

Infantile Scurvy Masquerading as Bone Tumour

Dear Editor,

Scurvy, a disease of dietary ascorbic acid deficiency, is uncommon today.¹ It is occasionally described in individuals with restrictive nutrition due to psychiatric neuromuscular or developmental disorders, traditional habits, malabsorption syndromes or dietary deficiency.²⁻⁴ Because of its rarity, varied clinical presentation and low index of suspicion on the part of the treating physician, it is frequently missed, delayed or misdiagnosed.⁵ In recent years, its diagnosis has been confused with osteomyelitis, septic arthritis, acute rheumatic fever, juvenile rheumatoid arthritis, domestic violence/child abuse, complex regional pain syndrome, vasculitis, neuroblastoma etc.⁶ We describe a case of advanced scurvy in a 6-year-old child with feeding difficulties mimicking a bone tumour of the distal end of the femur.

Case Report

A 6-year-old male child with a presumed diagnosis of malignant bone tumour of the distal femur was referred to our orthopaedic department. The parents reported that the child had developed a gradually progressive swelling around the right knee since the last 3 months. When the symptoms did not improve even after 1 month, the patient was referred to an orthopaedic surgeon. The radiograph at this time showed a large lesion involving the distal femur resembling a bone tumour. A fine needle aspiration cytology (FNAC) advised at this time documented “fragments of

fibromyxoid stroma with embedded cells having round to oval nuclei with granular chromatin and prominent nucleoli”. A provisional diagnosis of mesenchymal tumour with advice for a biopsy for characterisation of the lesion and confirmation of the diagnosis was suggested. It was at this stage that the child was seen by us.

The child complained of generalised musculoskeletal pains, weakness and inability to bear weight. He was listless and irritable, and resisted any examination. His growth was retarded with features suggestive of protein energy malnutrition. He also had widespread gingival swelling with bleeding. Besides the large swelling around the right knee (Fig. 1a), the child also had a swelling around the right shoulder. Both swellings were tense and globular with increased local temperature, stretched shiny skin and few dilated veins. They were firm to hard in consistency with ill-defined margins. The swelling appeared to be adherent to the underlying bone. Routine blood examination showed that the child had megaloblastic anaemia. The radiographs showed separation of the distal femoral and proximal humeral epiphysis of the right side (Figs. 1 b, c). There were also features of subperiosteal haematoma. A focussed nutritional history at this time revealed that due to feeding difficulties, because of generalised spasticity as sequelae of tubercular meningitis at the age of 2 years, the child had been on exclusive milk diet for many years. Based on the detailed history, clinical examination, laboratory and radiological evaluation, a provisional diagnosis of advanced scurvy with



Fig. 1. (a) Clinical photograph of the child showing a large tense swelling around the right knee. Radiographs of the right thigh with knee and right arm with shoulder shows (b) distal femoral epiphyseal separation with large soft tissue swelling mimicking a bone tumour (c) proximal humeral epiphyseal separation.

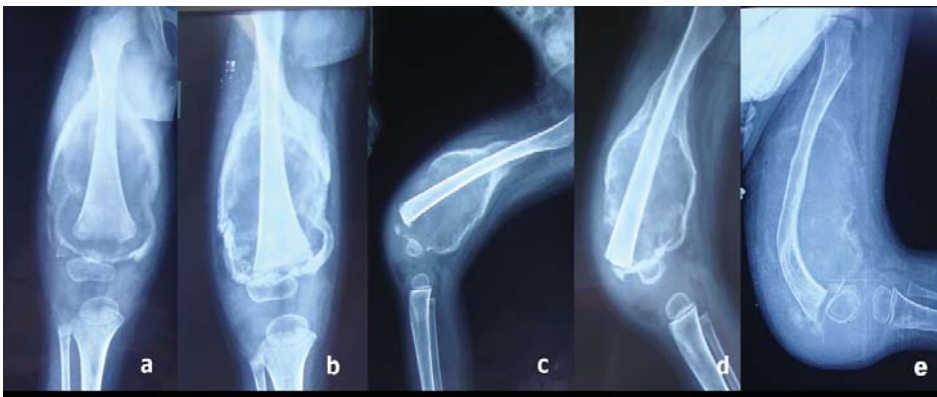


Fig. 2. Serial anteroposterior (a, b) and lateral (c, d, e) radiographs of the right thigh with knee after vitamin C administration, showing progressive calcification of the haematoma with remodelling.



Fig. 3. Serial anteroposterior radiographs of the right arm with shoulder after vitamin C administration, showing progressive calcification of the haematoma with remodelling.

grade IV protein energy malnutrition with megaloblastic anaemia with generalised spasticity was made.

The child was administered 200 mg of oral vitamin C, along with other supportive therapy. He was also started on a semi-solid diet rich in proteins and citrus fruits. The child's clinical condition started improving within 3 days of starting oral vitamin C supplementation. The psychomotor restlessness, muscular frailty and gingival bleeding diminished and the child became more alert and cheerful. The swelling also decreased in size and became harder in consistency. The local temperature decreased and the overlying skin became less tense. After one week, the patient was put in a one and a half hip spica cast for the femur and "U" cast for the proximal humerus. Serial radiographs demonstrated calcification of the haematoma with remodelling (Figs. 2 and 3). The cast was removed at 6 weeks and mobilisation started. At one year follow-up, satisfactory remodelling of the distal femur and proximal humerus was observed. Thus the diagnosis of scurvy was confirmed and extensive unnecessary investigations and interventions avoided.

