SIMULATED BLEEDING - A FORGOTTEN DISEASE IN A LAND OF PLENTY

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ABSTRACT

The human body is unable to synthesise Vitamin C and a diet deficient in Vitamin C leads to scurvy. Scurvy may mimic other medical conditions, like bleeding diasthesis or deep vein thrombosis, leading to delay in diagnosis and treatment, thus prolonging sufferings of patients. Often, scurvy could have been diagnosed if it is thought of and features of scurvy carefully looked for. Scurvy is easily treated with high dose of oral vitamin C. Recurrences may occur. Education of care providers cannot be overemphasised. We report three local cases of scurvy to highlight the existence of the disease in our modern society.

Keywords: *mentally defective, nutrition, bleeding, gum-hypertrophy, peri-follicular haemorrhages, corkscrew hairs*

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INTRODUCTION

Humans, primates and guinea pigs cannot synthesise vitamin C (ascorbic acid) due to the deficiency of the enzyme L-gloconolactone oxidase[1]. A diet deficient in vitamin C will lead to scurvy. It has been said that with the exception of famine, scurvy is probably the nutritional deficiency disease that has caused the most human sufferings. Scurvy plagued sailors on long ocean voyages and also appeared as a "disease of affluence" in the 1880s among infants receiving proprietary foods as substitutes for breast feeding[2].

Vitamin C has been isolated and synthesised in large scales since the 1930s[2]. As such, scurvy is uncommon in affluent societies. However, even in affluent Singapore, there is an unfortunate subgroup who are at risk of developing scurvy.

We report three patients with scurvy to highlight the existence of the disease in our modern society and hope to heighten the awareness of and to facilitate earlier recognition of this disease.

CASE REPORTS

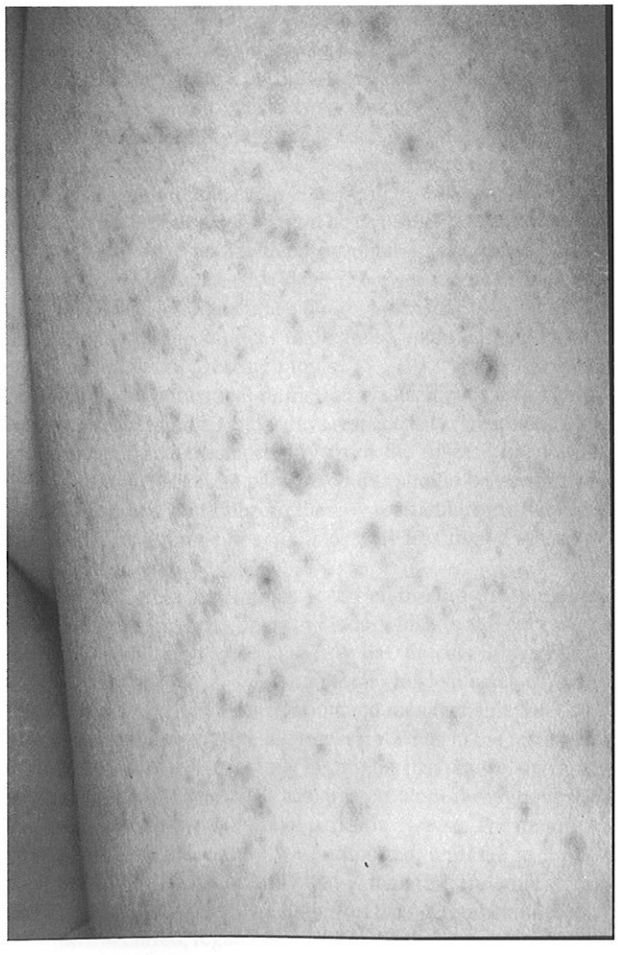
Case 1

YSC, a 32-year-old Chinese man, presented with a week's history of right thigh swelling, pain and spontaneous bruising with no preceding history of trauma. Two months prior to admission, he was noted to be lethargic and eating poorly. His meals generally consisted of bread with butter, and coffee with milk for breakfast, polished rice with little vegetables, canned meat or eggs for lunch and dinner. He had always been a slow learner, having completed primary education only and had previously worked as a storeman. He was single and stayed in an army cookhouse.

Clinically, he was pale and cachetic. He had gingivitis but the gums were not "spongy" or "bleeding". Soft tissue swelling and tenderness was noted over the medial right thigh with overlying ecchymosis. Peri-follicular haemorrhages and "corkscrew" hairs were noted over the limbs and the neckline.

