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CASE REPORT

Scurvy – Still an added co-morbidity in Children with Cerebral Palsy

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Abstract:

Scurvy is caused by prolonged severe dietary deficiency of ascorbic acid. Although it is uncommon today, it still exists in high risk groups, particularly in those with abnormal dietary habits, mental illness or physical disability. Health care providers must inquire about nutritional habits and keep in mind the risk of nutritional deficiency while treating a child with neurodevelopmental disabilities. Otherwise there is every possibility to miss the diagnosis of scurvy. Here we have described a case of scurvy in a child with cerebral palsy who presented with the clinical manifestations along with all the classical radiological features of scurvy.

Keywords: Scurvy, ascorbic acid, cerebral palsy

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Introduction

Deficiency of Vitamin C (ascorbic acid) results in the clinical presentation of Scurvy, the oldest nutritional deficiency to be recognized. A disease that was once rampant is now rarely seen, more so in the pediatric age group [1]. Its clinical features are: asthenia, weight loss, appetite decrease, irritability, gingival or mucous lesions, petechial hemorrhages, follicular hyperkeratosis, musculoskeletal pain due to multiple fractures and subperiosteal bleedings, pseudoparalysis (frog-like position of legs) and refuse to walk [2]. People with abnormal dietary habits, mental illness or physical disability are prone to develop this disease [3]. Given the present-day rarity of this disease, the consideration of scurvy as a diagnosis is often overlooked by physicians, leading to extensive

laboratory and radiographic testing and unnecessary delays in diagnosis and treatment [4].

Here we are reporting a case of scurvy with other nutritional deficiencies in a child with cerebral palsy which was diagnosed and treated promptly leading to rapid recovery and early discharge from hospital.

Case report

A 2 ½ yrs old girl from lower socio-economic status was admitted to our department of Pediatrics with history of intermittent low grade fever for last 2 months, swelling of both lower limbs for 1 month & occasional gum bleeding for last 1 month. The child was normal up to 6

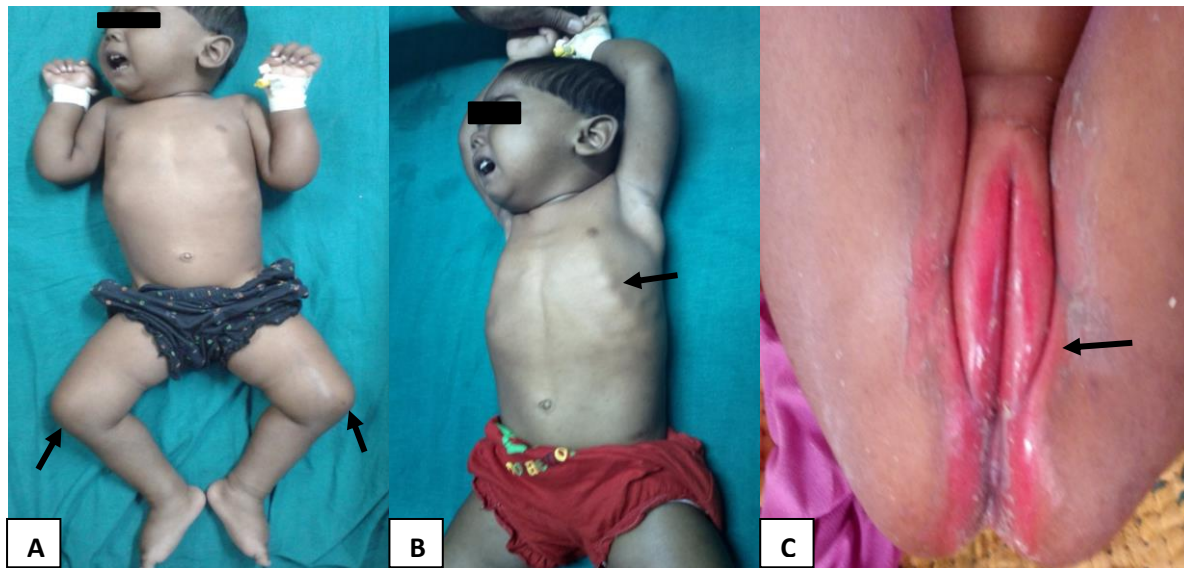


Figure 1. Child with bilateral knee swellings, scorbutic rosary and perianal dermatitis

months of age thereafter she developed high grade fever with chill & rigor and repeated episodes of generalized tonic-clonic convulsions. The child was not admitted but received treatment from local doctor. After 15 days of treatment with oral anticonvulsants and intravenous antibiotics, fever and convulsions were controlled, but patient became bed ridden. At that time complete evaluation for fever and the illness was not done. In spite of treatment with anticonvulsants, the patient developed intermittent episodes of convulsions. Since then the child was on oral anticonvulsant, but the parents stopped all medications for last one year without any medical consultation.

