Scurvy still exists in children: A case report

Scorbut infantile: A propos d'une observation

Sarra Ben Ahmed, Fatma Mezghani, Samar Rhayem, Faten Fedhila, Samir Hadded

Hôpital d'enfants Béchir Hamza de Tunis, Faculté de médecine de Tunis, Université Tunis El Manar

Abstract

Background: Scurvy is one of the oldest diseases known to mankind. Although rare lately, the clinical suspicion arises in front of a precarious situation or deficient nutrition and food restriction secondary to a psychiatric condition, even in patients with non-specific complaints.

We report the observation of a 6 -year- old boy, followed for autism since the age of 3 years and who was admitted for limping, hemorrhagic syndrome, arthritis and weakness. The diagnosis of child abuse was initially suspected but clinical and radiological abnormalities seen were characteristic of scurvy. Vitamin C level was undetectable. The child had an unbalanced diet.A favorable outcome was rapidly obtained following supplementation.

Scurvy is rare, but it should be mentioned among children with psychiatric disorders, presenting with musculoskeletal manifestations or hemorrhagic syndrome. It is essential to prevent it by systematic dietary supplementation of vitamin C in children with eating difficulties.

Keywords: Scurvy, hemorrhagic syndrome, arthritis

Résumé

Le scorbut (maladie de Barlow) est une pathologie historique, conséquence d'une alimentation carencée en vitamine C. Bien que rare ces derniers temps, le scorbut doit être suspecté chez certaines populations pédiatriques qui sont plus à risque comme les enfants autistes ayant une sélectivité alimentaire.

Nous rapportons l'observation d'un garçon de 6 ans, suivi pour autisme depuis l'âge de 3 ans et admis pour boiterie, syndrome hémorragique et arthrite. Le diagnostic de sévices à enfants a été initialement suspecté mais les anomalies cliniques et radiologiques observées étaient caractéristiques du scorbut. Le taux de vitamine C était indétectable. L'enfant avait une alimentation carencée. Une évolution favorable a été rapidement obtenue suite à la supplémentation vitaminique.

Le scorbut est une maladie rare, mais elle doit être évoquée chez les enfants atteints de troubles du comportement alimentaire, présentant des manifestations musculo-squelettiques ou un syndrome hémorragique. Il est indispensable de la prévenir par une supplémentation systématique en vitamine C chez les enfants à risque.

Mots clé : Scorbut, syndrome, hémorragique, arthrite

Correspondance
Sarra Ben Ahmed
Hôpital d'enfants Béchir Hamza de Tunis / faculté de médecine de Tunis
E-mail : sarrabenahmed21@gmail.com

INTRODUCTION

Scurvy is a historical disease that results from a deficiency of vitamin C (1). In the pediatric population, scurvy is the result of a serious abnormal diet behaviour (delay in starting weaning foods, improper dietary habits, developmental problems) (2). Despite the ease of treatment, scurvy frequently goes undetected for extended periods of time and can even be fatal. It requires an attentive physician to recognize the signs and symptoms and consider the diagnosis, especially in children with severe dietary restrictions secondary to developmental delay or autism spectrum disorder (3). Our case is an opportunity to recall the clinical and radiological manifestations of this forgotten historical disease.

OBSERVATION

We report a case of a 6-and-a-half-year-old child, admitted to our Pediatric Department for walking disorders and hemorrhagic syndrome. Parents were not blood related. Our patient was the second twin of an uncomplicated pregnancy. Both twins developed at the age of three autism disorder features and parents got divorced after. Neurosensory assessment was normal. Additionally, our patient had non-functional left kidney. The children lived in an underprivileged environment. The history of the disease dates back to three weeks, marked by the onset of multiple-daily gingivorrhagia, followed by the appearance of swelling in the left wrist, with deterioration of general condition, anorexia and unstated weight loss. On examination, the child was asthenic, feverish and unable to walk. He was eutrophic and had skin-mucous paleness. Cutaneous findings include petechiae, ecchymoses, hyperkeratosis, corkscrew hairs, and perifollicular hemorrhage, impetiginous lesions on the lower limbs, bruises on both arms (figure 1), swelling regarding the distal end of the left radius (figure 2), gingival hypertrophy (figure 3) with foul breath and a very poor oral state with multiple cavities.

