Pain, purpura and curly hairs

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

Departments of1Dermatology and2Dermatopathology, 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 × 10⁹/L), neutrophils 11.5 (1.5–7 × 10⁹/L), sodium 128 mmol/L; (135–144 μmol/L), creatinine 102 μmol/L; (90–120 mmol/L) and bilirubin 30 (1–22 μmol/L), and normal results for platelets 326 (150–400 × 10⁹/L) and potassium 4.8 (3.5–4.9 mmol/L). Liver function tests, coagulation parameters, auto-immune 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?
**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. 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.

Body stores of vitamin C last for between 1 and 3 months. Symptoms of deficiency will therefore present after 3 months of dietary exclusion. 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. 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'.

Patients can present with painful joints, caused by haemarthrosis. Bleeding from the mucous membranes and resultant iron-deficiency anaemia is also seen. Myalgias may occur because of reduced carnitine production, and bone pain may be present because of reduced osteoid bone formation and subperiosteal haemorrhage. Psychosis, pseudoparalysis, dyspnoea and syncope can occur, and sudden death has been described from cerebral haemorrhage or haemopericardium, 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 world and can present clinically with signs suggestive of a vasculitis.

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