The child had an uneventful antenatal, natal and postnatal period. She was exclusively breastfed until 6 months of age but was on a predominant milk based diet for last one year, with minimal intake of fruits and vegetables. There was no history of trauma.

Now on examination, the child had moderate pallor, excessive irritability with toxic look, high grade fever (39°C) and bilateral minimal non-pitting edema. There was no hepatosplenomegaly or lymphadenopathy. On musculoskeletal system examination both the

knee joints (left > right) were swollen & tender with the skin on the joints appearing shiny, red and warm (Figure 1A). There was severe wasting of muscles with tender nodular swelling at costochondral junctions present on both sides symmetrically. Both the lower limbs were semi-flexed at knee & hip joints with external rotation (Figure 1B). Any attempted movement of the affected joints was painful and restricted. There was bluish spongy gum with active bleeding, gum hyperplasia and perianal dermatitis (Figure 1C) and other features of spastic quadriplegia. She had microcephaly (head circumference: 42cm), body weight: 9 kg, length: 77cm, mid arm circumference: 12.5 cm. All the growth parameters fell below the 3rd percentile lines of WHO growth charts.

Blood reports showed Hb 6.6 g, leukocyte count 11200/mm³, platelet count 189000/mm³, PCV 26.3%, MCV 86 fl, MCH 28.3 pg, MCHC 30.1 g, CRP 162 ng/L, hypochromic microcytic RBCs. Liver function test, renal function test, sugar & electrolytes were normal (calcium: 9.2 mg/dl; alkaline phosphatase: 102 IU/dl; phosphate: 3.7mg/dl).

We made a provisional diagnosis of severe sepsis with severe malnutrition with ascorbic

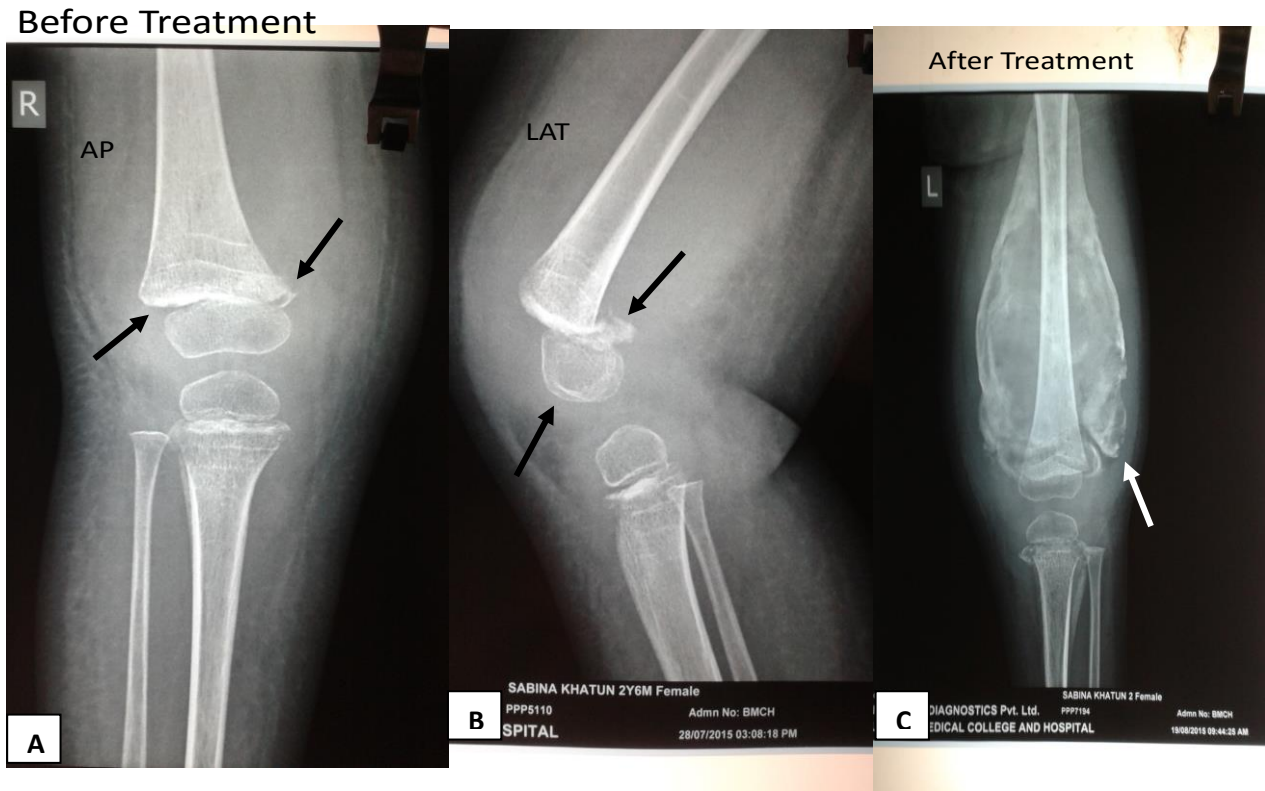


Figure 2. X-Ray AP & Lateral of knee joint showing Pelkan spur & white line of Frankel (arrows in A), Trumerfeld zone & epiphyseal slip (B), dumbbell shaped tumor (C).

