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The return of the old masquerader—scurvy: a case report of pediatric limping and bone pain

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Abstract

Background Scurvy has been a rare condition in the developed world due to the increasing availability of adequate nutrition and easy access to fruits and vegetables. Scurvy is caused by severe vitamin C deficiency and has been historically associated with malnutrition in high-risk populations such as sailors. As it is an uncommon nutritional deficiency, scurvy is seldom suspected and usually under-diagnosed especially in the pediatric population.

Case presentation We report a case of a pediatric patient with a history of autism spectrum disorder presenting with atypical symptoms of scurvy with gait abnormalities, diffuse rash, musculoskeletal deconditioning, and bleeding manifestations. After multiple extensive investigations and a prolonged hospitalization, a detailed dietary analysis revealed a severely restricted diet devoid of fruits and vegetables. Physical examination revealed specific signs of corkscrew hair with peri-follicular hemorrhages, hypertrophic, and bleeding gums with a concurrent low vitamin C level. Treatment with vitamin C supplementation was initiated and he was ambulant within 1 month of discharge.

Conclusion This case highlights the importance of nutrition history for determining micronutrient deficiency in children with special needs. Enhanced physician education and higher index of suspicion regarding nutritional deficiencies will avoid over-investigations and delays for the institution of specific treatment.

Keywords Ascorbic acid, Vitamin C, Pediatric scurvy, Malnutrition, Limping

Background

In pediatrics, nutritional deficiencies of vitamin C (ascorbic acid) leading to scurvy are rare in the modern era. Children at risk include those with restricted dietary habits or inadequate food intake secondary to developmental disorders or neuropsychiatric conditions [1, 2], dietary preferences [1], specific dietary needs (ketogenic diet for epilepsy) or gastrointestinal disorders such as inflammatory bowel disease or coeliac disease causing reduced absorption of vitamin C. In otherwise healthy children, we have seen a re-emergence of nutritional deficiencies secondary to the increasing consumption of highly processed food [3], atypical dietary preferences and new diet fads.

Vitamin C is important in multiple pathways including collagen synthesis, neurotransmitter processing, oxidative repair mechanisms and immune system function [4]. Vitamin C improves iron absorption and its deficiency can lead to the concomitant presentation of anemia [5]. In children, the most common presentations of scurvy involve the musculoskeletal system and can range from bone pain, joint swelling, limping and refusal to ambulate or weight-bear [6–10]. Many of these patients have been misdiagnosed by attending pediatricians due to

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the rarity of this condition [11–13]. We report an interesting presentation of a 5-year-old child with biochemical and radiographic findings of scurvy who presented with a myriad of multiple-organ symptoms that caused a diagnostic dilemma leading to prolonged and repeated hospitalization.

Case presentation

A 5-year-old boy with severe autism spectrum disorder (ASD) presented to the outpatient neurology clinic with concerns of intermittent bilateral lower limb weakness with pain for 2 years. He was non-verbal and had behavioral concerns. He was admitted from the clinic for further investigations as he became non-ambulant, and his symptoms were progressive over the preceding 2 months causing significant concerns for possible underlying degenerative neuromuscular condition.

He had intermittent symptoms of bone pain for 2 years with occasional gait abnormalities of limping and refusal to weight-bear. During these episodes of decompensation, he was reviewed by emergency physicians and pediatricians who diagnosed him with various conditions that ranged from transient synovitis, arthritis, myositis to growing pains to attribute to his symptoms. His neurological examination was always reported to be normal during these medical reviews.

He was admitted 1 year back for further work-up when his symptoms worsened causing him to be non-ambulant. Blood investigations revealed microcytic hypochromic anemia secondary to iron deficiency but the rest of the blood tests for kidney function tests, liver function tests, thyroid function tests and muscle enzymes were normal. Magnetic resonance imaging (MRI) spine, bone scans and positron emission tomography (PET) were performed and they ruled out any underlying spinal pathology or tumors. He was then discharged with physiotherapy (PT) sessions to re-condition his muscle strength. His parents defaulted to regular follow-up with the neurology team and discontinued PT.

