A case of scurvy in Singapore in the year 2006

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ABSTRACT
Scurvy is an ancient disease. Over the years, with advances in the understanding of the disease, general improvement in health standards and nutrition, scurvy is now rarely encountered. The few cases of scurvy reported in the 21st century mainly occurred in the neglected elderly, alcoholics and food faddists. We describe scurvy due to food selection in a 37-year-old woman with underlying eating and obsessive-compulsive disorders. With vitamin C replacement, psychiatric medication and cognitive behavioural therapy, there was a dramatic improvement in her condition. This case serves as a reminder to the clinician that, even though rare in today’s practice, ascorbic acid deficiency is still encountered, and when recognised, is an easily treatable disease.

Keywords: ascorbic acid deficiency, eating disorder, obsessive-compulsive disorder, scurvy

INTRODUCTION
A patient with scurvy due to underlying psychiatric illness and eating disorder is described. This case highlights that even though uncommonly encountered nowadays, awareness of its manifestation helps in prompt diagnosis and treatment with a good outcome.

CASE REPORT
A 37-year-old Chinese woman, with no significant past history, presented on May 6, 2006, with swelling, redness and pain over both lower limbs for three months with no preceding trauma. The pain was made worse by movement and she had great difficulty walking. She also complained of a swelling over the proximal right forearm after a minor knock. On systemic review, she had lethargy for the past three months and unquantified weight loss over the last three to four years. There was no epistaxis or symptoms of gastrointestinal bleeding. There was no fever. Urinary and bowel movements were normal. She had no history of malar rash, arthralgia or arthritis. She was a non-smoker and teetotaller, and was not on any regular medications or health supplements.

On examination, her height was 157 cm, weight 36 kg, and body mass index (BMI) 14.6 kg/m². She was comfortable and alert. Vital parameters were stable. She had conjunctiva pallor and no jaundice. Jugular venous pressure was not elevated. She had dry, coarse long hair. Thinning of scalp hair was noted. Gums and tongue were normal. An evolving haematoma was seen over the right proximal forearm and full passive range of movement was detected. (Fig. 1) Bilateral mild pitting pedal oedema was noted. Petechiae and purpura were seen over the dorsum of both feet with exquisite tenderness on palpation (Figs. 2–4). There was no increased warmth or fluctuance, and movement was restricted because of pain. The right knee was swollen but not warm, with no demonstrable synovial effusion. Other symptoms included pain and limitation of movement of the right knee. There was no lymphadenopathy or abdominal organomegaly.

The initial investigations showed anaemia of haemoglobin 8.8 g/dl (normocytic and normochromic), white cell count was 3.85 × 10⁹/L and platelet 237 × 10⁹/L. Coagulation profile was normal, urea and electrolytes were normal, and liver function test was normal except for an albumin level of 31 g/L. Erythrocyte sedimentation rate was 40 mm/hr and C-reactive protein 17 mg/L. Radiographs of the right knee, feet and right elbow revealed no fractures (Figs. 5–8).
Clinical photograph shows right knee swelling and purpura on both feet.

Clinical photograph shows the diffuse purpura on the lower legs and dorsum of the feet.

Clinical photograph shows petechiae on the leg. On closer inspection, the haemorrhages were observed to be around the hair follicles.

Frontal radiograph of both knees shows no fracture.

Lateral radiograph of the right knee shows no fracture.

At this point in time, with the history and preliminary investigations, the possible differential diagnoses considered were Henoch-Schönlein purpura, connective tissue disorder and platelet dysfunction.

The autoimmune panel test results for anti-nuclear antibody, anti-double-stranded DNA, anti-cardiolipin antibody, anti-neutrophilic cytoplasmic antibody, rheumatoid factor, and complement levels were all negative or within normal range. Bleeding time and platelet function assays were normal. Hepatitis B and C serology, VDRL/TPHA and HIV serology were also negative. Thyroid function test was normal. Iron level was 4 μmol/L, TIBC 49 μmol/L, B12 and folate levels were normal.

She was referred to a dermatologist, who assessed her on May 9, 2006. After a review of the history and physical examination, a diagnosis of scurvy was made. The petechiae were noted to be perifollicular and the
purpura was non-palpable. The right knee swelling and pain were attributed to chronic haemarthrosis and the pain and tenderness over dorsum of both feet, due to subcutaneous and periosteal bleeding. The iron deficiency anaemia was consistent with the diagnosis of scurvy as well. The impression was vascular fragility due to scurvy.

