Case Report

SCURVY

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Abstract : Scurvy is a disease of ancient times. Now-a-days, with advances in the understanding of the disease, improvement in the nutrition and health standard, scurvy is rarely noticed. Very few cases of scurvy occurred in neglected and developmentally delayed children. We present a 9 years old child who was a known case of cerebral palsy and presented with painful swelling of the right knee, with bleeding from the gums. With vitamin C replacement therapy there was a dramatic improvement in his condition. This case serves as a reminder to the clinicians that even though rare in today's practice, ascorbic acid deficiency is still encountered and when recognized, is an easily treatable disease.

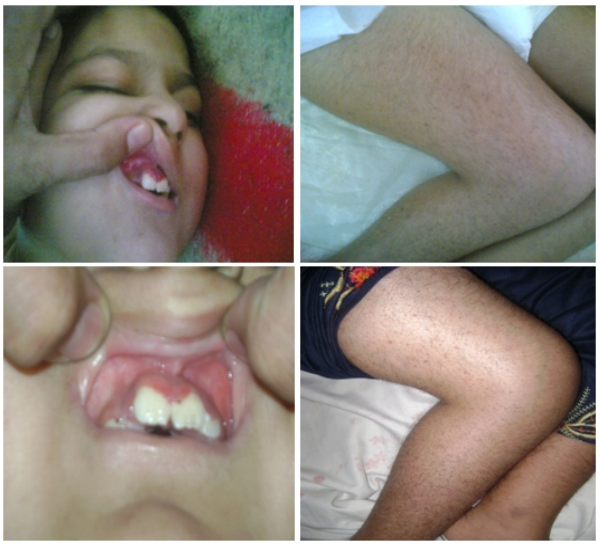
Introduction:

Scurvy is a nutritional disease that occurs due to low intake of vitamin C for prolonged period of time. The human body lacks the ability to synthesize and make vitamin C and therefore depends upon exogenous dietary source to meet vitamin C needs. Consumption of fruits and vegetables or diet fortified with vitamin C is essential to avoid ascorbic acid deficiency.

Vitamin C is functionally more relevant for collagen synthesis and its deficiency results in improved collagen formation. The typical pathological manifestation of vitamin C deficiency are noted in collagen containing tissues and organs like skin, cartilage, dentine, ostoid and capillary blood vessels. Defective collagen formation leads to defective dentine formation, hemorrahage into the gums, painful swelling of the long bones with pseudoparalysis and chostochondral beadings. The diagnosis of scurvy is based on the combinations of clinical and radiological findings. In questionable cases, the treatment with vitamin C can actually be diagnostic as well as therapeutic. Ascorbate (vitamin C), when given in the dose of 150 to 300 mg per day for one month, can lead to dramatic improvement in the clinical condition.

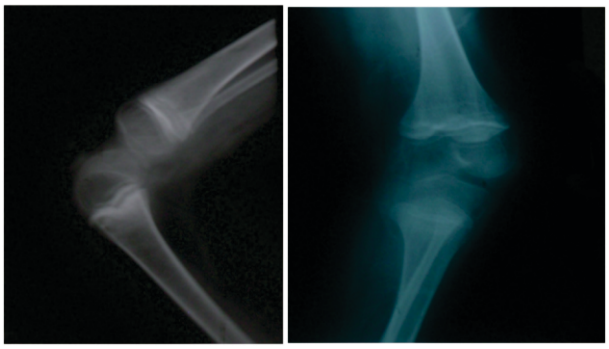
Case Report

A 9 years old Faizan, who is a known case of cerebral palsy, presented with complains of swelling of right knee joint and bleeding from the gums for the last 2-3 months. Physical examination showed, a thin emacia-ted child lying in a frog legged position. The legs were placed in a semi flexed position at the knee and hip joint. There was swelling over the knee and tibial area, more on the right side. Swelling was highly tender to touch with gross restriction of movements of the legs. It gradually involved the thigh and the hip joint. Further examination revealed multiple petichiae, which were associated with perifollicular hemorrhage and hyperkeratosis of the hair follicles. The gums were purplish in colour and there was swelling and oozing of blood from them. These findings were more marked to the upper incisors. There was depression of sternum as well as prominent chostrochondral junctions.



Birth history showed delayed cry at the time of birth. There past history of recurrent chest infections. Dietary history reveals intake of liquids and semi solid diet, with restriction of citrus fruits due to fear of chest infections.

Radiological examination of knee reveals typical findings of scurvy with thin cortex and ground glass appearance of medullary region. There was periosteal elevation and thickening at the area of provisional calcification, which appeared in the form of "White



Lines of Frankel".

Patient was started with oral vitamin C in the dose of 130mg/kg. After one month of treatment, when he was examined in the followed clinic, there was drastic improvement in his general condition as well as bone pain and swelling.

Discussion

Clinical manifestation of vitamin C deficiency "SCURVY" [1], is a disease of mainly dietary origin[2]. Vitamin C is a hydro-soluble sugar-like molecule found in citrus fruits, vegetables with green leaves, raw meat, and human and cow's milk. [1] It was isolated by the Hungarian scientist

Albert Szent-Gyorgyi in 1927. It was named "ascorbic acid "because of its recognized effectiveness against scurvy.

