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SCURVY*
A Case Report

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An 18-month-old girl presented with irritability, epistaxis, spongy appearance of the gums perifollicular papules with follicular hyperkeratosis, ecchymosis, painful swollen knees and scorbutic rosary. Her diet consisted mainly of wheat flour. X-ray of the knees showed findings compatible with scurvy. Ascorbic acid level was below 0.003 g/L. Ascorbic acid therapy resulted in a dramatic clinical improvement. Scurvy is an uncommon disease in our society today. It is important to recognize the signs and symptoms of scurvy because it is easily treated with vitamin C replacement. *Key words: scurvy-vitamin C.*

Scurvy is now an uncommon disease in the world, but this condition is still a problem in developing countries, where general malnutrition prevails[1]. Most cases occur in elderly persons who live alone, the mentally ill, alcoholics and those who consume an inadequate diet[2]. It can also occur in children with peculiar dietary habits resulting from parental misguidance or neglect[3,4].

We report a case of scurvy in an 18-month-old female with an inadequate diet because of her mentally ill mother.

Case Report

An 18-month-old girl was admitted to our hospital with a four-week history of irritability, weakness, fever, repeated epistaxis, bleeding from the gums and swelling of the knees.

She was born to nonconsanguinous parents after an uncomplicated delivery. Her mother was mentally retarded. The parent was questioned about the child's nutritional history. There was no breast-feeding. She had been fed on too-diluted formula for the first three months and on a mixture of water and wheat flour later. No vitamin supplements had been given for the previous months.

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On physical examination the patient weighed 10 kg (25^{th}-50^{th} percentile) and was 80 cm in length (25^{th}-50^{th} percentile). Her head circumference was 44 cm (below third percentile). Rectal temperature was 39 °C, blood pressure was 90/50 mm Hg and her heart rate was 156/min. Her behavior was aggressive. Ecchymosis and perifollicular papules with follicular hyperkeratosis were present on her thighs. Her lips were dry and fissured. The gums were friable and eroded (Fig. 1). There was edematous swelling along the shaft of the legs. The knees were swollen and painful (Fig. 2). Rosary was observed at the costochondral junctions. Deep tendon reflexes were normal.

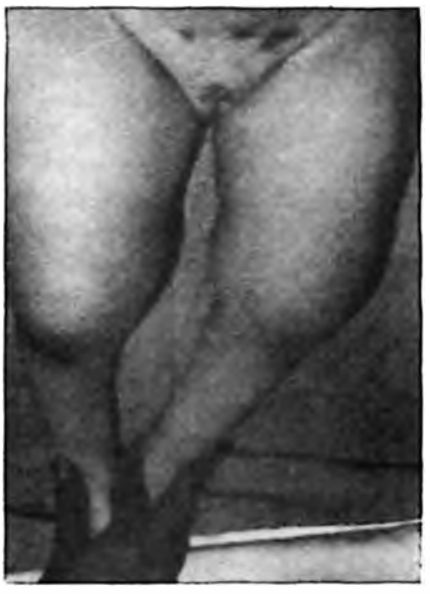


Fig. 1: The patient's gingivae show hemorrhage, edema and erosions.

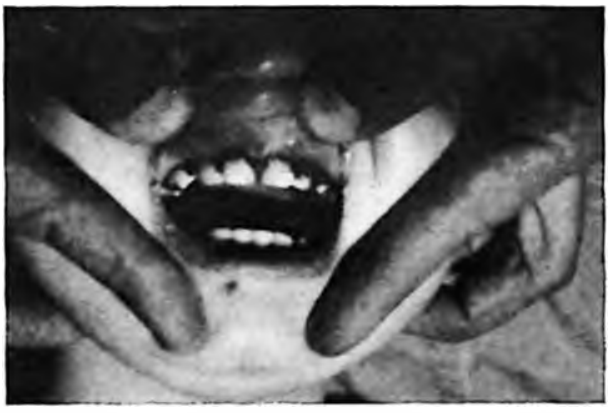
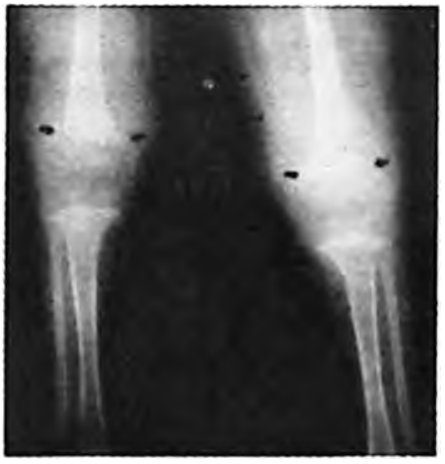


Fig. 2: Perifollicular papules with follicular hyperkeratosis, ecchymosis, and edematous swelling of the legs and knees are seen.

