



Minerva Access is the Institutional Repository of The University of Melbourne

Author/s:

Andrews, SL;Iyer, S;Rodda, C;Fitzgerald, J

Title:

Scurvy: A rare cause for limp in a child with autism spectrum disorder.

Date:

2018-12

Citation:

Andrews, S. L., Iyer, S., Rodda, C. & Fitzgerald, J. (2018). Scurvy: A rare cause for limp in a child with autism spectrum disorder.. *Journal of Paediatrics and Child Health*, 54 (12), pp.1375-1377. <https://doi.org/10.1111/jpc.14098>.

Persistent Link:

<https://hdl.handle.net/11343/284849>

CASE REPORT:

History:

A 6-year-old boy presented to a metropolitan hospital Emergency Department (ED) with a 10 day history of left sided limp, against a background of severe Autism Spectrum Disorder. He was non-verbal and incontinent. He had poor oral hygiene due to an aversion to mouth care. He had a longstanding very restricted diet and would only eat bread, cheese and water.

His mother had noticed a rash on his lower legs several weeks prior to the limp. He developed gum bleeding two weeks before presenting to hospital. There was no history of weight loss, fevers or lethargy.

Examination:

The mother brought the child to the ED as he was no longer weight-bearing. The child was distressed with examination due to his leg pain. He had a limp, pretibial rash and red swollen gums. There was no pallor, lymph node enlargement or hepatosplenomegaly. Despite no history of fevers at home, on presentation he had a temperature of 38.2 degrees.

Investigations:

The child was investigated with a full blood examination, electrolytes, coagulation profile, erythrocyte sedimentation rate and a C-Reactive protein. He had a raised ESR of 115, a CRP of 19, and a normal haemoglobin and white cell count. Left leg X-Rays demonstrated subtle soft tissue swelling displacing the fat pad from the femur with a faint metaphyseal lucency.

The child was admitted under the orthopaedic team and received an inpatient consultation from the general paediatric team. Osteomyelitis was suspected as the most likely diagnosis due to the combination of markedly raised ESR, fever and limp. Differential diagnoses of reactive arthritis, Henoch-Schönlein purpura, or evolving juvenile idiopathic arthritis were also considered. The gum bleeding was attributed to gingivitis, and Metronidazole was commenced.

The child underwent an MRI of his lower limbs and during the MRI the paediatric radiologist was called to review the rash and gum bleeding. The combination of clinical and systemic radiological abnormalities led to the eventual unifying diagnosis of scurvy, severe vitamin C deficiency.

The diagnosis was confirmed with a vitamin C level of 6 umol/L (Reference Interval 10-115). A dietary history of the patient was then taken and he was investigated for additional nutritional deficiencies. He had normal iron studies,

This is the author manuscript accepted for publication and has undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the Version of Record. Please cite this article as doi: [10.1111/jpc.14098](https://doi.org/10.1111/jpc.14098)

vitamin D, calcium, magnesium and phosphate levels. Folic acid, vitamin B12 and Vitamin A levels were not performed.

Management:

The patient was prescribed 100mg ascorbic acid TDS for 10 days, with a planned ongoing supplemental dose. He was discharged from hospital post diagnosis of scurvy and had follow up care with his private community paediatrician. The child re-presented several months later with gastroenteritis. He no longer had symptoms of scurvy and was taking a regular multivitamin that contained vitamin C.

Discussion:

Ascorbic acid (Vitamin C) is a water-soluble vitamin that is found in breast milk, citrus fruits and green vegetables. Vitamin C is a vital dietary micronutrient for humans. It is absorbed in the small intestine via active transport and the kidneys regulate the levels in the blood.¹ Ascorbic acid is essential in many metabolic pathways. It is involved in collagen synthesis, vitamin E and folate metabolism, fatty acid transport, neurotransmitter synthesis and prostaglandin metabolism.²

The clinical findings of ascorbic acid deficiency occur after months of poor dietary intake and are attributable to the effect of low levels of vitamin C on collagen containing tissues. Scurvy is a clinical diagnosis with features that may include bleeding and swollen gums, loosening of teeth, bone abnormalities, arthralgia, delayed wound healing, anaemia and classic skin findings of perifollicular hyperkeratosis, petechiae and purpura. Fever can occur in the later stages of the disease.³

Although Vitamin C deficiency remains a clinical diagnosis, there are radiological and biochemical investigations that support this diagnosis. ESR may be normal or elevated.⁴ This patient had X-Ray findings that can be found in, but are not pathognomonic for, scurvy. His presentation of fever, limp and elevated inflammatory markers raised the suspicion of the more common diagnosis of osteomyelitis. The patient underwent an anaesthetic and MRI to investigate for osteomyelitis/septic arthritis to determine need for acute surgery.