Scurvy has a prolific history in the annals of medicine. The first epidemic outbreaks of scurvy were seen amongst sea voyagers during the 14th and 16th centuries, because they lacked the adequate supplies of fresh vegetables, fruits, or animal foods. As it became rare at sea during the 19th century (after the provision of lemon juice onboard), an epidemic of land scurvy was witnessed, related to food deprivation occurring during wars and long expeditions.[3]

In the pediatric population, those at risk include children and teenagers with neurodevelopment disabilities or psychiatric illness, those with unusual dietary habits, food faddists, children of neglectful parents, those who avoid vitamin C–containing foods because of severe food allergies or gastrointestinal disease, patients with cancer, and those receiving long-term dialysis. Adolescents may be particularly at risk because of experimentation with restricted diets, their lack of concern regarding health, and lack of supervision.

Humans are unable to synthesize vitamin C and thus rely on dietary sources. Citrus fruits and juices are a major contributor to ascorbic acid in the US diet. Other good sources include collard greens, spinach, broccoli, tomatoes, potatoes, and strawberries. Red meat, chicken, fish, grains, eggs, and dairy products provide minimal vitamin C. However, many of these foods, particularly cereals, are fortified. Ordinary cooking decreases a food's vitamin C content by 20% to 40%.[4]

Bone involvement is typical for scurvy in children. The bony changes occur at the junction between the end of the diaphysis and growth cartilage. Osteoblasts fail to form osteoid (bone matrix), resulting in cessation of endochondral bone formation. Calcification of the growth cartilage at the end of the long bones continues, leading to the thickening of the growth plate. The typical invasion of the growth cartilage by the capillaries does not occur. Preexisting bone becomes brittle and undergoes resorption at a normal rate, resulting in microscopic fractures of the spicules between the shaft and calcified cartilage. With these fractures, the periosteum becomes loosened, resulting in the classic subperiosteal hemorrhage at the ends of the long bones. Intra-articular hemorrhage is rare because the periosteal attachment to the growth plate is very firm.

The principal manifestations of scurvy are primarily a result of abnormal collagen formation (vitamin C acts as a cofactor in the hydroxylation of proline and lysine residues of free procollagen chains[5], causing bleeding in the skin, mucous membranes, joints, muscles, or gastrointestinal tract and dystrophic hair deformities[6]. In addition, because of low levels of ascorbate, oxidative damage to coagulation factors occurs, and may contribute to bleeding.[7]

Fatigue is often the earliest sign of scurvy. The patient may complain of bruising on the lower extremities, swollen or bleeding gums, and coiled or fragmented hairs. Nonspecific symptoms include anorexia, shortness of breath, weight loss, myalgia, arthralgia, weakness, depression, and irritability. Prolonged disease can result in poor wound healing, edema, fever, and lethargy. Late findings include seizures, syncope, and sudden death.

The physical examination is notable for follicular nic for scurvy. Ecchymoses, petechiae, and purpura often

hyperkeratosis and perifollicular hemorrhages that are pathognomonic for scurvy. Ecchymoses, petechiae, and purpura often occur on the lower extremities but may be seen anywhere on the body. Gum abnormalities include swelling of the interdental papillae, sponginess, purplish discoloration, and hemorrhages. Epistaxis, hemarthrosis, muscle hemorrhage, and gastrointestinal bleeding may be present. Later findings are alopecia and softening or thinning of old scars. Ocular manifestations include conjunctival or intraocular hemorrhages. Examination of the fundus may reveal cotton-wool spots or flame-shaped hemorrhages[8]. The patient may have femoral neuropathy.

The diagnosis of scurvy is largely based on history and physical examination. An extensive workup is not necessary. At a minimum, a serum ascorbic acid level is usually obtained to help confirm the diagnosis. However, these levels do not correlate well with body reserves and tend to reflect recent dietary intake. The leukocyte ascorbate concentration is more closely related to tissue stores, but this test is technically difficult and not readily available. Urinary excretion of vitamin C is another way to detect body deficiency. A moderate to severe normocytic, normochromic anemia is common, but macrocytic anemia may occur in some cases as the result of concomitant folate deficiency. Other reasons for the anemia are gastrointestinal hemorrhage, altered metabolism of iron and folate, decreased red cell life span, and hemolysis[4]. Patients have a normal platelet count, aggregation studies, and bleeding time.

The characteristic radiologic changes occur at the growth cartilage-shaft junction of bones with rapid growth. The knee joint, wrist, and sternal ends of the ribs are typical sites of involvement. In the early phase of scurvy, the cortex becomes thin and the trabecular structure of the medulla atrophies and develops a ground-glass appearance[10] .The zone of provisional calcification becomes dense and widened, and this zone is referred to as the white line of Fränkel. The epiphysis also shows cortical thinning and the ground-glass appearance. As scurvy becomes advanced, a zone of rarefaction occurs at the metaphysis under the white line. The zone of rarefaction typically involves the lateral aspects of the white line, resulting in triangular defects called the corner sign of Park. This area has multiple microscopic fractures and may collapse with impaction of the calcified cartilage onto the shaft. The lateral aspect of the calcified cartilage can project as a spur. Subperiosteal hemorrhages are not visualized in the active phase. With healing, they become calcified and are readily observed.

In questionable cases of scurvy, the treatment can actually be diagnostic as well as therapeutic. Moreover, because patients have clinical improvement in as little as 2 to 3 days, a therapeutic trial is practical. Infants and children are given 150 to 300 mg ascorbate per day orally for 1 month. Adults have been treated with 1.0 g per day for 2 weeks. The patient's former diet must be corrected, and other nutritional deficiencies, particularly folate,must be explored. Most of the clinical manifestations of scurvy resolve within 2 weeks. Permanent bony changes may occur in infants and children with

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