The patient's hemoglobin level was 70 g/L and her hematocrit was 28 percent with normocytic and normochronic indices. White cell and platelet counts were normal. Electrolytes, renal and liver function tests, prothrombin time, activated

partial thromboplastin time and bleeding time were found to be normal. Bone marrow examination revealed erythroid hyperactivity. The serum ascorbic acid level was less than 0.003 g/L (normal 0.005-0.018 g/L). Roentgenograms of both knees including femur showed features of acute scurvy (Fig. 3). The clinical and laboratory findings led to the diagnosis of scurvy. Our patient was treated with parenteral ascorbic acid, 300 mg daily. After 10 days, the patient showed dramatic clinical improvement and her serum ascorbic acid level rose to 0.01. Vitamin C was continued with a dose of 50 mg/day for three months. Follow-up x-ray of both knees showed signs of healing with calcification of subperiosteal hemorrhage (Fig. 4).



(A)

(B)

Fig. 3: X-ray of both knees including femur taken on admission shows peripheral metaphyseal defects (Pelkan's spur), osteoporosis, rings around the epiphyses of the femur and tibia (Wimberger ring), and subperiosteal hematoma (A). One week later, linear displacement of the epiphysis against the shaft is seen (B).

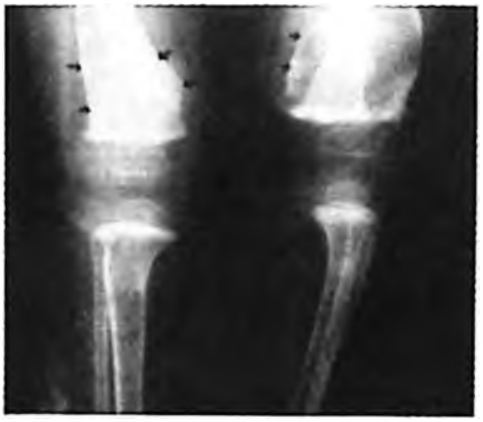


Fig. 4: X-ray of both knees after one month shows signs of healing with calcification of subperiosteal hemorrhage.

Discussion

Ascorbic acid (vitamin C) is an essential water soluble vitamin for human beings because of the absence of the microsomal enzyme L-Gulono-γ-lactone oxidase[5].

Vitamin C plays an important role in the formation of collagen; the synthesis of epinephrine, carnitine and steroids; folic acid metabolism; leukocyte function and iron absorption.

The infant is born with adequate stores of vitamin C if the mother's intake has been adequate. Breast-milk is an adequate source of vitamin C[1]. Infants fed with formula must also receive vitamin C supplements. Without ascorbic acid scurvy can occur. Today scurvy is a very rare disease. Nevertheless, cases of vitamin C deficiency are still being reported both in developed and developing countries[7,8]. Scurvy is also very rare in Turkey. Our patient's unusual eating habits due to her mother's mental retardation caused scurvy. Her mother did not know anything about the feeding of a child. Cases of scurvy have been previously published in patients with peculiar dietary habits as was observed in our patient.

The clinical findings of scurvy are generally first evidenced by petechial hemorrhages. These hemorrhages are usually perifollicular and occur in hyperkeratotic areas. Next to occur are ecchymosis and purpura, usually at sites of pressure. As the level of ascorbic acid continues to decrease, coiled and fractured hairs with hyperkeratosis develop. Gingivitis and bleeding from the gums may occur; these findings are important manifestations of scurvy. The manifestations of scurvy can progress to pseudoparalysis due to acutely painful subperiosteal hemorrhage in the long bones, lower extremity edema, muscle tenderness, conjunctival and intraocular hemorrhages, arthralgias, hemorthroses, gastrointestinal hemorrhage, Sjögen-like syndrome, poor wound healing and femoral neuropathy. Rarely, convulsions, hypotension and death can result.

Severe to moderate anemia secondary to hemorrhage may occur. Concurrent with these signs, the patient usually complains of general weakness, fatigue and weight loss[1]. The case illustrated here was presenting with aggressive behavior, depressed affect, irritability, perifollicular papules with follicular hyperkeratosis, ecchymosis, gingival erosions, painful swelling of knees and legs and rosary at the costochondral junction. There was also dietary history. Thus, the presumptive diagnosis of scurvy was made. The dignosis was confirmed by measuring the plasma ascorbic acid level, roentgenographic findings and clinical response to vitamin C.

The recommended daily allowance for vitamin C is 35 mg for infants, 45-50 mg for children and 60 mg for adults[1, 9-11]. Daily intake of 60 mg of ascorbic acid results in a 1500 mg pool. Clinical scurvy develops when the body's pool of vitamin C is depleted below 300 mg[10]. Replacement of the body store of vitamin C is the treatment for scurvy. Scorbutic children should be treated with 150-300 mg/day of vitamin C either intravenously or orally for a month, and 30-60 mg/day thereafter until complete recovery occurs[1, 10, 11]. Our patient was treated with 300 mg/day of vitamin C parenterally for 10 days, thereafter 50 mg daily for three months. A well-balanced diet containing fresh fruits and vegetables was recommended.

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