He re-attended neurology clinic after 1 year with additional symptoms of bilateral ankle swelling with tenderness which were concerning then of a possible rheumatological condition. In view of his multi-system presentations, he was admitted again for further detailed investigations.

On physical examination, he appeared small for his age with failure to thrive and was generally very irritable. His height was 100 cm (25% for age) and weight was 12.7 kg (3% for age) with a corresponding body mass index (BMI) of 12.7 (< 3% for age) which meant he was significantly underweight (Table 1). There was pitting edema of the ankles with tenderness over the bilateral ankle joints. He was unable to bear weight but his hip joint examination was normal.

He had conjunctival pallor suggestive of anemia. There was no clinical evidence of anemic heart failure as he did not have generalized edema or appear breathless. There was gum hypertrophy with contact bleeding on examination of his oral cavity. He had petechial rash over his forearms with peri-follicular hemorrhages on closer inspection of his skin. The rest of his systemic examination including the neurological examination was normal.

Blood investigations revealed microcytic hypochromic anemia with low iron levels (Table 2). There was no evidence of coagulopathy as his platelet and PT/INR levels were normal. More detailed blood investigations were done to investigate for the cause of persistent iron-deficiency anemia and failure to thrive. Multiple nutritional deficiencies including low folate, vitamin B12, calcium, phosphate, and vitamin D levels were detected (Table 2). Long bone radiographs revealed diffusely osteopenic bones with metaphyseal fraying and splaying, horizontal linear sclerotic bands over metaphyses (Fig. 1). MRI of the ankles and spine showed periosteal reaction over the distal tibia with edema of the surrounding muscles and subcutaneous fat. Bone marrow analysis (BMA) and bone biopsy of the ankle did not reveal any underlying osteomyelitis or malignancy.

Further blood tests to screen for other nutritional deficiencies revealed low vitamin C levels that cemented his eventual diagnosis of scurvy. In view of his new diagnosis, his skin was examined closely again which revealed the pathognomic sign of corkscrew hair over hyperkeratotic

Table 1 Anthropometric measurements of our patient

Parameters (units)	Observed	Expected (at 50% for age)	Centiles	Interpretation
Weight (kg)	12.7	15.8	3%	Underweight
Height (cm)	100	103	25%	Normal height
BMI	12.7	15.2	< 3%	Low BMI, Low weight for height measurements

Table 2 Blood investigations for our patient

	Patient's value	Reference range
Hemoglobin	5.7	11.4–14.2 g/DL
White blood cell count	8.65	$5.22–13.35 \times 10^{9}$/L
Platelet count	290	$140–440 \times 10^{9}$/L
Creatinine kinase	37	20–310 U/L
Aldolase	5	1.3–6.3 U/L
Iron	1.5	2.8–22.9 µmol/L
Free thyroxine	15.6	10.3–25.7 pmol/L
Thyroid stimulating hormone	2.38	0.50–4.50 mIU/L
INR (International Normalized ratio)	1.12	2.0–3.5
Prothrombin time	14	12.1–14.5 s
Vitamin B12	198	209–1190 pmol/L
Folate	9.7	> 27.90 nmol/L
Calcium	2.14	2.30–2.63 mmol/L
Phosphorus	1.2	1.3–1.9 mmol/L
Vitamin D (25 Hydroxyvitamin D)	6.7	20.0–100.0 ng/mL
Vitamin K	0.15	0.2–3.2 ng/mL
Vitamin B1	80	70–180 nmol/L
Vitamin A	0.18	0.20–0.43 MG/L
Vitamin C	< 0.1	0.4–2.0 mg/dL

follicles. It was missed previously as the skin was not examined at such close range.