However, MRI findings of oedema of the metaphyseal marrow, periosteum and surrounding soft tissue are pathognomonic for scurvy.⁵ Scurvy often causes symmetrical changes to the long bones metaphysis, with oedema around the bone that extends into the surrounding soft tissue. Severe cases progress to frank subperiosteal haematomas or pathological metaphyseal fractures.⁶

The patient was prescribed Metronidazole for gingivitis. The first line treatment for gingivitis is improved oral hygiene, and the possibility of short-term use of

chlorhexidine mouthwashes. Antibiotics are not recommended the treatment of gingivitis, unless acute ulcerative gingivitis is present.⁷

Ascorbic acid levels reflect the levels of ascorbic acid in plasma, but not in the tissues. Normal ascorbic acid levels can occur if there has been recent improvement in dietary intake, despite ongoing low tissue levels. Treatment of scurvy is fortunately simple with patients requiring 100mg three times a day for one week followed by improvement in the diet, or ongoing supplemental dosing. Resolution of symptoms occurs within several weeks.⁸

A full dietary history for this patient was not taken on admission to hospital. Once the diagnosis of scurvy was made the child was screened for some nutritional deficiencies, yet as he was also at risk of vitamin A, B12 and folate deficiencies this should have been investigated. This case highlights the need to take a careful dietary history in children with Autism Spectrum Disorders, and manage accordingly.

Questions:

Q1. MRI findings of vitamin C deficiency include:

- A. Oedema of the metaphyseal marrow
- B. Oedema of periosteum and surrounding tissues
- C. Subperiosteal heamatomas
- D. Pathological metaphyseal fractures
- E. All of the above

Answer: E. All of the above.

Q2. Vitamin C (Ascorbic acid)

- A Is absorbed in the large intestine
- B Serum levels reflect tissue levels
- C Is a fat soluble vitamin
- D Levels are regulated by the kidneys
- E Is not an essential dietary micronutrient for humans

Answer: D. Ascorbic acid is absorbed in the small intestine, serum levels do not reflect tissue levels if there has been recent intake, is a water soluble vitamin and is an essential dietary micronutrient for humans.

Q3. Which answer is incorrect in regards to Scurvy?

- A Occurs after months of a vitamin C deficient diet
- B Children with restrictive eating patterns and underlying gastrointestinal disorders are at risk of this condition
- C Vitamin C deficiency requires confirmation of biochemical and radiological results to make the diagnosis
- D Replacement therapy with vitamin C improves symptoms rapidly
- E The clinical findings of scurvy are predominantly due the impact of low vitamin C levels on collagen synthesis

Answer: C. The diagnosis of scurvy remains a clinical diagnosis.

Key points:

- Restrictive eating patterns are common in children with developmental disorders and can lead to significant micronutrient deficiencies.
- Detailed history taking, and holistic view of the patient can assist in earlier detection of rare but important differential diagnoses.

- Scurvy is a rare diagnosis with classic clinical features.

