SCURVY

CASE REPORT PROF-1946

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ABSTRACT... Scurvy is a condition caused by a lack of vitamin C (ascorbic acid) in the diet. It is extremely rare in industrialized socities but still exists in higher risk groups including economically disadvantaged populations with poor nutrition. We are reporting a case of 4 year old Asad who presented to our department with this condition.

Key words: Scurvy, perifollicular hemorrhage, scorbutic rosary.

INTRODUCTION

Scurvy is a state of dietary deficiency of vitamin C (ascorbic acid). The human body lacks the ability to synthesize and make vitamin C and therefore depends on exogenous dietary sources to meet its needs. Vitamin C is required as a redox agent, reducing metal ions in many enzymes and removing free radicals. Clinical manifestations of its deficiency seen in infants as well as in older children and adolescents include a "rosary" at the costochondral junctions, anemia, purpura, petechiae, ecchymoses, perifollicular hemorrhages, hyperkeratosis of hair follicles, swollen joints, poor wound and fracture healing, arthralgia, muscle weakness and generalized tenderness particularly in the legs.

CASE REPORT

Four year old Asad Ali, resident of Qasoor, presented to Department of Pediatrics, Lahore General hospital, with 2 months history of inability to walk along with progressive pallor, irritability, low grade fever and gum bleeding. He also complainted of generalized bony pains, tenderness and swelling more pronounced in lower limbs. There was no history of blood transfusion, bleeding from any other site, trauma, urinary and bowel complaint. He is the 5th issue of a consanguineous marriage, born vaginally with normal birth events and there is no family history of similar illness. He is developmentally normal and vaccinated according to EPI schedule. He belongs to lower socio-economical class and his father is a daily wage laborer. He was breast fed uptil 2 year of age when inadequate weaning was started.

Examination revealed pale, anxious child with swelling

and tenderness over all the bones more pronounced in the lower limbs. He had bruises and petechae more over the lower limbs. There was no jaundice or lymphadenopathy. His pulse rate was 105 beats per min; respiratory rate was 32 breath per min; temperature was 101° F; blood pressure 90/70mm of Hg and his anthropometry revealed height and weight below 5th percentile.

Complete blood counts showed microcytic hypochromic anemia with hemoglobin of 6.2 g/dl, WBC count of 5500/mm³ with 65% lymphocytes, and platelets count of 211000/mm³. His renal, liver function tests and urine complete were within normal limits. His electrocardiography, echocardiography, chest X-ray, bleeding time, prothrombin time and activated partial thromboplastin time were also within normal limits. X-ray of his knee joint revealed a thick sclerotic metaphyseal line above a widened physis, small beak-like excrescences at the metaphysis of both femora and pencil thin cortex, all of which were suggestive of scury. His bone marrow aspiration revealed megaloblastic erytheropoisis with no evidence of infiltration.

DISCUSSION

Scurvy is a disease resulting from a deficiency of vitamin C (ascorbic acid), which is needed for a variety of biosynthetic pathways. In the synthesis of collagen, ascorbic acid is required as a cofactor. Defective collagen fibrillogenesis impairs wound healing and bone formation. Defective connective tissue also leads to fragile capillaries, resulting in abnormal bleeding. Scurvy can be prevented by a diet that includes certain citrus fruits such as oranges or lemons.

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Other sources rich in vitamin C are fruits such as guava, papaya, tomatoes, strawberries, bell peppers, blackcurrants, and kiwifruit¹.

Early clinical manifestations of the disease include pallor, irritability, and poor weight gain as was seen in our case². In advanced disease, the major clinical manifestation is extreme pain and tenderness of the arms and the legs³. The child is miserable and tends to remain in a characteristic immobilized posture from subperiosteal pain, with semiflexion of the hips and the knees (pseudoparalysis). Four year old Asad had all of these features.

The body was both wasted and edematous, and petechiae and ecchymoses⁴ are commonly present. Hyperkeratosis, corkscrew hair, and sicca syndrome are typically observed in adult scurvy but rarely occur in infantile scurvy. The case of an infant with diffuse, nonscarring alopecia of the scalp and radiologic features of scurvy have also been reported⁵. although sicca syndrome was not found in Asad but he had alopecia, radiological features, petichae, bruises and gum bleeding which were suggestive of scurvy.

Normochromic, normocytic anemia develops in 75% of patients, resulting from blood loss into tissue, coexistent dietary deficiencies (folate deficiency), altered absorption and metabolism of iron and folate. The same peripheral picture was seen in our patient and he was transfused twice to raise his low hemoglobin.

Scorbutic rosary at the costochondral junctions of the ribs and may occur in children. Fractures, dislocations, and tenderness of bones are also common. Bleeding into the joints causes exquisitely painful hemarthroses. Subperiosteal hemorrhage⁶ may be palpable, especially along the distal portions of the femurs and the proximal parts of the tibias of infants. These findings were not present in our patient. Gum hemorrhage⁷ occurs only if teeth have erupted and usually involve the tissue around the upper incisors. The gums have a bluish-purple hue and feel spongy. Gum swelling, friability, bleeding, and infection with loose teeth also occur, as do mucosal petechiae. All of these finding were present in our patient.

Cardiac complications include cardiac enlargement, electrocardiographic (ECG) changes (reversible STsegment and T-wave changes), hemopericardium, and sudden death. Bleeding into the myocardium and pericardial space has been reported. High-output heart failure due to anemia can be observed. No such cardiac anomaly was observed in our patient.

At the time of admission Asad was considered as a case of juvenile idiopathic arthritis (JIA) due to inflamed tender swollen joints fever and pallor. We also had a suspicion of acute leukemia due to marked pallor, petechae, bruise, fever and bony tenderness for which bone marrow aspiration was done. During investigation for JIA the Xrays showed the findings suggestive of scurvy in lower end of femurs. Vitamin C (ascorbic acid) level estimation could not be done because of nonavailability of this test. However on the basis of history, examination, radiological investigations and exclusion of other diagnosis, the diagnosis of scurvy was made. He was transfused packed cells twice and was given vitamin C with folic acid, multivitamin and dietary advice. Asad responded guite dramatically to our treatment and his symptoms of irritability, tenderness and easy fatigability improved considerably during his stay at hospital. He is still on our follow up and doing well.

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Malik M. Khalid, SCURVY; RADIOLOGICAL DIAGNOSIS (Case Report) Prof Med Jour 16(3) 466-468 Jul, Aug, Sep, 2009.

