Case Report

A Rare Case of Scurvy in an Otherwise Healthy Child: Diagnosis Through Oral Signs

Christian Bacci, DDS[1] • Stefano Sivolella, DDS[2] • Jacopo Pellegrini, DDS[3] • Lorenzo Favero, MD, DDS[4] • Mario Berengo, MD, DDS[5]

Abstract: *The purpose of this paper was to report the case of a 2-year-old Caucasian female who was referred with a presumed diagnosis of pediatric rheumatoid arthritis. The patient presented widespread gingival swelling with bleeding, sharp pain, and halitosis. The patient also presented pain and swelling of the right knee joint, and psychomotor restlessness associated with muscular frailty. Little compliance on the part of both the patient and parents was also noted. Oral manifestations, together with an accurate medical history, led to the diagnosis of infantile scurvy, caused by an inadequate dietary supply of vitamin C. Administering 250 mg of ascorbic acid orally twice a day led to the remission of gingival swelling and of the other symptoms. The parents were advised to feed the child appropriate foods. Nutritional problems are traditionally linked to an insufficient availability of food, but can also be associated with child- or family-related psychological problems. (Pediatr Dent 2010;32:) Received June 5, 2009 / Last Revision August 17, 2009 / Accepted August 20, 2009*

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Scurvy, a rarely found pathosis caused by a deficiency of vitamin C, is characterized among other symptoms by oral manifestations (swelling and gingival bleeding) and rheumatic-like symptoms.[1-4] In Western countries, it can be present in environmental or family conditions of malnutrition due to difficulty in accessing primary food resources. It can also be related to psychological or developmental problems.[5,6]

The purpose of this paper was to report a case of nutritional vitamin C deficiency which had been misdiagnosed as a rheumatologic disease. Oral signs of massive gingival swelling and bleeding helped in the definition of the correct diagnosis.

Case description

A 2-year-old Caucasian female patient was referred to the Pediatric Rheumatology Unit, Department of Oral Surgery, University of Padova, Padova, Italy, for a gingival biopsy. An oral examination revealed widespread bilateral, upper, and lower gingival swelling, sharp pain with a tendency to bleed, and halitosis (Figure 1). The patient also presented pain and swelling of the right knee joint and psychomotor restlessness associated with muscular frailty. Marginal compliance on the part of the patient and her parents was also noted; the parents, in fact, seemed to have an extremely permissive parenting style and be unable to follow a physician's instruction. Bowel movements and urine production were normal. Symptoms had been present for approximately 30 days. A febrile pharyngitis 1 week earlier was the only past medical history of remarkable note.

The treatment prescribed initially by the family physician, based on the diagnosis of acute rheumatic fever, consisted of administration of 300 mg of acetylsalicylic acid 3 times a day, which is not commonly recommended, and penicillin. The prescribed medical therapy was without benefit. Due to a worsening of the symptoms (mainly oral bleeding and knee pain on passive and active motions), she had then been referred to the Pediatric Rheumatology Unit for further evaluation. The patient presented no fever, and her hematological exams showed hypochromic microcytic anaemia and an increase in the erythrosedimentation rate and c-reactive protein. A bone marrow biopsy, performed in the Pediatric Rheumatology

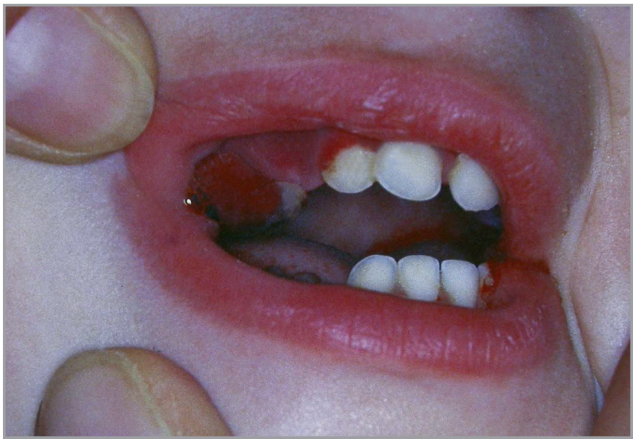

Figure 1. Baseline: gingival swelling with bleeding.

Drs. [1]Bacci and [2]Sivolella are consultants, [3]Dr. Pellegrini is resident, [4]Dr. Favero is full professor, and [5]Dr. Berengo is associate professor, all in the Department of Medical-Surgical Specialities, Unit of Oral Surgery, Institute of Clinical Dentistry, University of Padova, Padova, Italy.

