CHRONIC ADULT SCURVY WITH UNCOMMON SKIN LESIONS

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Chronic scorbutic features as isolated deficiency were seen in an uneducated food faddist female. Unusual sclerodermatous and discolored lesions on both legs were accompanied by typical perifollicular haemorrhages, follicular hyperkeratosis, corkscrew hairs, hypertrophic spongy bleeding gums and arthritis. Lesions were reversible on administration of 1000 mg ascorbic acid a day.

Key words : Scurvy, Sclerodermatous lesions.

Scurvy has a long history beginning with significant mortality in long voyages, to the famous study of James Lind[1] in 1753 using citrus fruits for its cure, and final isolation of the active ingredient ascorbic acid by King and Waugh in 1932.[2] Even after six decades, ascorbic acid deficiency still occurs mostly as a part of multiple clinical deficiencies and rarely in isolated form because of poverty, ignorance, poor intake by psychiatric patients, elderly, alcoholics, infants and food faddists.[3,4] We report a case of ascorbic acid deficiency in an uneducated female who had food fads.

Case Report

A 30-year-old married female of poor socio-economic status had recurrent painful swellings, increasing discoloration on both legs and associated difficulty in walking for three years. She also complained of body-aches, swollen bleeding gums, painful swelling of right knee and lower legs for one month. Her husband claimed that she avoided eating vegetables and fruits as these nauseated her. She had been very depressed and ate very little, particularly during the last one month. Examination revealed pallor, hypertrophic bleeding gums (Fig. 1), poor oro-dental hygiene and foul oral odour. Right knee was swollen, hot, tender with limited movements and patellar tap. Multiple, discrete, dusky discolored and tender subcutaneous nodules, brawny, indurated and discolored lesions, isolated petechae with surrounding oedema were located on the legs. Perifollicular haemorrhages, follicular hyperkeratosis and corkscrew hairs were visible on the extensor aspects of thighs (Fig. 2) and arms. Nails showed multiple splinter haemorrhages.

Investigations revealed haemoglobin 10.8gm%, normochromic macrocytic, while blood counts, sedimentation rate, serum biochemistry including liver enzymes, serum proteins, renal function, uric acid and calcium were normal. Rheumatoid factor, ANF, LE cells and VDRL test were negative. Serum complement was within normal limits. Urine and stools were normal. Coagulation studies showed normal bleeding time, clotting time, Hess test, clot retraction, prothrombin time, prothrombin index, thrombin time and partial thromboplastin time with kaolin. Joint aspirate was haemorrhagic but sterile with normal WBC count, C3 concentration and negative rheumatoid factor. X-ray right knee showed a slight increase in the joint space. Skin biopsy revealed follicular keratotic plugging, normal dermis and scanty extravasated red cells around hair follicles. Following 1000 mg ascorbic acid daily, gum hypertrophy and bleeding cleared within three weeks. Fresh crops of nodular lesions and petechae were arrested though indurated discolored lesions took longer time to regress.

Comments

In a well controlled experimentally induced scurvy of human volunteers utilizing isotopic

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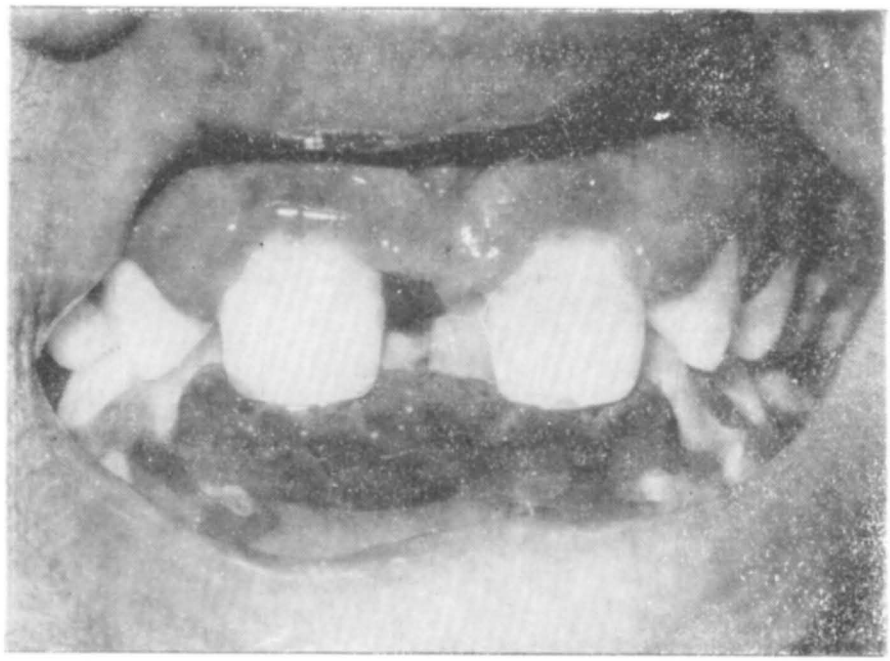

