

Scurvy: Presenting as Musculoskeletal Pain

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ABSTRACT

The incidence of scurvy in the paediatric population is very low. This report describes a case of scurvy in toddler girl who was admitted with severe pain in legs and difficulty in walking along with bleeding from gums. After extensive investigation, she was found to have vitamin C deficiency resulting from restrictive eating pattern. There was significant improvement in presenting symptoms and general health after introduction of vitamin C and fresh fruits and vegetables to her diet. Nutrition-based diseases can still occur in any socioeconomic setting. Nutritional screening remains an important part in every child's general health care.

Key words: Scurvy. Vitamin C deficiency. Children. Nutrition.

INTRODUCTION

Scurvy results from a deficiency of ascorbic acid (vitamin C). One of the earliest documentations was by Jacques de Vitz in 1218, which described epidemics among population affected by war. Scurvy was often seen in sailors on long ocean voyages described from the 15th century onwards.^{1,2} Many men died from the disease until it was discovered that scurvy could be effectively cured and prevented by consuming vitamin C through lemons, oranges and limes. A marked reduction in the occurrence of scurvy has occurred over the last century due to improved knowledge about the pathophysiology and treatment of scurvy. However, the disease still exists even in industrialized countries.^{3,4} Delay in the diagnosis of an uncommon disease, even when patient presents with typical clinical features, might be caused by lack of awareness of the disease.⁵

This report describes a girl child who presented with typical findings of scurvy.

CASE REPORT

A 4-year-old girl presented with refusal to walk secondary to pain in her lower extremities, generalized irritability, low-grade fever, malaise and bleeding from gums. Her parents complained that the pace at which she was able to move around was significantly reduced even a year before she was afflicted with the illness. Her condition gradually worsened over the last few months leading to total inability to walk and consequently became bedridden. The parents apprised of having fed the patient with diluted cow's milk and routine home-

based diet devoid of fruits and vegetables. Physical examination revealed moderately unwell and very irritable girl having pale, bloated skin with edematous, violaceous gums and loosening of a few teeth. The patient's weight was below the third percentile for her age group. Dermatologic finding included pallor and xerosis. There was no evidence of petechiae, bruising or bleeding from any other mucosal site. Moreover, the patient had severe pain in both legs, which were tender to touch. The rest of neurological examination was normal. There was no evidence of hepatosplenomegaly or lymphadenopathy.

Diagnosis initially considered included hematological malignancy, chronic osteomyelitis or juvenile rheumatoid arthritis. The laboratory data results were as follows: hemoglobin (5.2 g/dL), white blood cell count of 9,500/ μ L, neutrophil count of 45%, lymphocyte count of 55%, and platelet count of 328,000/ μ L. The erythrocyte sedimentation rate was 30 mm first hour, the C-reactive protein level was 6.05 mg/dL. Peripheral smear revealed microcytic hypochromic anemia. The metabolic profile, PT/APTT and bone marrow biopsy was also normal.

Information was obtained from her parents regarding her eating patterns. It was revealed that she had developed restricted dietary habits from last 2 years that became more severe few months prior to admission. At presentation, her diet consisted of diluted milk, bread, biscuits and water, devoid of any kind of fresh fruits or vegetables. This showed that her diet was deficient in many nutrients including vitamin C. Nutritional deficiency, particularly of vitamin C, became an additional diagnostic consideration.

The radiographs of both knees showed osteopenia, with a thick sclerotic metaphyseal line of increased density, a transverse metaphyseal line of decreased density (scurvy line) above a widened physis, small beak-like excrescences at the metaphysis of both femora, and

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Figure 1: Anteroposterior radiograph of both knees shows a thick sclerotic metaphyseal line above a widened physis, small beak-like excrescences at the metaphysis in both femora and epiphyseal shell with a central lucency.

epiphyseal shell with a central lucency (Wimberger's sign of scurvy, Figure 1). The combination of evaluation confirmed the diagnosis of scurvy. The patient showed dramatic improvement after only 7 days of treatment with oral ascorbic acid and dietary modification. Follow-up radiographs also showed healing with continued dietary rectification.

DISCUSSION

This case highlights the importance of taking a detailed dietary history when evaluating diseases involving the musculo-skeletal system and bleeding tendencies.

Normal collagen synthesis depends upon the hydroxylation of proline and lysine residues in the endoplasmic reticulum to form hydroxyproline and hydroxylysine, respectively. Prolyl and lysyl hydroxylase, the enzymes that catalyze the hydroxylation, require ascorbic acid (vitamin C) to function correctly. Humans cannot synthesise vitamin C, therefore, they require an exogenous source for daily metabolic requirements. With no ascorbic acid available, the enzymes cannot hydroxylate proline and lysine, and so normal collagen synthesis cannot be performed.

Although, the incidence of scurvy is extremely rare in industrialized countries,⁶ it is still present in economically disadvantaged populations with poor nutrition including elderly persons living alone and alcoholics.^{2,5} Scurvy has historically been less frequent in the paediatric population. Infants who are fed evaporated or boiled milk, in which ascorbic acid is easily destroyed by heat as well as children with poor diet, as a result of psychiatric or developmental disorders, are at risk.⁷ Scurvy is one of the accompanying diseases of malnutrition. In this patient, scurvy resulted from poor oral intake during last 2 years due to poor dietary habits.

