

Neonatal Scurvy- A Rare Case Report

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Abstract

Scurvy, a disease of vitamin C deficiency, is now considered as a rare disease in paediatric population. The human body is dependent on the external supply of vitamin C. The newborn infant receives this from breast milk. Therefore, infantile scurvy is generally observed only in infants over the age of three months. The occurrence of scurvy before that age is extremely rare. We described a case of neonatal scurvy in a 28 days old boy who was on parenteral nutrition for initial 10 days of age due to neonatal sepsis with perinatal asphyxia.

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Introduction

Vitamin C is an essential nutrient for humans and it must be acquired through an adequate diet to prevent hypovitaminosis C, deficiency and its consequences- including the potentially fatal deficiency disease scurvy.^{1,2} Vitamin C deficiency is defined as a serum concentration of less than 11.4 $\mu\text{mol/L}$, and prevalence varies across the world, with rates as low as 7.1% in the United States and as high as 73.9% in north India.³ Being rare as compared to other nutritional deficiencies,⁴ it is seldom suspected and this, along with its extensive variable presentation, frequently leads to delayed recognition of this disorder.^{2,5} Vitamin C (l-ascorbic acid or ascorbate) is intimately concerned with the maintenance of intercellular connective tissues, osteoid, dentine and collagen.⁶⁻⁸ It is also involved in neurotransmitter metabolism, cholesterol metabolism, biosynthesis of carnitine and nonheme iron absorption.⁹ Human beings lack the enzymatic process for conversion of glucose to ascorbic acid via gulonolactone oxidase unlike other animals (e.g. rats), therefore vitamin C supplementation in the form of fresh fruits, vegetables, or dietary supplements is essential for humans.^{10,11}

The early manifestations are irritability, loss of appetite, musculoskeletal pain and tenderness in the legs (pseudoparalysis). The child is frequently irritable and

does not like handling which was evident in our case. A “scurvitic rosary” at the costochondral junctions and sternum depression are other characteristic bony features.⁴ Anemia is another hallmark of scurvy, iron deficiency anemia is common and may be secondary to a combination of bleeding and decreased absorption.⁶ Haemorrhagic manifestations of scurvy include petechiae, purpura and ecchymoses at pressure points; epistaxis; gum bleeding and the characteristic perifollicular hemorrhage.⁶ The diagnosis of vitamin C deficiency is usually based on the characteristic clinical picture, the radiological appearance of long bones and the history of poor vitamin C intake.⁴ The radiographic findings of infantile scurvy are multiple: ground glass appearance of the shaft of the long bones, thin and dense cortex giving the appearance of pencil outlining of diaphysis, white line of Frankel at the end of the metaphysis with occasional lateral prolongation of the white line (Pelkan spur), ringing of the epiphysis and the more specific radiological feature is a zone of rarefaction proximal to white line at the metaphysis (Trümmerfeld zone). Subperiosteal haemorrhages are not visible using plain radiographs during the active phase of scurvy. However, during healing, the elevated periosteum becomes calcified and radio opaque.⁶ Radioisotope bone scan can also be useful due to the specific uptake pattern.^{12,13} The best evidence of the presence of scurvy is the resolution of the manifestations of the disease after ascorbic acid treatment.¹⁴⁻¹⁶ Vitamin c supplements of 100-200 mg per day orally or parenterally ensure rapid and complete cure. The clinical improvement is seen within a day to week in most cases,¹⁴ but the treatment should be continued for up to three months for complete recovery.⁶

