Rare disease

A good growth in a child with scurvy

Diletta Valentini,¹ Domenico Barbuti,² Annalisa Grandin,³ Laura Tanturri De Horatio,² Alberto Villani³¹Department of Pediatrics, Ospedale Bambino Gesù, Rome, Italy;²Department of Radiology, Bambino Gesù Roma, Rome, Italy;³Department of Pediatrics, Bambino Gesù Roma, Rome, ItalyCorrespondence to Diletta Valentini, diletta.valentini@opbg.net

Summary

We report on a 13-month-old boy who experienced pain while mobilising, and had bruising and swelling of the lower limbs. Laboratory examinations revealed anaemia and skeletal x-rays showed irregularity and thickening of the provisional zones of calcification of lower and upper limbs. The boy had been fed with only goat milk, homogenised meat, fruits and vegetables, all of which had been boiled together. Forty-eight hours after starting oral vitamin C supplementation, the patient showed dramatic clinical improvement. The clinical presentation, laboratory and imaging findings, together with the good response to vitamin C intake, allowed us to confirm the diagnosis of infantile scurvy. Scurvy is a disease that can be found among children, especially among groups with restrictive eating pattern. Fortunately, once diagnosed, scurvy is an easily treatable disease by administration of vitamin C and a correct diet that is rich in fruits, vegetables and fresh meat, all of which contained vitamin C.

BACKGROUND

A case of scurvy is discussed, which occurs rarely in developed countries. Scurvy is a disease caused by dietary deficiency of vitamin C. Its incidence in the paediatric population is uncommon, but case reports continue to appear.^{1 2} Groups at risk include infants who are fed evaporated or boiled milk, in which vitamin C is destroyed by exposition to heat, and children with dietary restrictions stemming from psychiatric or developmental disorders.³

Ascorbic acid is a necessary cofactor in collagen biosynthesis. The capillary fragility, responsible for the clinical manifestations of scurvy, is attributable to the depletion of pericapillary collagen.

The disease is characterised by petechiae, bruising and bleeding gums, and bone disease is a frequent manifestation of the condition in children and can be debilitating.⁴

CASE PRESENTATION

A 13-month-old boy was admitted to the Emergency Unit of our hospital in a poor general condition, unable to stand, affected by pain during mobilisation, and with bruising and swelling of the lower limbs. Brain and medulla MRI excluded the presence of cerebral masses and/or a medullary compression. The child was then admitted to the Pediatric Department.

The child was breast fed and received artificial milk since birth. Goat milk was given by the fourth month. He received vitamin A and D till he was 8 months old and iron supplementation from the fifth to the seventh month of life. Until the 10th month, he had normal growth and development. At 10 months, he showed increasing inappetence and, because of that, he was fed with only goat milk, homogenised meat, fruits and vegetables, all of which had been boiled together. Purpuric lesions, pain at mobilisation and soft tissue swelling of lower limbs had appeared over the 12th month.

Upon physical examination, the child was irritable, suffering and pale, with bruising on the legs and in a frog-like position. He weighed 11.5 kg and was 86 cm tall (75th and 97th percentile, respectively). Laboratory examinations revealed anaemia (haemoglobin 8.7 g/dl), low levels of iron (11 mg/dl; normal 40–140) and serum ferritin (17 ng/ml; normal 17–390), and an increase of erythrocyte sedimentation rate (30 mm/h) and lactate dehydrogenase (711 U/l; normal 100–600), and C3 and C4 levels (184 and 48 mg/dl, respectively; normal 65–130 and 8–32). Abdominal ultrasound and bone marrow aspiration excluded tumours.

INVESTIGATIONS

We performed radiographic studies because of the clinical suspicion of traumatic lesion. X-rays of the lower and upper limbs showed metaphysis thickening of the provisional zones of calcification ('Fraenkel's line'), radiotransparent transverse bands under metaphyseal limit, adjacent to Fraenkel's line (scurvy lines), and lateral metaphyseal spurs at the cartilage-shaft junctions (corner sign). The ossification centres showed central rarefaction with heavy ring shadows on the margins ('Wimberger's sign') (figure 1).