Blood assessment shown he had normochromic normocrocytic anemia (hemoglobin: 7,7g/dl), thrombocytosis (Blood platelets: 530 000/mm3), mild chronic kidney failure (Renal clearance: 56ml/min/1,73 m2cs) and biological inflammatory syndrome (White blood cells: 20 000/mm3, CRP: 113mg/l). Hemostasis assessments as well as blood smear were normal (Prothrombin time = 90%).Given the socio-psychological status, child abuse was suspected. The x-rays of the wrist revealed an epiphyseal radiopaque band, metaphyseal spurs, epiphyseal densification and a thickening of soft tissue opposite the lower end of the radius and ulna with no fracture. The ultrasound of the left wrist revealed tenosynovitis of the extensors of the left wrist. Scurvy was then suspected. Diet history shown long-term abnormal behaviour with restrictive food that excluded fruits and vegetables and relied only on cereals. Phosphocalcic balance revealed vitamin D deficiency (9.26 ng / ml) and vitamin c were undetectable. Vitamin c supplementation (1 gram a day) associated with nutritional and other vitamins support lead to immediate and complete improvement with disappearance of the hemorrhagic syndrome, weakness and arthritis. The child was able to walk on the fourth day. He was less aggressive. Psychological and nutritional care has been established.

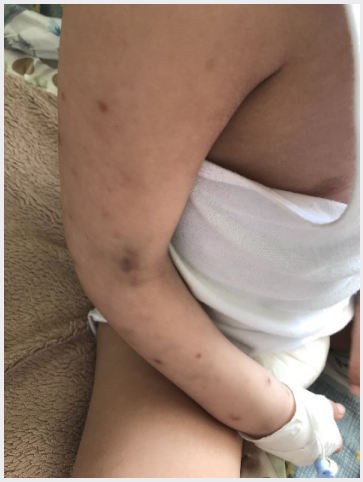

Figure 1. bruises on the right arm.

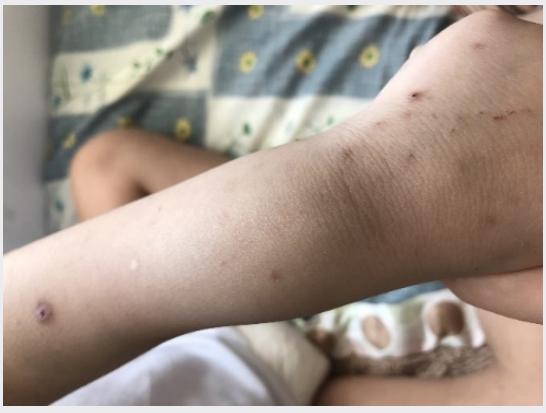

Figure 2. swelling regarding the distal end of the left radius.

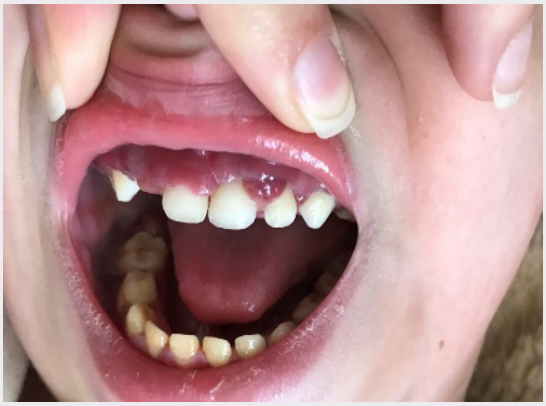

Figure 3. gingival hypertrophy.