Discussion

Vitamin C (Ascorbic acid) is a water soluble, enolic form of alpha-keto lactone. Humans are not able to synthesise vitamin C and therefore require an exogenous source for daily metabolic requirements. Deficiency of vitamin C intake results in scurvy characterised by defective collagen synthesis, osteoblasts dysfunction and impaired tissue repair. Musculoskeletal manifestations are present in 80% of patients and are prominent in the paediatric population.⁷

Scurvy has been called the eternal masquerader due to its varied clinical presentation.⁸ As most treating physicians may not have come across this condition during their clinical career, they have a low index of suspicion for it.⁵ This results in it being frequently missed, delayed or misdiagnosed.⁵ In our case, the child was initially thought to be a case of malignant tumour of the distal femur. The large and progressive swelling drew the treating physicians away from the other subtle findings like: gingival bleeding, generalised musculoskeletal pains and another swelling around the right shoulder. Moreover, because they did not suspect it, they were not able to interpret the radiographs properly to the extent that the child had to undergo an unnecessary FNAC.

Restrictive nutrition due to psychiatric, neuromuscular, developmental, traditional, habitual, malabsorption syndromes or dietary deficiency has been reported to be a cause of scurvy in recent years.^{2-4,9} Our patient had generalised spasticity of all the 4 limbs, and because of the resulting feeding difficulties, the parents had been keeping him on an exclusive milk and semi-solid diet which did not include any fresh fruit or vegetables. This fact was not picked up in the initial evaluation because a detailed dietary history was not elucidated at any time.

The classical radiological changes of scurvy; ground glass appearance, cortical thinning Fraenkels white line, Wimbergers sign and Pelkan spur (or corner sign) have been well described in the literature.¹⁰ However, epiphyseal separation and subperiosteal haemorrhage as seen in our case is less reported.⁶ The delayed ossification of the calcified degenerating cartilage columns creates a zone of weakness which results in epiphyseal separation. However, there is general consensus that these epiphyseal separations do not result in any permanent residual deformity or disturbance of longitudinal bone growth after conservative treatment.⁶ This is because the periosteum has its strongest attachment at the growth plate which is detached together with the epiphysis. Therefore, the epiphysis remains lined up to the denuded shaft by a periosteal envelop containing blood.⁶ Ascorbic acid administration results in rapid calcification and remodeling of this envelop with no residual deformity.

In conclusion, we describe a rare case of scurvy in a 6-year-old child that had been referred to us with a provisional diagnosis of a bone tumour. History revealed that the child had been on an oral milk diet because of feeding difficulties. Plain radiographs showed epiphyseal separation of the distal femur and the proximal humerus. Within 3 days of starting oral vitamin C supplementation and citrus fruits, the child showed significant improvement. The case highlights the importance of a detailed history, high clinical suspicion and through clinical examination in high-risk patients for the diagnosis of this now rare condition. Ascorbic acid dietary supplementation should be started on clinicoradiological suspicion. Correct diagnosis and treatment can result in excellent outcome and avoid extensive unnecessary investigations and interventions.

REFERENCES

1. Clemetson CA. Barlow's disease. *Med Hypotheses* 2002;59:52-6.
2. Swanson AM, Hughey LV. Acute inpatient presentation of scurvy. *Cutis* 2010;86:205-7.
3. Bari A, Javaid BK, Rehman S, Naz S. Scurvy: presenting as musculoskeletal pain. *J Coll Physicians Surg Pak* 2009;19:190-200.
4. Vitale A, La Torre F, Martini G, Calcagno G, Fede C, Conti G, et al. Arthritis and gum bleeding in two children. *J Paediatr Child Health* 2009;45:158-60.
5. Bursali A, Gursu S, Gursu A, Yildirim T. A case of infantile scurvy treated only with vitamin C: a forgotten disease. *Acta Orthop Belg* 2009;75:428-30.
6. Aroojis AJ, D'Souza H, Yagnik MG. Separation of the proximal humeral epiphysis. *Postgrad Med J* 1998;74:752-5.
7. Weinstein M, Babyn P, Zlotkin S. An orange a day keeps the doctor away: scurvy in the year 2000. *Pediatrics* 2001;108:E55.
8. Gupta P, Taneja K, Iyer PU, Murali MV, Tuli SM. Scurvy the eternal masquerader. *Ann Trop Paediatr* 1989;9:118-21.
9. Willmott NS, Bryan RA. Case report: scurvy in an epileptic child on a ketogenic diet with oral complications. *Eur Arch Paediatr Dent* 2008;9:148-52.
10. Brailsford JW. Some radiological manifestations of early scurvy. *Arch Dis Child* 1953;28:31.

Rehan Ul Haq, ^{1,2}MS (Orthopaedics), Ish Kumar Dhammi, ^{1,2}MS (Orthopaedics), Anil K Jain, ^{1,2}MS (Orthopaedics), Puneet Mishra, ^{1,2}MS (Orthopaedics), K Kalivanan, ^{1,2}MS (Orthopaedics)

¹University College of Medical Sciences, Delhi, India

²Guru Tegh Bahadur Hospital, Dilshad Garden, Delhi, India

Address for Correspondence: Dr Rehan Ul Haq, Department of Orthopaedics, UCMS and GTB Hospital, Dilshad Garden, Delhi, India 110095.
Email: docrehan@rediffmail.com