acid deficiency in a child with spastic quadriplegic variety of cerebral palsy (CP) and started treatment with intravenous antibiotics, nutritional supplementation & other supportive care for anemia & CP.

X-ray both knee joints showed Cortical thinning with white line of Frankel and a characteristic zone of rarefaction under the white line at the metaphysis (Trumerfeld zone). Pelkan spur and diffuse soft tissue swelling were present (Figure 2A and 2B). Chest X-ray was normal. Ultrasonography of both lower limbs showed soft tissue edema with fluid collection in the subcutaneous as well as muscular layers. MRI brain showed cerebral atrophy with hyper dense basal ganglia and thalamus. Blood culture was sterile.

We also sent serum vitamin C and 25 (OH) cholecalciferol levels which came 3 mmol/L (normal 11-104 mmol/L) and 10 mcg/ml (normal 20-100 ng/ml) respectively.

The diagnosis of scurvy and vitamin D deficiencies were made, and the child was treated with 200 mg of vitamin C daily. Vitamin D 6 lakh IU was also administered. Her mother was educated about dietary modification. Perianal dermatitis was treated with antifungal & mid-potent steroid without any improvement which improved only after starting zinc supplementation (2mg/kg/day) after sending serum zinc level which came 45 mcg/dl (normal 65-130 μ g/dl). Two weeks after vitamin C administration, the child's general condition and joint swelling improved. Repeat X-ray of the knee joint showed features suggestive of healing but a huge dumble shaped tumor suggestive of calcified sub-periosteal hemorrhage was noted in left knee x-ray (Figure 2C).

Discussion

In the 21st century, the most devastating nutritional problem for many is not lack of nutrients, rather excessive calorie intake leading

to obesity and other metabolic syndromes. However, despite modernization among almost all societies, nutritional deficiencies including scurvy still occur in certain populations (those with neurodevelopmental disabilities, psychiatric illness or having unusual dietary habits) [5].

Scurvy though is less common in the pediatric population, but case reports still appear [6,7]. A review of the literature by Noble et al. reveals twenty three case reports of scurvy in children with behaviorally restricted diets including children with autism, mental retardation and cerebral palsy [8].

Various factors contribute to nutritional deficiencies in non ambulant children with severe spastic cerebral palsy like poor intake, oral motor dysfunction, feeding problems, and use of antiepileptic drugs [9].

Scurvy is common in them as they subsist on predominant milk based diets (due to pseudobulbar palsy and difficulty swallowing solids) and boiled cow's milk is a very poor source of vitamin C [1]. The diagnosis of scurvy is based on history of poor dietary intake of vitamin C, classic clinical features and radiological findings and prompt response to treatment with vitamin C [10, 11].

Scurvy can present with different manifestations, which mandates careful assessment of the diet for each and every child with cerebral palsy. Subperiosteal hematomas in scurvy may be palpable as painful swellings over the distal end of the femur and tibia. Our patient had large, uncomplicated intramuscular hematoma that healed by producing a dumbbell shaped tumor.

Laboratory test abnormalities in scurvy are nonspecific; anemia is frequent manifestation. Our patient had moderate anemia that required blood transfusions. The causes for anemia in scurvy could be multifactorial, resulting from blood loss, concomitant vitamin deficiencies, and decreased iron absorption [12]. Vitamin or mineral deficiencies other than vitamin C deficiency may accompany scurvy; therefore we will have to check for concomitant micronutrient deficiencies, testing levels of zinc, iron, folate,

and vitamin B12. In our case, serum 25 (OH) cholecalciferol and serum zinc were low. A low level of vitamin D occurs due to lack of sunlight exposure, dietary deficiencies and chronic anticonvulsant therapy [1].

Weinstein et al. [10] recommend oral doses of 100 to 300 mg of vitamin C daily until body stores are replenished per serum levels. Once a regimen of vitamin C is begun, improvement of symptoms usually begins in 24 hours, with pain diminishing in two to four days, and gingival lesions recovering in two to three weeks [13]. With vitamin C supplementation, metaphyseal abnormalities of scurvy will completely resolve [14]. The large shells of periosteal bone are common radiographic findings particularly during the healing phase of disease [15] as seen in our case.

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