Thereafter, he was managed by a multi-disciplinary team for nutritional rehabilitation. In view of symptomatic anemia, he required a blood transfusion. Subsequently, he was also initiated on multiple nutritional supplementations: vitamin C 400 mg/day, iron drops 80 mg/day (6 mg/kg/day), calcium 500 mg/day, vitamin D3 2000 unit/day, folic acid 5 mg/day. They were titrated with the advice of the dietician during the inpatient review. He received these supplements for 3 months with biochemical and clinical improvement in his nutritional status. Endoscopy did not reveal any underlying gastrointestinal pathology or malabsorptive conditions to account for the multiple nutritional deficiencies. He was noted to have poor dietary habits and a detailed nutritional assessment revealed a limited variety of food intake. He rarely consumed any fruits or vegetables, rejected all forms of milk feeds and predominantly consumes carbohydrates in the form of plain crackers, rice or bread. This has been his diet since he started weaning diet at 6 months of life.

His restricted dietary preferences were attributed to his underlying condition of ASD which caused him to be a picky eater. He was discharged with the dual diagnoses of scurvy and vitamin D-dependent rickets which explained his predominantly musculoskeletal symptoms. On follow-up, his anemia had resolved within 4 months of iron therapy (from 5.7 g/DL to 11.1 g/DL). His musculoskeletal symptoms completely resolved and he was ambulant after initiating nutrition supplementation within 1 month post-discharge.

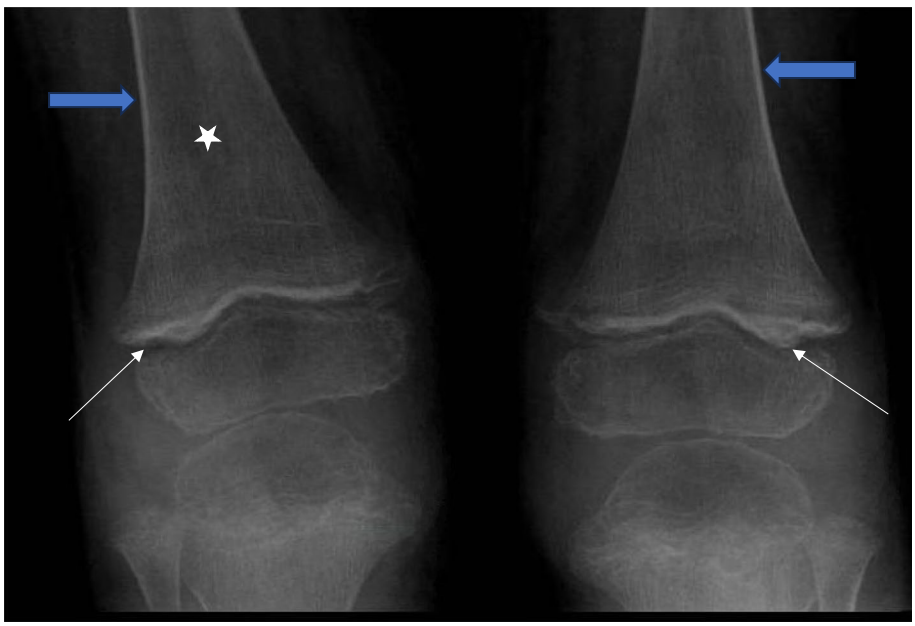

Fig. 1 Anteroposterior radiograph of knee joint reveals metaphyseal dense bands in bilateral distal femoral metaphyses (white arrows). Bones appear generally osteopenic (star sign) and there is pencil-thin cortex (blue arrows). Similar findings are also seen in bilateral proximal tibial physes. The findings are characteristic for scurvy

Discussion

Deficiency of vitamin C results in the clinical manifestations of scurvy which is the oldest known condition related to malnutrition [14]. English sailors [15, 16] knew that the disease could be prevented by taking fresh lime. Compared to other nutritional deficiencies, it is seldom suspected which leads to delayed recognition of this disorder [2]. At-risk children are those with abnormal dietary habits, mental illness or physical disabilities. The disease spectrum is quite varied as vitamin C is required in multiple biological processes [14]; but the most common presentations usually involve the musculoskeletal system in pediatrics [2, 17, 18] with refusal to ambulate [13] or limping [1, 6, 19] gait, bone pain, joint swelling, hemarthrosis or pseudo-paralysis [20, 21]. Scurvy is an eternal and great masquerader and is often misdiagnosed with osteomyelitis, arthritis and leukemia [21–26].