She was started on oral vitamin C 500 mg twice a day. On obtaining further history, it was discovered that the patient had not been taking fruits and vegetables for years, and that she has some obsessive-compulsive behaviour. She stayed in a rented flat with her elderly mother. She had, however, not left the house for the last five years, as she felt “unclean” going out. She was perpetually worried about cleanliness and would wash her hands repeatedly for about half an hour each time and would bathe for about two hours. She would also eat only a meal a day to avoid having to wash her hands so often. She first started to worry about her weight at 17 years of age, and started taking smaller amounts of food. About five years ago, she stopped taking vegetables and fruits, as she deemed these too “cooling” for her and may cause her abdominal pain and diarrhoea. So for the past years, her main diet consisted of rice, fishcake and luncheon meat, which her elderly mother would buy from a nearby eatery. She weighed 55 kg at 18 years of age, but on admission at age 37 years, she weighed a mere 36 kg. She claimed that her menses were regular. On probing, she also complained of intermittent sadness for the past ten years, regretting quitting school when she was in secondary two and feeling that her future was bleak as a result.

She was referred to a psychiatrist, who started her on fluoxetine. During her stay in the psychiatric ward, the dietician, medical social worker and the psychologist saw her. She had multiple sessions of cognitive behavioural therapy and her meals were supervised. She accepted her new diet, which included fruits and vegetables, and was able to finish all her meals in the ward. She started to gain weight and on May 15, 2006 (ten days after admission), she weighed 40.9 kg. A bone mineral density study revealed osteoporosis with a T-value of -2.6 at the left femoral neck and -2.5 at the lumbar spine. She was hence commenced on calcium and vitamin D supplement. On review on May 16, 2006, the right elbow and knee swelling were subsiding and the perifollicular haemorrhages were reduced (Figs. 9–11). Upon review on June 6, 2006, her general condition and the cutaneous signs had improved remarkably.

DISCUSSION
This is an interesting case of scurvy presenting with easy bruising, right forearm haematoma, right knee haemarthrosis, perifollicular haemorrhages and non-palpable purpura. The patient was also found to have
iron deficiency anaemia and osteoporosis. Ascorbic acid deficiency is uncommon in current medical practice. However, it is still occasionally seen with inadequate intake of fruits and vegetables in the neglected elderly, alcoholics and those on faddish diets.\textsuperscript{15-16} Scurvy in patients with eating disorders, namely anorexia nervosa and bulimia, have also been described.\textsuperscript{5,6}

In our patient, it is the result of underlying psychiatric and eating disorders. The clinical signs and symptoms were a result of capillary fragility that occurs with ascorbic acid deficiency. The vitamin is required for collagen synthesis, acting as a cofactor for prolylhydroxylase, which catalyses the hydroxylation of proline and lysine in procollagen. In addition, vitamin C also aids in iron absorption and increases the conversion of cholesterol to bile acid and increases the bioavailability of selenium. Clinical manifestations are usually latent for months after onset of severe deficiency. The early symptoms include myalgia, malaise and weakness. Early signs include follicular hyperkeratosis, corkscrew hair and later on, patients develop perifollicular purpura and ecchymoses. Old wounds starts to break down, new wounds heal poorly; lower limb oedema may be seen; gingivitis and in severe cases, gingival necrosis occurs; and there were reports of Sjogren-like syndrome of keratoconjunctivitis sicca and xerostomia. Inflicted patients often have difficulty walking and may be incapacitated due to haemarthrosis and subperiosteal bleeding.\textsuperscript{7}

Serum ascorbic acid level and vitamin C urinary excretion tests are no longer available in Singapore.
In this case, the diagnosis was made with a meticulous review of the history and physical examination, and the remarkable clinical response to vitamin C replacement confirmed it. Recommendation for dosage of ascorbic acid for treatment of acute scurvy varies. 200 mg/day results in marked improvement in symptoms within several days, other authors suggest 1 g/day for the first 3–5 days followed by 300–500 mg/day for at least a week. Symptoms resolve in 3–5 days and physical findings in 1–2 weeks. Our patient had almost complete clearance of pain and limitation of movements, and significant improvement in the skin.

Scurvy, once a much dreaded and fatal disease, is now uncommon, but clinicians will still need to be on the alert, and keep nutritional deficiencies in our list of differential diagnoses. Nutritional illnesses can cause a myriad of physical signs which affect the skin, joints, bones, nails, hair, and eyes. Some signs are unique and hence diagnostic, whereas most others are not specific. For instance, follicular hyperkeratosis seen in scurvy can also be seen in vitamin A and essential fatty acid deficiencies; angular stomatitis can be observed in thiamine, riboflavin, pyridoxine, zinc, and folic acid deficiencies. In addition, nutritional deficiency may not occur singly. For example, a person whose diet is devoid of fruits and vegetables may also have folic acid deficiency, besides ascorbic acid deficiency. In this case, the constellation of signs of perifollicular haemorrhages, ecchymoses and haemarthrosis, together with a compatible history, led to the diagnosis of scurvy, even before the laboratory tests were performed to exclude other differential diagnoses. This case highlights the importance to diagnose and treat nutritional deficiencies promptly, as they are readily amenable to treatment.

REFERENCES