DISCUSSION

We reported an original case of scurvy in a six-and-a-half-year-old child. This is an atypical presentation suggesting other differential diagnoses of hemorrhagic syndrome. This observation emphasizes the value of a careful interrogation and especially a dietary investigation to support the diagnosis. The current literature consists of mostly case reports or small case series (1). Our patient had almost all signs of the disease while authors usually report minimal clinical features (2). Our diagnosis were rapid and accurate while authors often report diagnostic delay and unnecessary explorations (2-4).Typically, the first signs of scurvy are nonspecific, including fatigue, anorexia and weight loss (3,4). Those signs where not reported in our patient because of his psychiatric condition and his familiar situation. Scurvy is a preventable disease caused by poor intake and/or absorption of vitamin C. Fruits and vegetables are the primary dietary source of this essential vitamin (2). Food selectivity and dietary avoidant behavior in our patient lead to severe deficiency. Sensory sensitivities are common in autism and this contributes to the picky eating habits as they create challenges adjusting to new foods, textures and tastes. Indeed, scurvy is particularly found in children with food selectivity, as evidenced by the study of the team from Boston Children's Hospital, where the diagnosis of scurvy was made in seven children, all with a developmental disorder and autism was the most common (1). The eventual insistence on eating the same foods predisposes to nutritional derangements and a diet devoid of fresh fruits and vegetables or vitamin C-fortified foods is a major risk factor for scurvy (2).For all these reasons, autism is the most common developmental disorder seen in 57 % of cases of scurvy (1). Given the clinical polymorphism of scurvy, several etiologies can have the same presentations (2). Child abuse was the first diagnosis we discussed. In the absence of a fracture, both mucosal and cutaneous signs with no hemostasis disorder, musculo skeletal signs made the diagnosis almost certain. As previously seen in our patient, cutaneous findings are frequently found in the lower extremities because the capillaries also face hydrostatic pressure (5). Many mucosal and cutaneous manifestations are secondary to blood vessel fragility. Therefore, the purpura is vascular, and no hemostasis disorders are associated (6). Walking inability is secondary to musculo-skeletal damage. A case series from Thailand reported that 96% of children diagnosed with scurvy presented with inability to walk. In this cohort, 96% also had limb pain, 43% had gingival bleeding, 46% had lower extremity joint swelling, 36% had gum hypertrophy, and 3.6% had petechial hemorrhage (7). In typical cases of childhood scurvy, X-ray shows demineralization and subperiosteal hemorrhage secondary to increased bleeding risk (8). In our patient, metaphyseal spurs could lead to corner fracture explaining the pain (Pelkin spur) (8) and disorganized bone production lead to sclerotic epiphyseal rim (Winberger Ring) (8). Scurvy is preventable and treatment is uncomplicated. There is not a standardized protocol, but the literature suggests 100-300 mg daily in children and 500–1000 mg daily in adults for 1 month, or until symptoms resolve (9). Clinical features and vitamin c deficiency in our patient were severe explaining the higher doses we prescribed. Working closely with a nutritionist provides parents the guidance they need to expand their child's repertoire of foods and in most cases to add supplemental vitamins to the regimen (1). Similarly, to our patient, the hemorrhagic syndrome resolves within 48 hours and the overall improvement occurs within 15 days in the reported cases (10).

CONCLUSIONS

Our case emphasizes the importance of obtaining a diet history in children with psychiatric disorders, presenting with musculoskeletal manifestations or hemorrhagic syndrome. It is essential to prevent scurvy by systematic dietary supplementation of vitamin C in children with eating difficulties.

REFERENCES

1. Ma NS, Thompson C, Weston S. Brief Report: Scurvy as a Manifestation of Food Selectivity in Children with Autism. J Autism Dev Disord. 2016;46(4):1464-70
2. Lund RM, Becker ML, Shapiro S, Allison T, Harris JG. Scurvy presenting with limp and weakness: a case report. BMC Pediatr. 2019;19(1):228
3. Olmedo JM, Yiannias JA, Windgassen EB, Gornet MK. Scurvy: a disease almost forgotten. Int J Dermatol. 2006;45:909-13
4. Léger D. Scurvy: reemergence of nutritional deficiencies. Can Fam Physician.2008;54(10):1403-6
5. Karthiga S, Dubey S, Garber S, Watts R. Scurvy: MRI findings. Rheumatology. 2008; 47(7):1109
6. ouati Y, Oquendo B, Raphael M. Purpura and Scurvy. Ann Fr Med Urgence .2018;8:34-6
7. Ratanachu-ek S, Jeerathanyasakun Y, Sukswai P, Wongtapradit L. Scurvy in pediatric patients: a review of 28 cases. J Med Assoc Thail. 2003;86:734-40
8. Ghedira Besbes L, Haddad S, Ben Meriem C, Golli M, Najjar MF, Guediche MN. Infantile scurvy: two case reports. Int J Pediatr. 2010:717518
9. Agarwal A, Shaharyar A, Kumar A, Bhat MS, Mishra M. Scurvy in pediatric age group-A disease often forgotten?. J Clin Orthop Trauma. 2015;6(2):101-7
10. Brennan CM, Atkins KA, Druzgal CH, Gaskin CM. Magnetic resonance imaging appearance of scurvy with gelatinous bone marrow transformation. Skeletal Radiol. 2012;41(3),357-60