Correspond with Dr. Bacci at christian.bacci@unipd.it

Unit, demonstrated no pathological changes; X rays of both knees showed no pathological findings. Articular ultrasonography revealed very mild synovial effusion. The Pediatric Rheumatology Unit initially presumed a diagnosis of pediatric rheumatoid arthritis.

Our differential diagnosis regarding the gingival swelling included: drug-influenced gingivitis; pyogenic granuloma; necrotizing ulcerative gingivitis; aphthous ulcers; gingival disease of specific bacterial or viral origin; Crohn's disease; Behcet's disease; dental abscess; multifocal osteomyelitis; multiform erythema (Stevens-Johnson disease); scurvy; Langerhans-cell histiocytosis; hematological cancer (leukemia-associated gingivitis); diabetes mellitus-associated gingivitis; and gingival manifestation of systemic conditions (hyperparathyroidism; leukocyte T-cell and combined antibody deficiency; phagocyte dysfunction; disorders affecting neutrophils; immunosuppressive medications; HIV disease; Langerhan's cell histiocytosis; hypophosphatasia; and Ehlers-Danlos syndrome).

Acetylsalicylic acid was the only medication taken; therefore pharmacologically induced gingival hypertrophy was excluded; multiform erythema was possible, but knees are usually not affected.

Dental abscess and infectious gingivitis are usually unilateral; instead, in this case, hypertrophy had spread to all quadrants. Hematological exams and the bone marrow biopsy made possible the exclusion of hematological cancer, Langerhans-cell histiocytosis, and endocrine disorders. The lack of lesions on the lips and other skin irritations made exclusion of multiform erythema possible.

A more in-depth medical history was then conducted, revealing that the child seemed to be more excited recently, and the parents less able to satisfy her. The child's behaviour was not considered to be related to any underlying developmental disorder and, therefore, no referral to psychiatry was considered. Her diet was limited to well-accepted foods, including milk, water, and biscuits every day for more than 1 month.

A nutrition problem was suspected, and the hypothesis of avitaminosis C, which explained all the symptoms, was formulated. Vitamin C was measured, revealing a value of 0.12 mg/dl (normal range=0.20-1.90 mg/dl), confirming the diagnosis of critical avitaminosis C or scurvy (also called Barlow's disease or hemorrhagic scurvy) (1). The oral picture was then defined as "ascorbic acid-deficiency gingivitis."

After informed consent was obtained from the parents, the child was immediately administered 250 mg of ascorbic acid twice a day orally.[7,8] The child's psychomotor restlessness decreased notably on the first day; by the third day, the child's gums were no longer painful and not bleeding on minimum contact (Figure 2). In 3 weeks, the patient was able to walk normally again, with her hemoglobin and inflammation markers normalized.

Figure 2. Resolution of swelling after a 3-day ascorbic acid administration.

Discussion

Ascorbic acid is needed for a variety of biosynthetic pathways; in particular, ascorbic acid is required as a cofactor for 2 enzymes responsible for the hydroxylation of the proline and lysine amino acids in collagen. Oral mucosa and skin petechiae, perifollicular hemorrhage, and ecchymosis are usually the first signs of scurvy, followed by edema and bleeding of the gums. Subperiosteal haemorrhages can lead to intense bone pain and the inability to move the legs actively or passively.[2,4,7,8]

Scurvy is by now a pathosis relegated almost exclusively to books on the history of medicine or present in rare cases in conditions of extreme malnutrition.[2,3]

Children's nutritional deficiencies can be influenced by many factors. Classically, a lack of family economic resources is considered the main factor.[5] The family environment, however, has to be considered not only from an economic perspective, but also from other points of view.[5,6] There are many factors that can contribute to this problem, including: capacity of establishing family rules; positive parental role modelling to encourage good eating habits; parental occupational status; the availability of certain kinds of foods (fresh fruits and vegetables); maternal education; intelligence and depression; cultural characteristics; maternal input into family economic decisions; and child characteristics (psychiatric and developmental pathologies).

Living in smaller communities,[1] religious rules,[4] and dietary restrictions for therapeutic purposes (eg, allergy, atopic dermatitis) could lead to improper nutrition habits. The family environment of the patient involved in this case report was characterized by an absence of economic problems. The nutrition deficiency was explained by the parents' inability to establish a suitable dietary regime for their child. The child was otherwise healthy and there were no economic concerns.

In this case report, oral manifestations, combined with a more accurate medical history, dramatically helped in the formulation of the correct diagnosis and the resolution of the pathology.

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