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Fig.-1



Fig.-2

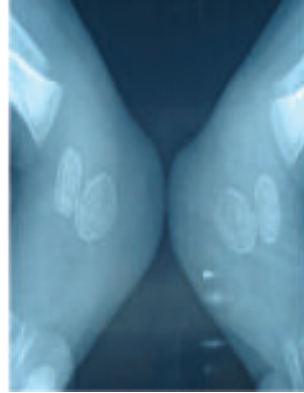


Fig.-3



Fig.-4

The requirement of vitamin C is increased during infections and diarrhoeal disease.¹⁷ When a mother's intake of vitamin C during pregnancy and lactation is adequate, the newborn will have adequate tissue levels of vitamin C related to active placental transfer;¹⁸ subsequently maintained by the vitamin C in breast milk or commercial infant formulas.¹⁹ Breast milk contains sufficient vitamin C to prevent deficiency throughout infancy.¹⁹ Therefore infantile scurvy is generally observed only in infants over the age of six months. The occurrence of scurvy before that age is extremely rare.²⁰ Infants at risk include those who are fed with evaporated or boiled milk, exclusive meat feeding and children with dietary restrictions due to neuropsychiatric or developmental disorders.¹¹ Neonates whose feeding has been delayed because of clinical conditions can also suffer from ascorbic acid deficiency.⁶ The neonate we described here was a 28 days old baby who was on dietary restrictions for a long time after birth due to perinatal asphyxia with neonatal sepsis and later presented to us with swelling of various joints and reduced activity.

Case:

A 27-days-old baby was admitted because of inability to move the limbs and swelling of both knee, elbow and wrist joints since 15 days of age. The baby was born by LUCS at term due to eclampsia of mother. The baby was asphyxiated and was admitted in a local hospital. Subsequently he developed sepsis in the NICU and was treated accordingly. According to the mother the baby was on intravenous fluid up to 10 days of his age. At the age of 14 days the baby was discharged with the diagnosis of PNA with HIE (stage-II) with microcephaly with PDA. Two days after going home, the mother noticed the above complaints which was increasing in severity. The clinical examinations

revealed marked tenderness in addition to the swelling of the knee, elbow and wrist joints which caused a certain degree of pseudo paralysis of the lower extremities. His OFC was 31.7 cm, LAZ was on 3rd centile and WLZ was on 25th centile. He gained only 100 gm of weight since his birth.

X-ray of the affected joints were done. Ground glass appearance of the long bones (fig -1,2,4), Ringing of the epiphysis (fig-3), Trümmerfeld zone (fig-1,2,3) and frankel's white line (fig-1,2,3) were observed. His CBC was normal and VDRL was negative for him and his parents. So a daily dose of 100 mg of vitamin C was given routinely to the infant. And within three days the tenderness was decreased significantly and after one week there was obvious clinical improvement with disappearance of tenderness and pseudoparalysis and gradual diminishing of the knee, wrist and ankle swelling.

Discussion

By the middle of the 20th century, technological developments including food processing and transportation combined to make scurvy a rarely encountered disease.²¹ Scurvy is less common in the pediatric population, but case reports still appear. The baby we described here is a 28 days old neonate who presented with enlargement of large joints and pseudoparalysis. It was a common presentation of scurvy among children.²² The baby also had microcephaly. Scurvy is well described in children with neurodevelopmental delay.²³ This baby had H/O withdrawal of oral feeding for prolonged period of time after birth due to hospital admission owing to perinatal complications which may had enhanced the condition.⁵ Radiological findings of the affected joints were typical of scurvy.¹⁴ We could not assess S. vit

C level due to lack of laboratory facilities though it is not essential to diagnose the case.¹⁴⁻¹⁶ After starting treatment with oral vit C, clinical improvement was observed within a week. Our case was unique in its character as scurvy below one year of age is seldom reported and neonatal scurvy is the rarest one. The median age for childhood scurvy was found to be 42 months by Tripany S et al.¹⁴ and 44.65 months \pm 30.50 months by Lampopas O et al.²² Two cases of neonatal scurvy were reported so far. First one was by Hirsch M et al.²⁴ This case was also diagnosed by typical radiological findings and was thought to be of congenital in origin.²⁴ the second one was described by Bhat BV et al.²⁵

Conclusion

Although rare, scurvy is still encountered in the pediatric population. Early diagnosis is vital as the disease is potentially fatal but easily curable disease. We highly recommend to consider administration of vitamin C when any baby is on long term parenteral nutrition.

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