joint Bone Spine* 2005; **72**: 124–8.
- 8 Chisholm C, Brouha B, Lee P *et al.* Lower extremity purpura in a woman with psychosis – quiz case. *Scurvy. Arch Dermatol* 2010; **146**: 1167–72.
- 9 Arron ST, Liao W, Maurer T. Scurvy: a presenting sign of psychosis. *J Am Acad Dermatol* 2007; **57** (Suppl. 2): S8–10.
- 10 Spodick DH. Another cardiac disorder in scurvy. *N Engl J Med* 1970; **282**: 686.
- 11 Léger D. Scurvy: reemergence of nutritional deficiencies. *Can Fam Physician* 2008; **54**: 1403–6.