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Case Report

Scurvy "in the Land of Citrus Fruits"

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Abstract

Nutritional deficiencies including scurvy are still prevalent in developing countries though not as common as they once were. Vitamin C deficiency has varied presentations including musculoskeletal problems, skin manifestations and bleeding tendency. We report a case of a five years old developmentally normal child who presented with debilitating bone pains and inability to walk. Overall clinical and radiological features were suggestive of scurvy. Treatment with vitamin C resulted in significant improvement.

Key words: Ascorbic acid, Gum hyperplasia, Scurvy, Metabolic complications, Nutrition, Pocalcemia, Vitamin C.

Introduction

Vitamin C is a water soluble vitamin. Citrus fruits are rich source of vitamin C. It is not stored in the body. Foods containing vitamin C are commonly consumed and the vitamin is taken up by all the tissues. The classic disease was reported to occur in sailors due to diet deficient in vitamin C. Since the discovery of link between vitamin C deficiency and scurvy, it is rarely seen in developed countries. Case is not the same in developing countries where scurvy is still prevalent but overlooked. Occurrence of scurvy is unfortunate in our country which is amongst the best citrus producing countries. It is important to identify individuals at risk and recognize the clear and classic sign and symptoms associated with scurvy. Failure to diagnose this disease can potentially lead to expensive and unnecessary medical tests, as well as missing a very simple and easy to treat condition. This result in serious morbidity and even death due to life threatening bleeding or serious infections. Early diagnosis and timely management can not only prevent complications but also results in improved quality of life.

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Case Report

A 5 years old boy presented to the pediatric medicine department with complaints of progressively worsening pain in both legs for four months and inability to walk and sit for last one month. Child was in his usual state of health four months back, when he started complaining of pain in the legs and started losing interest in play. Pain was aggravated on walking and thus he preferred keep sitting. No joint swelling, stiffness or skin rash was noticed. He was taken to a local health facility where he was diagnosed as having growing bone pains and parents were counselled regarding benign nature of the problem. There was temporary relief with analgesics but general condition of the child did not improve much. Over the course of weeks his pain worsened to the extent that he was extremely irritable and confined to bed due to inability to move because of pain.

There was history of fracture femur one and a half year back following fall from the chair. He was treated at orthopedic department in another hospital. The fracture took longer than usual to heal. He was able to return to his usual activities following healing. He was born full term and vaccinated partially and was breast fed for one month only and then diluted cow's milk was fed for one year. Weaning started at about 7 months. His usual diet included roti, biscuits and tea. Fruits, milk, meat and eggs were consumed rarely. Both parents were laborers, had four children and the children were taken care of by the old grandmother in their absence. On examination he had wasted appearance. He was a sick looking, pale and irritable child without any respiratory distress. His weight was < 5th percentile and height 10^{th} centile for age. He had low grade fever of 100°F. His heart rate, blood pressure and respiratory rate were normal. The child was difficult to be examined due to extreme pain. He could not bear weight on legs. There was tenderness, and swelling in both lower limbs. There was no joint swelling, petechiae or bruises. There was generalized hypotonia. Reflexes were normal in upper limbs however they could not be assessed in lower limbs due to extreme pain and tenderness.

Further examination revealed a non-healing ulcer on the forehead which according to his mother was a small scratch initially that failed to heal and increased in size over time. Skin was dry and hyperpigmented without any evidence of bleeding. He had prominent costochondral junctions with sharp angulations (figure 1). Examination of oral cavity revealed purple colored spongy gum swellings with bleeding and hematoma formation (figure 2). Systemic examination was unremarkable. Patient was admitted and I/V antibiotics started. Labs results showed Hb of 5.6 gm/dl, normal leukocyte count, platelet count and coagulation profile. Peripheral smear showed microcytic hypochromic anemia. Serum ferritin level was low. Serum calcium, phosphate and vitamin D levels were within normal limits. Findings on X-rays lower limbs revealed the diagnosis. There was reduced bone density, cortical thinning which was more marked in the region of epiphysis (Ring sign), small fractures at the corner (pelkun spur) and dense calcification zone at physis (figure 3 and 4).

Clinical presentation and radiological findings of our patient strongly favor diagnosis of scurvy which needs serum vitamin C level for its confirmation. Vitamin C levels are not done in our setup so we made the diagnosis of scurvy on clinical presentation and radiological findings. Oral Vitamin C was started and child showed marked improvement within 2 weeks and had no walking difficulty or pain.

Figure 1: Prominent costochondral junction: Scorbutic rosary

Figure 2: Gingival hypertrophy

Figure 3: Radiological findings

Figure 4: Pencil thin cortex, Ring sign, Pelkun spur

Discussion

Vitamin C is a water soluble vitamin. Its common sources are oranges, strawberries, tomatoes, leafy green vegetables and germinating pulses.[1] Cooking destroys the vitamin. It is not found in meat. Inadequate intake of vitamin C results in scurvy. Vitamin C storage is depleted in 1 to 3 months.[1,2] It is a potent antioxidant and is also required for normal collagen synthesis. The recommended daily allowance is 50 to 60 mg per day during first year of life. The recommendation for the content of vitamin C in formula is 10 to 30 mg/100 kcal.[1,3] Complementary feeding with fruits and formula or breast milk can help to meet vitamin C requirement. Vitamin C is destroyed during the industrial processing of almond beverages.[4] Scurvy like illness was described in Egyptian medical scrolls and later on it was described as sailor's disease.[5] Initial symptoms of scurvy are vague and difficult to diagnose early. Malaise, fatigue, and body aches occur in early stages.[6] Musculoskeletal problems, skin manifestations, gum diseases and bleeding tendency occur later. Frequently, easy bleeding of spongy gums is encountered.[7] These symptoms if go unrecognized can result in severe debilitating course rendering the patient confined to bed, loosening of teeth and skin ulcers occur. Life threatening bleeding can occur due to fragile vessels. Cause of death is intracranial bleed or infections of open wounds.[7,8]

Musculoskeletal manifestations are common in scurvy during childhood. In 19th century clinicians concluded that the condition called "acute rickets" was actually scurvy.[9]

The constitutional symptoms in infantile scurvy include failure to thrive, anorexia and irritability. Debilitating leg pains result in pseudoparalysis. Joint pain and swelling may be the first presentation.[9,10] Radiographs show sheathing of the diaphyseal shafts, broadening of the calcification front at the anterior end of the ribs and ends of the metaphyses, and demineralization with metaphyseal fractures.[5,10] Subperiosteal hemorrhages are seen during healing phase and involve paraepiphyseal regions. These are palpable as painful swellings over the distal ends of long bones. Epiphyseal separation may also result.[11]

Lab diagnosis requires estimation of serum ascorbic acid level. The normal range is 5–16 mg/l. Classical clinical features occur when levels fall below 2.5 mg/l. Leukocyte ascorbic acid assays are a more accurate indicator of the body pool but are not done on a routine basis.[11,12] Treatment with vitamin C supplementation results in dramatic recovery. Anemia and other coexisting nutritional deficiencies are also treated. Scurvy line is lost with healing leaving behind a double line of ossification at the original site.[12] most of the reported cases had developmental or feeding issues but our case was a developmentally normal child. Disease occurred due to primary malnutrition.

Conclusion

Scurvy can have varied symptoms and signs. Due to multisystem involvement the differential diagnosis includes malignancy, connective tissue disorders or autoimmune diseases. Symptomatic infants are often misdiagnosed as victims of child abuse. High index of suspicion is needed to diagnose this easily treatable condition.

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