These clinical and radiologic findings led to the suspicion of a metabolic bone disease and, therefore, investigations were carried out for rickets and/or scurvy. The dosage of vitamin C was 10.4 µmol/l (normal: 30–80) and the doses of vitamin D, 1,25 (OH)₂D₃ and 25-OH-D₃, were 17.8 pg/ml (normal: 19.9–67) and 31.6 ng/ml (normal: >32.0), respectively.

Considering the above examinations, especially radiographic findings, infantile scurvy was diagnosed.

DIFFERENTIAL DIAGNOSIS

Diagnosis of scurvy can be difficult as it mimics other diseases. Among them, rheumatic disorders were excluded on laboratory examinations. Traumatic events and the

Figure 1 Metaphysis thickening of the provisional zones of calcification ('Fraenkel's line'), radiotransparent transverse bands under metaphyseal limit, adjacent to Fraenkel's line (scurvy lines), and lateral metaphyseal spurs at the cartilage-shaft junctions (corner sign). The ossification centres showed central rarefaction with heavy ring shadows on the margins ('Wimberger's sign').

'battered child syndrome' were taken into account, however, they were not supported by a positive history of trauma and the radiological findings were not characteristic. Brain and medulla MRI, abdominal ultrasound and bone marrow aspiration excluded a neoplastic disease.

One aspect of this case that needs to be addressed is whether this child's presentation can be attributed to a combination of rickets and iron deficiency.⁵ Although a component of this child's bone disease may have been exacerbated by vitamin D deficiency, the radiographic findings were not suggestive of rickets and the normal levels of calcium, phosphate, alkaline phosphatase and parathyroid hormone do not support vitamin D deficiency rickets as the primary cause of the child's bone disease.

TREATMENT

We quickly initiated oral administration of high doses of vitamin C (1 g/daily) and iron (6 mg/kg/daily).

OUTCOME AND FOLLOW-UP

Within 48 h of beginning therapy, the patient showed dramatic clinical improvement. He was able to stand and walk independently after 2 weeks of treatment. His haemoglobin increased to 13.5 g/dl and serum levels of iron and ferritin normalised.

Three months after starting vitamin C supplementation, the radiological findings were completely normalised (figure 2). The dosages of vitamin C and D were normal.

DISCUSSION

In 1883, the British physician Thomas Barlow described the clinical features of infantile scurvy.⁶ Nowadays, it is also known as 'Barlow's disease'.

Currently, the depletion of vitamin C is an under-recognised condition in Western populations. There are groups at risk, including infants who are fed with evaporated or boiled milk, in which ascorbic acid is destroyed by heat, as well as children with poor diets as a result of psychiatric or developmental disorders.⁷ Scurvy is still present in economically disadvantaged populations with poor nutrition.⁸

In our case, there was no economic disadvantage, but the inadequate intake of vitamin C was caused by an eating disorder that occurred during weaning. Patient's diet, in fact, has been only artificial milk and heated goat milk, up to the sixth month of life. Subsequently, there was a lack of dietary sources of vitamin C, because fruits, vegetables and meat were added to milk and cooked with it. As is known, this procedure leads to the destruction of vitamin C.

A particular aspect of our case is that despite the child growing well, he has a nutritional deficiency of ascorbic acid. Signs of scurvy develop after 1–3 months of inadequate vitamin C intake. Most cases of scurvy are seen between the ages of 6 and 24 months; in our patient, the onset occurred at 13 months.

Initial manifestations are aspecific: irritability, digestive disorders and decreased appetite. The earliest manifestations are dermatologic such as skin and mucosal petechiae, and ecchymosis.⁹ The gingival changes are frequent and characterised by swelling, bruising, bleeding and tooth loss.^{10 11}

Our patient, despite some of these initial events, has never shown relevant signs or symptoms to the oral cavity, but the presence of petechiae on the tongue reported by the mother.

Bone abnormalities, more frequent in children than in adults, are responsible for bone pain and functional

Figure 2 Three months after starting vitamin C supplementation, the radiological findings were completely normalised.

impotence.¹² Because of this, infants often hold their legs in a 'frog legs' position.¹³ Our patient has showed this antalgic position.