Fig. 1. Gum hypertrophy.

Fig. 2. Arthritis of right knee, follicular hyperkeratosis, corkscrew hairs and perifollicular haemorrhages.

techniques, multisystem manifestations such as petechial haemorrhages,follicular hyperkeratosis with coiled corkscrew hairs, perifollicular haemorrhages, arthritis, dependent oedema, hypertrophic spongy bleeding gums, peripheral neuropathy, sicca syndrome, occular haemorrhages,mild anaemia,general aches and impaired vascular reactivity were recorded.[5] During deprivation, personality changes of fatigue, lassitude, classical neurotic triad of hypochondriasis, depression and hysteria were found earlier than psychomotor changes.[6] Walker[7] noted painful, woody indurated lesions, oedema and discoloration of legs in seven patients. Similar lesions were found in the patient under report and are presumably the result of extravasated blood into the tissues.

Scurvy appears when the large ascorbic acid body pool of 1500 mg falls below 300 mg and whole blood level below 0.3 mg/100 ml.[8] The body stores of ascorbic acid in a healthy man are utilized at an average rate of 3% of the existing pool per day. Thus, after a period of three

months of deprivation, the stores become largely depleted leading to frank scurvy.[8] Late appearance of scurvy in our patient could be because of consumption of smal amounts of vitamin C, but the balance was completely tilted on account of her recent refusal to eat at all. Plasma levels of ascorbic acid have poor correlation with pool size, whole blood level and clinical sings of deficiency.[8] The minimal amount of ascorbic acid necessary to prevent scurvy is less than 10 mg daily.

The mechanism of action of ascorbic acid remains obscure, albeit as a known cofactor in certain enzymes such as in the hydroxylation of proline to hydroxyproline, an important step in the biosynthesis of collagen and chondroitin sulfate of the ground substance.[4,9] Electron microscopy has revealed discontinuous and ballooned endothelial cells with diminished perivascular collagen support which accounts for the haemorrhages and oedema.[9] Follicular hyperkeratosis and corkscrew hairs could be due to abnormal cross linking of keratin.[4] Most of the features of chronic scurvy including rare painful sclerodermatous woody discolored lesions closely simulate cellulitis and vasculitis, though follicular hyperkeratosis and petechial lesions are the most pathognomic features.[4]

References

1. Hughes RE : James Lind and the cure of scurvy. An experimental approach, Med Hist, 1975; 19 : 342.
2. King GC and Waugh WA : Chemical nature of vitamin C, Science, 1932; 75 : 357.
3. Connelly TJ, Decker A and McDonald JW : Bachelor scurvy, Intern J Dermatol, 1982; 22 : 209.
4. Warshauer DM, Hayes ME and Shumer SM : Scurvy, a clinical variant of vasculitis, Cutis, 1984; 39 : 539-541.
5. Hodges RE, Hood J, Canham JE et al : Clinical manifestations of ascorbic acid deficiency in man, Amer J Clin Nutr, 1971; 24 : 432-443.
6. Kinsman RA and Hood J : Some behavioural effects of ascorbic acid deficiency, Amer J Clin Nutr, 1971; 24 : 455-464.
7. Walker A : Chronic scurvy, Brit J Dermatol, 1968; 80 : 625-630.
8. Baker EM, Hodges RE, Hood J et al : Metabolism of C14 and H3 labelled L-ascorbic acid in human scurvy, Amer J Clin Nutr, 1971; 24 : 444-454.
9. Hashimoto K, Kitabchi AE, Duckworth WC et al : Ultrastructure of scorbutic human skin, Acta Dermato-Venereol, 1970; 50 : 9-21.