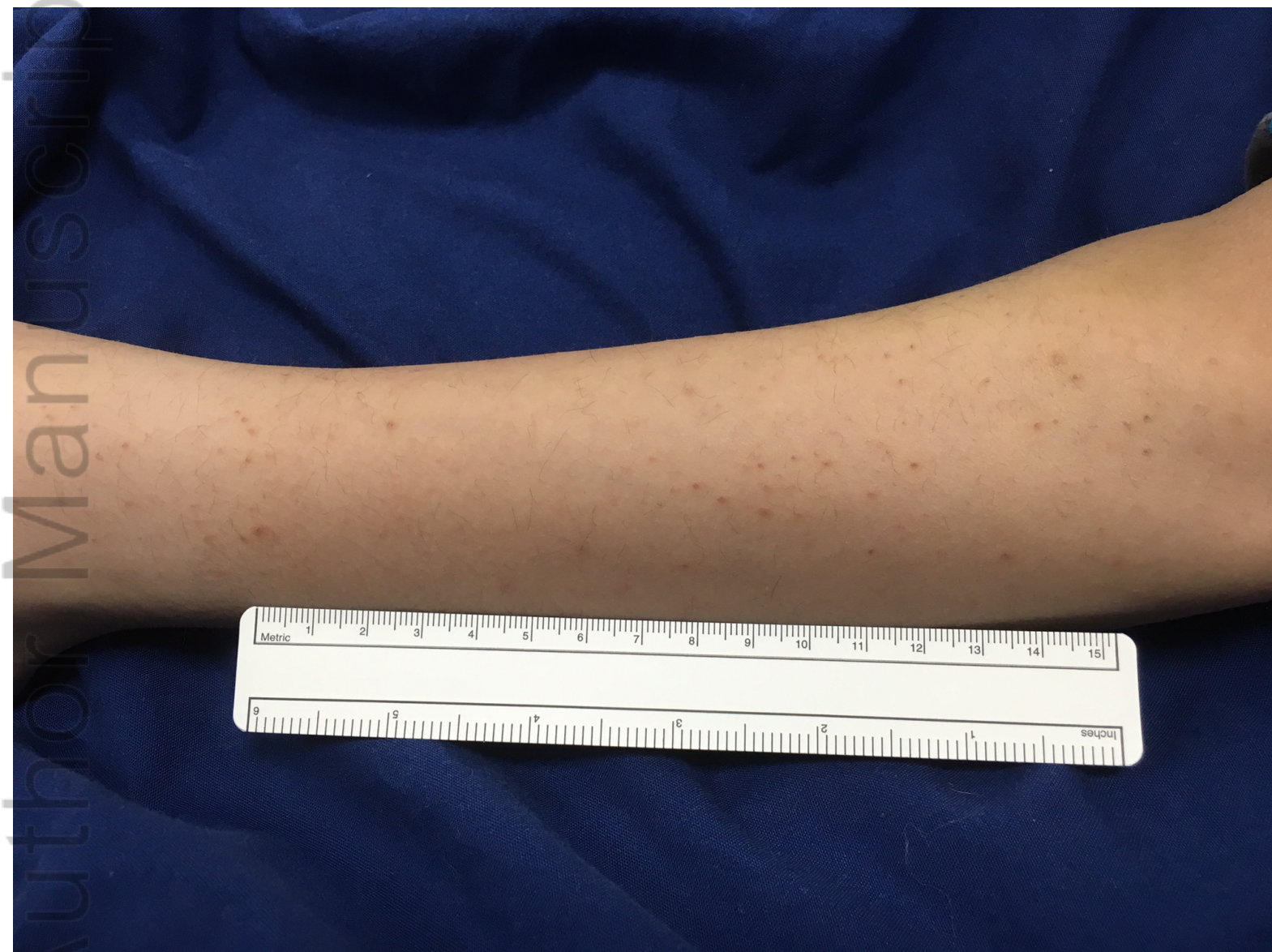
References:

- ¹Pazirandeh S, Burns DL. Overview of water-soluble vitamins. Waltham, MA: UpToDate; 2017. Available from: www.uptodate.com.
- ² Sherwood L. Human Physiology. From Cells to Systems. Eighth Edition. Alexander S editor. California: Brooks/Cole; 2013.
- ³ Aghajanian P, Hall S, Wongworawat MD, Mohan S. The Roles and Mechanisms of Actions of Vitamin C in Bone: New Developments. J Bone Miner Res. 2015 Nov;30(11):1945-55
- ⁴ Hafez D, Saint S, Griauzde J, Mody R, Meddings J. A Deficient Diagnosis. N Engl J Med. 2016 Apr 7;374(14):1369-74
- ⁵ Gulko E, Collins LK, Murphy RC, Thornhill BA, Taragin BH. MRI findings in pediatric patients with scurvy. Skeletal Radiol. 2015. Feb;44(2):291-7
- ⁶ Gongidi P, Johnson C, Dinan D. Scurvy in an autistic child: MRI findings. Pediatr Radiol. 2013 Oct;43(10):1396-9
- ⁷ Therapeutic Guidelines Limited (2018). Oral and Dental Therapeutic Guidelines (Periodontal Disease). Retrieved from <http://www.tg.org.au>
- ⁸ American Academy of Pediatrics. Water-soluble vitamins. In: Pediatric Nutrition, 7th ed., Kleinman RE, Greer FR (Eds), American Academy of Pediatrics, Elk Grove Village 2011. p.527.





MRI.jpg



Skin findings .jpg