Investigations revealed severe anaemia with a haemoglobin of 6.7 g/dL, a normal prothrombin time (PT) and activated partial thromboplastin time (PTT), and a platelet count of 684 x 10^9/L. Serum ferritin, folate, and vitamin B_{12} levels were normal. A vitamin C loading tests revealed marked ascorbic acid deficiency with only 4 µmol excreted in the urine over 2 hours after vitamin C loading (normal > 284). He was treated with high doses of vitamin C and haematinics. His thigh swelling improved gradually and resolved over a period of one week.

Fig 1 – Peri-follicular haemorrhages (Case 1)



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Case 2

TPK, a 41-year-old Chinese man, presented with a 3-day history of inability to walk or eat. Over the preceding 6 months he was noted to be restless and agitated in contrast to his usual timid behaviour. There was no history of falls or trauma. He had profound mental retardation and needed much assistance in the activities of daily living. He ate porridge and noddles with little meat and had no fruits or vegetables for his meals.

The patient had clinical features of Down's Syndrome. Multiple ecchymosis over the buttocks and peri-follicular haemorrhages over the limbs were noted. His gums were spongy and exhibited contact bleeding.

Investigations revealed a haemoglobin of 9.1 g/dL, MCV of 97 fL, total white count of 3.6 x 10^9/L and platelets of 110 x 10^9/L. Prothrombin time and partial thromboplastin time were within normal limits. Serum iron, TIBC serum folate and Vitamin B_{12} were normal. The vitamin C saturation test revealed a urinary vitamin C of 3 µmol/2 hours (Normal > 284).

Over a 6-day period, he improved with initiation of treatment with vitamin C. He defaulted follow-up subsequently. One and a half years after the initial presentation, he was readmitted with left lower limb pain and swelling and was initially suspected as having deep venous thrombosis. On closer scrutiny however, he had bruising in the posterior aspect of his leg, peri-follicular haemorrhages and corkscrew hairs. The vitamin C saturation test again confirmed the diagnosis of scurvy. He responded well to vitamin C therapy.

Fig 2 – Corkscrew hairs at the hairline and posterior neck (Case 2)

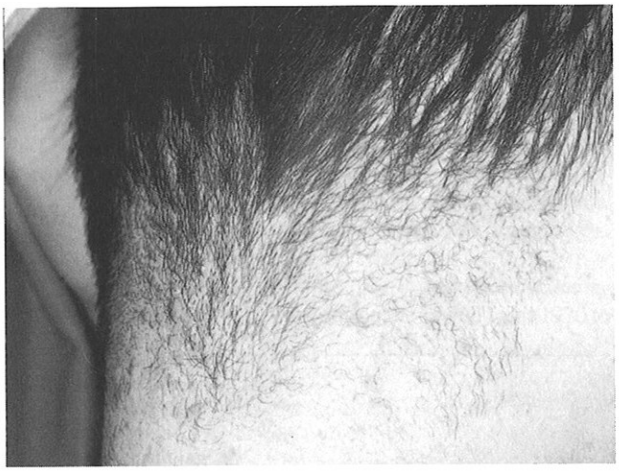


Fig 3 – Bruising at the inferior and medial surface of the thigh (Case 2)

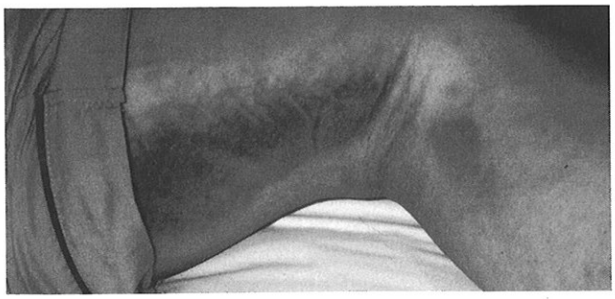
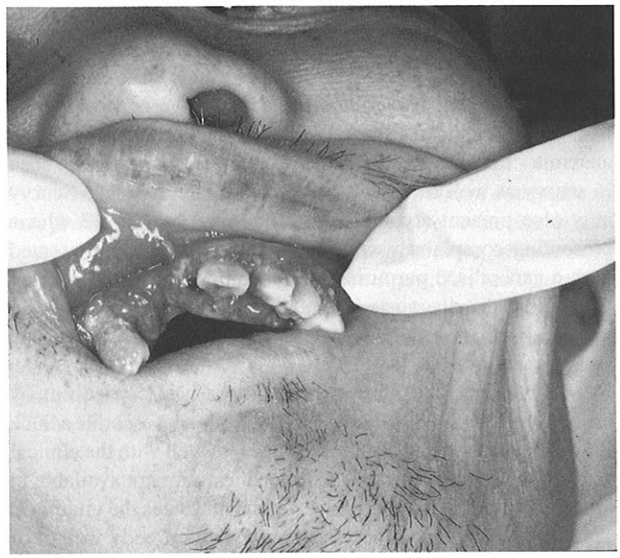