Our patient had non-specific symptoms of anorexia, malaise, lethargy and irritability; all of which have been reported previously in scurvy [6]. Specific symptoms to scurvy that were present in our patient were corkscrew hair, peri-follicular hemorrhages, purpura and swollen bleeding gums [1, 11, 27].

Our patient's X-rays revealed typical features of scurvy: osteopenia [28] with metaphyseal dense bands (Fig. 1). Other common radiological findings include periosteal elevation, subperiosteal hemorrhages [2] or calcifications [18, 29, 30], metaphyseal spurs, metaphyseal calcification [18] and sclerotic epiphyseal rim [28]. MRI findings in our patient were also consistent with scurvy as he had periosteal reaction with surrounding edema [18]. Other MRI findings can be varied from non-specific focal changes in bone marrow [31] or metaphyseal signal changes [28, 31].

Common differentials that were considered in our patient were arthritis in view of ankle joint tenderness and swelling. However, the inflammatory markers were normal: CRP 4.3 (0–5 mg/L), ESR 14 (1–10 mm/h). Furthermore, there were no joint stiffness or limitations in the range of motion of the ankle joints. Osteomyelitis was another differential due to the prolonged duration of symptoms with bone pain but the bone biopsy ruled out any infective process. Another worrying differential considered was leukemia but this was ruled out with a normal BMA and bone biopsy. As the patient had easy bruising with petechiae, hematological conditions were considered; but the blood investigations revealed a normal platelet count and coagulation profile.

The diagnosis of scurvy in our patient was based on a combination of specific symptoms with clinical and radiographic findings. The diagnosis was confirmed with the concomitant low blood ascorbic acid levels.

Vitamin C deficiency may occur concurrently with other nutritional deficiencies such as thiamine (vitamin B1), pyridoxine (vitamin B6), cobalamin (vitamin B12), and vitamin D [32]. It is thus important to screen for macronutrient and micronutrient deficiencies to ensure adequate nutritional supplementation is administered in such high-risk children.

Treatment is relatively simple with oral or intravenous vitamin C supplementation. Many cases have reported resolution of musculoskeletal impairments within 1 month of initiating nutritional supplementation [17, 33] which was similar to our patient.

Conclusion

As scurvy is a great mimicker of more concerning differentials, many cases have been subjected to over-investigations and invasive procedures with costly medical bills. Scurvy should be considered when a child presents with musculoskeletal symptoms such as limping or refusal to ambulate especially if the patient has normal neurological and rheumatological physical examination or has high-risk conditions that lead to significant dietary restrictions. A high index of suspicion is required when faced with such atypical constellation of symptoms with focus on a meticulous clinical history and nutritional assessment to consider the possibility of nutritional disorders attributing to these symptoms.

Abbreviations

ASD	Autism spectrum disorder
MRI	Magnetic resonance imaging
PET	Positron emission tomography
PT	Physiotherapy
BMA	Bone marrow analysis

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Authors' contributions

XC: data curation, editing, writing of original draft preparation, and investigation. CHWW: data curation and investigation. SRC: conceptualization, investigation, supervision, reviewing, and editing.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
Ethics approval was not required by our local institution as this is a single case report. Written and verbal parental consent was taken for sharing of patient's data and for publication.

Consent for publication
Written and verbal consent was taken from parents.

Competing interests
The authors declare that they have no competing interests.

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