Symptoms develop after weeks to months of vitamin C depletion. Lassitude, weakness, irritability, weight loss, and vague myalgias and arthralgias may develop early. Follicular hyperkeratosis, coiled hair, and perifollicular hemorrhages may develop. Gums may become swollen, purple, spongy, and friable; they bleed easily in severe deficiency, as it was seen in this patient.^{3,5} Eventually, teeth become loose and avulsed. Secondary infections may develop. Wounds heal poorly and spontaneous hemorrhages may occur, especially as ecchymosis in the skin of the lower limbs or as bulbar

conjunctival hemorrhage. Musculoskeletal manifestations are present in 80% of patients with scurvy.^{4,6,8,9} Moreover, bone disease is a more frequent manifestation of the condition in children than adults, as seen in this patient.

Diagnosis is usually made clinically in a patient who has skin or gingival signs and is at risk of vitamin C deficiency. Laboratory confirmation may be available. Anemia is common.

Skeletal X-rays can help to diagnose childhood but not adult scurvy. Changes are most evident at the end of long bones, particularly at the knee. The radiographic findings of paediatric or infantile scurvy include a transverse metaphyseal line of increased density, (scurvy line), metaphyseal excrescences of the beaks, sub-epiphyseal infractions, increased density of periostitis and epiphyseal shell with a central lucency (Wimberger's sign of scurvy).^{4,8} The radiographic findings including osteopenia, thick sclerotic metaphyseal line, metaphyseal excrescences of beaks and Wimberger's sign were observed on the radiograph of this patient. The resolution of the metaphyseal abnormalities after vitamin C supplementation was also consistent with radiographic findings of the healing stage of scurvy.

A low vitamin C level in the plasma is specific for the diagnosis of scurvy; however, this is not always a reliable indicator because plasma levels may be normal with recent intake of ascorbic acid. Measuring vitamin C levels in the Buffy-coat of leukocytes better reflects the body stores; however, this method is technically more difficult.² Ascorbic acid levels were not available in this patient and it was described to treat on clinical suspicion and radiological findings.

The best evidence for scurvy is the resolution of the manifestations of the disease within few days after treatment with ascorbic acid as occurred in this patient.⁷ Because of the extremely rare occurrence of scurvy in modern society, it is difficult to differentiate it from other diseases. The diagnosis of scurvy is made by clinical and radiographic findings and may be supported by additional findings such as reduced levels of vitamin C in the serum or Buffy-coat of leukocytes, followed by clinical improvement in symptoms with vitamin C. For scurvy in children, ascorbic acid 100–300 mg orally is given for 1–2 weeks, until signs disappear, and followed by a nutritious diet supplying 1–2 times, the Daily Recommended Intake (DRI). In scurvy, therapeutic doses of ascorbic acid restore the functions of vitamin C in a few days. The symptoms and signs usually disappear over 1–2 weeks.

Worldwide, children presenting with pain, limp, weakness and apparent multi-system disease present a diagnostic challenge and are often investigated extensively. It is not widely appreciated that such

symptoms may be a common presentation of vitamin C deficiency in children and this disease is still seen in developed countries also. Clinicians and radiologists must be aware of this extremely rare but still present condition, because it is potentially fatal and can easily be cured with vitamin C supplementation. We suggest that in any child presenting with musculoskeletal manifestation, the possibility of nutritional cause, particularly scurvy, secondary to abnormal eating pattern should be considered before undertaking extensive investigations.

REFERENCES

1. Rajakumar K. Infantile scurvy. a historical perspective. *Pediatrics* 2001; **108**:E76.
2. Pimentel L. Scurvy: historical review and current diagnostic approach. *Am J Emerg Med* 2003; **21**:328-32.
3. Olmedo JM, Yiannias JA, Windgassen EB, Gornet MK. Scurvy: a disease almost forgotten. *Int J Dermatol* 2006; **45**:909-13.
4. Fain O. Musculoskeletal manifestations of scurvy. *Joint Bone Spine* 2005; **72**:124-8.
5. Akikusa JD, Garrick D, Nash MC. Scurvy: forgotten but not gone. *J Paediatr Child Health* 2003; **39**:75-9.
6. Halligan TJ, Russel NG, Dunn WJ, Caldroney SJ, Skelton TB. Identification and treatment of scurvy: a case report. *Oral Surg Oral Med, Oral Pathol Oral, Radiol Endod* 2005; **100**:688-92.
7. Weinstein M, Babyn P, Zlotkin S. An orange a day keeps the doctor away: scurvy in the year 2000. *Pediatrics* 2001; **108**: E55.
8. Choi S, Park S. Scurvy: a case report. *Korean J Radiol* 2007; **8**: 443-7.
9. Tamura Y, Welch DC, Zic JA, Copper WO, Stein SM, Hummell DS. Scurvy presenting as painful gait with bruising in a young boy. *Arch Pediatr Adolesc Med* 2000; **154**:732-5.
10. Larralde M, Santas Munoz A, Boggio P, DiGruccio V, Weis I, Schygiel A. Scurvy in a 10-month-old boy. *Int J Dermatol* 2007; **46**:194-8.

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