Fig 4 – Gingival hypertrophy (Case 2)



Case 3

CHL, a 42-year-old Chinese man with a past history of diabetes mellitus, hypertension and epilepsy requiring insulin, nifidepine and phenytoin respectively, presented with a generalised rash. Clinically, he had a generalised purpuric rash distributed predominantly over the lower limbs. He had no gum hypertrophy, corkscrew hairs or peri-follicular haemorrhages.

Investigations revealed a haemoglobin of 12.7 g/dL, MCV of 95.5 fL. The platelets count was 280 x 10^9/L. Prothrombin time and partial thromboplastin time were normal. The vitamin C saturation test was abnormal with urinary vitamin C excretion of 4 µmol/2 hours (Normal < 284).

He was treated with high doses of vitamin C and the purpuric rash improved over a few weeks.

DISCUSSION

Vitamin C is present in milk, some meats (kidney, liver, fish) and many fruits and vegetables. It is easily destroyed by cooking and prolonged storage. Though food rich in vitamin C is easily available in Singapore, there are many factors contributing to the persistence of scurvy. Food like milk, fish and liver are more expensive and may be avoided by the lower income group. Food fads and ignorance of food values may lead to consumption of vitamin C deficient foods like bread, canned food and polished rice.

The mentally retarded, demented and the elderly sick are often dependent on their carers. Either through neglect or ignorance of food values, non nutritious food may be regularly consumed thus making them at risk of developing a deficiency state. Some of these patients (as illustrated by Case 2) may be fussy about food or refuse food. These patients often present late because of their carer's fear of the stigma of having a mentally defective relative. After treatment, recurrence is not uncommon unless social circumstances are changed and the carers are educated about food values.

Substance abusers often self-neglect and may not have the appetite or money for nutritious food. Less commonly, food faddists may consume unbalanced diets leading to deficiency states.

Scurvy may mimic numerous other medical conditions such as vasculitis, systemic bleeding disorders and deep vein thrombosis[5]. It may present as a haemarthrosis[6], purpuric rash[7], refractory anaemia[8] and inability to walk[9]. Case 2 was initially diagnosed to have deep vein thrombosis. But closer examination revealed bruising, peri-follicular haemorrhage and corkscrew hairs characteristic of scurvy. The first two cases had documented anaemia - this may be attributed to the refractory anaemia seen in scurvy as well as other nutritional deficiency states. Scurvy may also present atypically, as illustrated by Case 2 whose presenting complaint was inability to walk. Case 3 presented with a generalised purpuric rash. It cannot be overemphasised that unless the diagnosis is thought of, scurvy may be easily missed. Furthermore, unnecessary investigations may be done and treatment delayed.

Laboratory confirmation of scurvy is best carried out by estimation of white cell ascorbic acid[3] or platelet ascorbic acid[1]. Plasma levels of the vitamins correlate less well with the clinical status. However, the above-mentioned tests are not available in most laboratories. An alternative diagnostic test is the vitamin C saturation test. Eleven milligram per kilogram body weight of vitamin C is given to the patient orally and the patient's urine sample from the 4th to the 6th hour is obtained. This sample is then analysed for its vitamin C content. Under normal circumstances, more than 284 μmol is excreted over 2 hours.

CONCLUSION

Scurvy is uncommon in Singapore, but cases still surface sporadically. It is most gratifying to diagnose and treat a case of scurvy. On the other hand, it is most embarrassing (for the doctor) and unfortunate (for the patient) should a classical case of scurvy be missed. Scurvy causes much suffering to those afflicted and yet it is such a treatable disease. It is important to recognise that recurrences may occur, and patients' and carers' education cannot be overemphasised.

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