Anaemia is multifactorial in origin and is a characteristic sign of scurvy. The most common cause is iron deficiency because ascorbic acid improves absorption of iron by reducing it to the more absorbable ferrous state. In fact, in our patient, anaemia only improved after vitamin C supplementation.¹⁴

The radiological investigations in our case showed irregularity and thickening of the provisional zones of calcification of upper and lower limbs. The ossification centres show central rarefaction with heavy ring shadows on the margins. Those are the so called signs of Fraenkel and Wimberger, pathognomonic of scurvy.^{14 15}

Learning points

- ▶ Although rare, scurvy is a disease that can be found among children, especially among groups with restrictive eating pattern.
- ▶ The disease requires careful attention because unknown and untreated scurvy can be fatal.¹⁶
- ▶ Fortunately, on correct diagnosis, scurvy can be easily treated by administration of vitamin C and a correct diet, rich in fruits, vegetables and fresh meat, all containing vitamin C.

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Weinstein M**, Babyn P, Zlotkin S. An orange a day keeps the doctor away: scurvy in the year 2000. *Pediatrics* 2001;**108**:E55.
2. **Rosati P**, Boldrini R, Devito R, *et al*. A child with painful legs. *Lancet* 2005;**365**:1438.
3. **Hampl JS**, Taylor CA, Johnston CS. Intakes of vitamin C, vegetables and fruits: which schoolchildren are at risk? *J Am Coll Nutr* 1999;**18**:582–90.
4. **Shetty AK**, Steele RW, Silas V, *et al*. A boy with a limp. *Lancet* 1998;**351**:182.
5. **Barlow T**. On Cases Described as "Acute Rickets," which are probably a combination of Scurvy and Rickets, the Scurvy being an essential, and the rickets a variable, element. *Med Chir Trans* 1883;**66**:159–220.1.
6. **Rajakumar K**. Infantile scurvy: a historical perspective. *Pediatrics* 2001;**108**:E76.
7. **Cole JA**, Warthan MM, Hirano SA, *et al*. Scurvy in a 10-year-old boy. *Pediatr Dermatol*. Published Online First: 16 Jun 2010. doi: 10.1111/j.1525-1470.2010.01095.x.
8. World Health Organization. Complementary Feeding of Young Children in Developing Countries: A Review of Current Scientific Knowledge. WHO/NUT/98.1. Geneva: World Health Organization, 1998.
9. **Chatproedprai S**, Wananukul S. Scurvy: a case report. *J Med Assoc Thai* 2001;**84**(Suppl 1):S106–10.
10. **Larralde M**, Santos Muñoz A, Boggio P, *et al*. Scurvy in a 10-month-old boy. *Int J Dermatol* 2007;**46**:194–8.
11. **Adewumi AO**, Ashoor IF, Soares FM, *et al*. Eruption hematoma as a possible oral sign of infantile scurvy. *Pediatr Dent* 2010;**32**:151–5.
12. **Duggan CP**, Westra SJ, Rosenberg AE. Case records of the Massachusetts General Hospital. Case 23-2007. A 9-year-old boy with bone pain, rash, and gingival hypertrophy. *N Engl J Med* 2007;**357**:392–400.
13. **Bursali A**, Gursu S, Gursu A, *et al*. A case of infantile scurvy treated only with vitamin C: a forgotten disease. *Acta Orthop Belg* 2009;**75**:428–30.
14. **Ghedira Besbes L**, Haddad S, Ben Meriem C, *et al*. Infantile scurvy: two case reports. *Int J Pediatr*. Published Online First: 29 Sep 2010. doi: 10.1155/2010/717518.
15. **Riepe FG**, Eichmann D, Oppermann HC, *et al*. Special feature: picture of the month. Infantile scurvy. *Arch Pediatr Adolesc Med* 2001;**155**:607–8.
16. **Mimasaka S**, Funayama M, Adachi N, *et al*. A fatal case of infantile scurvy. *Int J Legal Med* 2000;**114**:122–4.

This pdf has been created automatically from the final edited text and images.

Copyright 2011 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Please cite this article as follows (you will need to access the article online to obtain the date of publication).

Valentini D, Barbuti D, Grandin A, De Horatio LT, Villani A. A good growth in a child with scurvy. *BMJ Case Reports* 2011;10.1136/bcr.10.2010.3383, date of publication

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow