

Scurvy Presenting As Pseudoparalysis Without Other Classical Clinical Features : A Case Report

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Abstract

A 28 months old boy presented in the casualty for evaluation of lower limb weakness and difficulty in walking since last 2 weeks and being referred as acute flaccid palsy suspected as Guillen Barre Syndrome for further management. The clinical presentation and imaging findings, together with the dramatic response to ascorbic acid intake, allowed us to confirm the diagnosis of infantile scurvy.

INTRODUCTION

Scurvy is a common nutritional disorder in developing countries due to high prevalence of malnutrition. Vitamin C performs various important functions in the body. It is required for synthesis of supporting tissues of mesenchymal origin like osteoid in bone, chondroid in cartilage, dentin in teeth and collagen in capillary walls. It enhances iron absorption from the gastrointestinal tract, acts as an antioxidant, required in the coagulation pathway, also in tyrosine metabolism and synthesis of corticosteroids¹. Amla, guava and citrus fruits are rich dietary sources of vitamin C¹. Human breast milk has five times more vitamin C than cow's milk, thus breast fed infants are less prone to develop scurvy. Delay in starting weaning foods, improper dietary habits and recurrent diarrhea predispose to scurvy

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CASE REPORT

A 28 months old boy presented in the casualty for evaluation of lower limb weakness and difficulty in walking since last 2 weeks and being referred as acute flaccid palsy suspected as Guillen Barre Syndrome for further management. On examination, he was irritable and thinly built. Height and weight were in the 85th percentile for his age and sex. There was no muscle weakness noticed. There was marked tenderness in the lower limbs and difficulty in walking with a wide-based, waddling gait, requiring assistance to ambulate. He had normal abdominal examination results, without organomegaly. He had no apparent joints swelling or gum hypertrophy, but there was pain with motion of his knees and hips. The remainder of his neurological

examination was normal. The past history and development history was normal. His dietary history indicated that he was mainly fed with diluted feeds during infancy and very less green vegetables and citrus fruits being included in the diet.

Imaging of lower limbs showed classical features of scurvy (fig.1) and hence diagnosis –pseudo paralysis of scurvy made and accordingly child was treated with 200 mg of daily ascorbic acid for one month and dietary modifications were also advised. Seventy-two hours after starting oral vitamin C supplementation, there was significant improvement in the patient's general health. He markedly improved within 2-3 weeks with follow up x ray showing signs of healing. He was able to walk in 2 weeks of treatment. Imaging of lower limbs also showed signs of healing (fig.2). The clinical presentation and imaging findings, together with the dramatic response to ascorbic acid intake, allowed us to confirm the diagnosis of infantile scurvy.

DISCUSSION

Scurvy is the disease caused by deficiency of vitamin C in diet. The typical pathologic manifestations of vitamin C deficiency are noted in collagen-containing tissues and organs such as skin, cartilage, dentine, osteoid and capillary blood vessels. Pathologic changes are a function of the rate of growth of the affected tissues; hence the bone changes are often observed only in infants during periods of rapid bone growth. Defective collagen synthesis leads to defective dentine formation, hemorrhaging into the gums, and loss of teeth. Hemorrhaging is a hallmark feature of scurvy and can occur in any organ. Hair follicles are one of the common

sites of cutaneous bleeding ¹² .

Bone involvement is typical for infantile scurvy. 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 ¹²³ .

Scurvy usually presents between 6 and 24 months of age ²³ . Initially there may be nonspecific symptoms such as Loss of appetite, poor weight gain, and diarrhea. The Specific symptoms occur later which include irritability, pain and tenderness of the legs, swelling over the long bones and hemorrhage. The pain results in pseudo paralysis ²³ . Bori A et al recently described a case report of scurvy with musculoskeletal pain and difficulty in walking ⁴ . But, in our case, the child was though presented as lower limb weakness and difficulty in walking and there was no gingival hypertrophy or other signs of increased capillary fragility. There was marked tenderness and child was even not allowing to touch the limbs for examination. This is very characteristic of scurvy and the other close differential for this is leukemia which should be ruled out ⁴⁵ . Kumar et al described a case report of scurvy being diagnosed and treated as juvenile idiopathic arthritis ⁶ . Even in our case, the child was referred as suspected Guillen Barre Syndrome. Thus ,one has to be aware of presentation of scurvy as pseudoparalysis due to pain in the lower limbs to avoid misdiagnosis

IMAGING FINDINGS

Radiologic findings in infantile scurvy are diagnostic ³⁴⁵ . 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. 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 ³⁴⁵ . These all findings were present in our case imaging of lower limbs (figure 1). Subperiosteal hemorrhages are not visualized in the active phase. With healing, they become calcified and the affected bone assumes a dumb-bell or club shape, which readily observed ¹²³⁴⁵ as in the X-ray of the lower limb of the child after 4 weeks of treatment (figure 2).

Figure 1

Figure 1: features of scurvy on X-ray of long bones



Figure 2

Figure 2



After treatment of 1 month of ascorbic acid healing present in the long bones in the same patient.

DIAGNOSING SCURVY

Diagnosis of vitamin C deficiency is usually based on the characteristic clinical picture, the radiographic appearance of the long bones, and a history of poor vitamin C intake³⁷⁸. Laboratory tests are usually not helpful to ascertain a diagnosis of scurvy³⁷. Serum ascorbic acid levels can be used for support of diagnosis but it is not confirmative. A fasting serum ascorbic acid level greater than 0.6 mg/dL aids in precluding vitamin C deficiency, but an even lower concentrations does not necessarily indicate deficiency. White blood cell/platelet layer (buffy layer) of centrifused oxalated blood ascorbic acid concentration is a more accurate measure of a vitamin C nutritional state. A level of zero indicates latent scurvy, even in the absence of clinical signs of deficiency. Saturation of the tissues with vitamin C can be estimated from the urinary excretion of the vitamin after a test dose of ascorbic acid. In normal children more than 80% of test dose appears in urine within 3-5 hours. Generalised nonspecific aminoaciduria is common in scurvy³⁴.

Other differential to be considered are copper deficiency, arthritis, syphilis, leukemia, Henoch-Schonlein purpura⁴⁵.

PREVENTION

A diet adequate in vitamin C can prevent the development of scurvy. The dietary requirements of vitamin C sufficient to prevent deficiency vary with the age of the individual. The following are the Food and Nutrition Board of the National Academy of Sciences, National Research Council's minimum recommended daily dietary allowances of vitamin C⁸⁹.

Infants - 30-40 mg
Children and adults - 45-60 mg
Pregnant women - 70 mg
Lactating mothers - 90-95 mg

TREATMENT

Scurvy responds rapidly to oral or parenteral ascorbic acid. Daily intake of 3-4 oz of orange or tomato juice produces healing in children with scurvy³.

Dose is 100-200 mg per day for 2 weeks. Large doses may

precipitate hemolysis in individuals with glucose-6-phosphate dehydrogenase deficiency. Interfere with the absorption and metabolism of vitamin B-12, can cause renal stones and diarrhea. Limited data exist on vitamin C toxicity in children⁶. The following values for tolerable upper intake levels were extrapolated from data for adults based on body weight differences: for children 1-3 yr, 400 mg; 4-8 yr, 650 mg; 9-13 yr, 1200 mg, and 14-18 yr, 1800 mg³⁸⁹.

SUMMARY AND MESSAGE

Thus, this case highlights that scurvy should be kept as differential in any child presenting with lower limb weakness with marked tenderness which may mimic acute flaccid paralysis. It can be easily confirmed with X ray of long bones with characteristic features as described and it is easily treatable with oral vitamin C. It is important to remember that scurvy can present atypically i.e. with painful limbs, pseudo-paralysis and without bleeding gums or scorbutic rosary. Scurvy should be kept as a differential diagnosis in a young child presenting with bleeding manifestations or only with difficulty in walking without other features. Taking a detailed dietary history is of utmost importance in pediatric practice for the diagnosis.

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