THE OLDEST NUTRITIONAL DEFICIENCY DISEASE: A CASE REPORT OF SCURVY

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Abstract

Scurvy resulting from dietary deficiency of vitamin C is characterized by anaemia, tender limbs swelling and hemorrhagic manifestations such as petechiae and bruises. We report a case of scurvy in a 13 years old boy with cerebral palsy who presented with history of limbs bruising and swelling for 2 months. On examination, he was cachexic and pale with poor dentition and swollen gum. There were extensive bruises and swollen limbs as well. His platelet count and coagulation profile were normal. Limbs X-ray revealed classical scorbutic changes in the long bones. Serum ascorbic acid level was low <5 µmol/L (normal range 28-120 µmol/L) and he was successfully treated with vitamin C for 3 months. Though scurvy is now rarely seen in modern times, it is important to keep in mind that certain vulnerable populations with compromised nutrition are at risk. A thorough dietary history is helpful in clinching the diagnosis. It is important for clinician to recognize the disease as it is potentially fatal but can be easily reversible.

Introduction

Scurvy was the first vitamin deficiency disease ever to be described by Sir James Lind in 1753, who introduced citrus fruits to his seaman with scurvy. However, ascorbic acid was only isolated later in 1928 by Dr. Albert Szent-Györgyi [1,2].

Due to improvement in the nutritional and socioeconomic status of most populations in modern times, scurvy is now rarely seen. However, children suffering from neurodevelopmental disabilities are prone to vitamin C deficiency due to various factors, such as poor oral intake, oral motor dysfunction, feeding problems, non-ambulatory status and use of antiepileptics.

As the clinical features of vitamin C deficiency may overlap with various systemic diseases, one can easily miss the diagnosis leading to extensive laboratory and radiographic testing causing unnecessary delays in diagnosis and treatment.

We report a case of scurvy in a 13 years old Chinese boy with cerebral palsy who presented with extensive limb bruising and swelling to our outpatient clinic.

Case Report

A 13 years old Chinese boy with dystonic cerebral palsy and epilepsy was admitted to the pediatric ward due to extensive limbs bruising and swelling for 2 months. There was no preceding history of trauma, fever or altered consciousness. He was on long term phenobarbitone for his epilepsy. He was taken care of by his mother at home and he
On admission, he was severely malnourished, restless and pale. He had petechial rashes and hyperkeratosis over his forehead and lower limbs. He had poor dentition with swollen gums. There were extensive bruising and tenderness over all four limbs.

A detailed dietary history revealed a restrictive diet profile of mainly minced meat and fluids such as condensed milk. He was not given any vegetables or fruits.

**Figure 1. Follicular hyperkeratosis and perifollicular hemorrhages**

**Figure 2. Inflammed gingival with easy gum bleeding**

**Figure 3 and 4. Extensive bruises and diffuse swelling of bilateral upper limbs**
Blood investigations revealed a low hemoglobin of 5.5 g/dL, total white cell count of $4.2 \times 10^9/L$, normal platelet count of $233 \times 10^9/L$. His renal function was normal except for low serum sodium of 117 mmol/L, other electrolytes were normal. The coagulation profile were within normal range.

X-ray of the long bones (figure 5) showed classical scorbutic changes which include osteopenic changes with white line of Fraenkel (a thickened white line at the metaphysis) and a characteristic zone of rarefaction proximal to the white line of Fraenkel (Trumerfeld zone).

Serum ascorbic acid done showed a level < 5 µmol/L (normal range 28-120 µmol/L). He also had low serum iron of 8.4 µmol/L and low serum zinc level of 7.4 µmol/L (normal range 11-22 µmol/L).

Figure 5. Scorbutic changes in patient’s right elbow radiograph. Noted the ground glass appearance of the shaft of long bone suggestive of (A) osteopenia. There was a (B) thickened white line at the metaphysis (White line of Fraenkel) with the characteristic zone of rarefaction, the (C) Trumerfeld zone, showing a linear break of the bone proximal to the white line representing area of debris of broken-down bone trabeculae and connective tissue.
Ascorbic acid (Vitamin C) supplement was initiated at 500mg twice daily for 3 days then 300mg daily for 2 weeks and maintained on 100mg daily for 3 months. By the 3rd week of treatment, his limb swelling and bruises had markedly improved. 2 months later, his skin lesions and limb swelling had completely resolved.

**Discussion**

Vitamin C deficiency results in the clinical presentation of scurvy, the oldest recognized nutritional deficiency disease. Although a rare disease, scurvy is still common in children with cerebral palsy as they subsist on predominant milk based diets due to poor intake, oral motor dysfunction and feeding problems.

Capillaries are fragile in scurvy patients and there is tendency of bleeding. The initial manifestations are non-specific (Table 1) such as irritability, anorexia, and low grade fever. Dermatological findings include petechiae, and follicular hyperkeratosis with perifollicular haemorrhages. As the disease progress, patients develop gingival involvement where their gums become swollen, loosen and easy bleeding leading to poor dentition. Musculoskeletal symptoms usually occur in the later stage of the disease but it is the most common reason that leads the child to medical attention. Progressive limb swellings and joint pain cause further immobility.

Advanced and untreated scurvy may lead to severe complications such as bleeding, poor wound healing and death [3].

**Table 1. Clinical manifestations of scurvy in infants and young children** [4] (World Health Organization, 1999)

<table>
<thead>
<tr>
<th>Most frequent symptoms</th>
<th>Possible symptoms</th>
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<td>General irritability</td>
<td>Anorexia</td>
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<tr>
<td>Tenderness of the limbs, especially of the legs</td>
<td>Low-grade fever</td>
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<td>Pseudo paralysis, usually involving the lower extremities</td>
<td>Mild diarrhoea, sometimes bloody</td>
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<td>Involvement of costochondral junctions: changes such as beading of ribs</td>
<td>Petechial haemorrhages in the skin</td>
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<td>Haemorrhage around erupting teeth (in infants without teeth gums appear normal)</td>
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<tr>
<td>Anaemia</td>
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The diagnosis of scurvy can be made based on meticulous dietary history taking, classical clinical features with radiological findings and clinical improvement following treatment with vitamin C [5]. Once the diagnosis is made, vitamin C supplements of 100-200mg/day should be initiated. The clinical improvement is seen within a week in most cases; spontaneous bleeding and constitutional symptoms begin to improve within days while bony changes and ecchymosis may take several weeks to resolve. Treatment should be continued for up to 3 months to ensure complete recovery [6,7].

**Conclusion**

Although scurvy is now rarely seen in modern times, it is important to keep in
mind that vulnerable populations with compromised nutrition are at risk. There should be a high index of suspicion in these groups of susceptible populations. A thorough dietary history, characteristic clinical features and a confirmed low serum ascorbic acid level is helpful in clinching the diagnosis. It is important for clinicians to recognize the disease as it is potentially fatal but can be easily prevented and reversible on prompt treatment.

References


