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Case Report / Olgu Sunumu

Known But Forgotten Disease: A Ten-year-old Scurvy Case

Bilinen Ama Unutulan Hastalık: On Yaşında Skorbüt Olgusu

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ABSTRACT

In this case report, the symptoms, diagnosis, and treatment process of a ten-year-old male patient with cerebral palsy diagnosed with scurvy due to vitamin C deficiency are presented. A 10-year-old male patient with a diagnosis of cerebral palsy and epilepsy was admitted with complaints of pain, swelling, and inability to step on the right ankle and left knee for 3 weeks. Swelling and edema were present in the left knee and right ankle. There were petechial lesions and ecchymosis on the legs. In addition, gingival bleeding and hypertrophy were observed. Laboratory examinations revealed anemia and acute phase reactant elevation. Non-steroidal anti-inflammatory therapy was initiated, but no regression was observed. Radiographs of the left knee and magnetic resonance imaging were performed, and dense lines were detected in the metaphyseal line. When the patient's dense lines finding, myalgia, petechiae, anemia that is unresponsive to iron treatment, gingival hypertrophy, and bleeding were evaluated, it was found to be compatible with scurvy disease. Despite the prevalence of easy access to food and healthy nutrition information, scurvy disease should be considered in patients with selectively fed autism spectrum disorder or neurodevelopmental retardation, as in our case.

Keywords: Child, dense line, metaphyseal irregularity, petechiae, vitamin C

ÖZ

Bu olgu sunumunda, C vitamini eksikliğine bağlı skorbüt hastalığı tanısı alan serebral palsili 10 yaşındaki erkek hastanın semptomları, tanı ve tedavi süreci sunulmaktadır. On yaşında serebral palsi ve epilepsi tanılı erkek hasta, üç haftadır sağ ayak bileği ve sol dizinde ağrı, şişlik, basamama şikayetleri ile başvurdu. Sol diz ve sağ ayak bileğinde şişlik ve ödem mevcuttu. Bacaklarda peteşiyal lezyonlar ve ekimoz mevcuttu. Ayrıca dişeti kanaması ve hipertrofi gözlendi. Laboratuvar incelemelerinde anemi ve akut faz reaktan yüksekliği saptandı. Non-steroidal anti-enflamatuvar tedavi başlandı ancak gerileme olmadı. Sol diz grafileri ve manyetik rezonans görüntüleme yapıldı ve metafiz hattında yoğun çizgiler tespit edildi. Hastanın yoğun çizgi bulgusu, miyalji, peteşiler, demir tedavisine yanıt vermeyen anemi, dişeti hipertrofisi ve kanaması değerlendirildiğinde skorbüt hastalığı ile uyumlu bulundu. Gıdaya kolay erişim ve sağlıklı beslenme bilgilerinin yaygınlığına rağmen, olgumuzda olduğu gibi seçici beslenen otizm spektrum bozukluğu veya nörogelişimsel geriliği olan hastalarda bu semptomların varlığında skorbüt hastalığı düşünülmelidir.

Anahtar Kelimeler: Çocuk, yoğun çizgi, metafiz düzensizliği, peteşi, C vitamini

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INTRODUCTION

Vitamin C is a powerful antioxidant involved in fatty acid transport, collagen and neurotransmitter synthesis, prostaglandin metabolism, and nitric oxide synthesis. Its need is met by the daily intake of foods.[1]

Vitamin C deficiency is most commonly observed because of a lack of intake.[2,3] In children with malabsorption, autism, and similar spectrum disorders, neurodevelopmental retardation and food selection can be observed because of difficulties in solid food intake.[1,2]

Vitamin C is an effective cofactor in collagen synthesis, and its deficiency causes collagen-related symptoms. In insufficient intake, symptoms begin after 3 months, and petechiae, gingivitis, arthralgia, edema, and anemia are observed most frequently.[1,3] In addition, characteristic findings on magnetic resonance imaging and radiography, with sclerotic and lucent metaphyseal bands, guide the diagnosis.[4]

Although the occurrence of scurvy is rare, it remains essential to detect scurvy in children selectively fed. In this case report, a ten-year-old male patient diagnosed with cerebral palsy diagnosed with scurvy due to vitamin C deficiency because of insufficient intake is presented to draw attention to this issue.

CASE REPORT

A ten-year-old male patient with a diagnosis of cerebral palsy (not autism spectrum disorder in addition to cerebral palsy) and epilepsy was admitted with complaints of pain, swelling, and inability to step on the right ankle and left knee for 3 weeks. It was learned that he applied to the orthopedics department, and the synovial fluid samples were determined and it was not compatible with septic arthritis. Ecchymosis was observed in both ankles and the left leg churis area (Figure 1).

There was a difference in diameter in the joints and legs (Table 1). Gingival bleeding and hypertrophy were observed (Figure 1). It was learned that he had applied to multiple dentists because of bleeding 3 weeks ago.

In laboratory tests, hemoglobin was 7 g/dL, C-reactive protein was 49.3 mg/L, sedimentation rate was 79 mm/h, and peripheral smear showed clustered platelets and no signs of hemolysis (Table 1). When it was learned that the patient had been followed up with iron deficiency for about a year, 5 mg/kg/day iron treatment was started. White cells, platelets, and bleeding profiles were normal. Epstein-Barr virus, TORCH, and brucella were negative, and no growth was observed in blood/urine cultures. Antinuclear antibody and p/c-Anca values were negative,

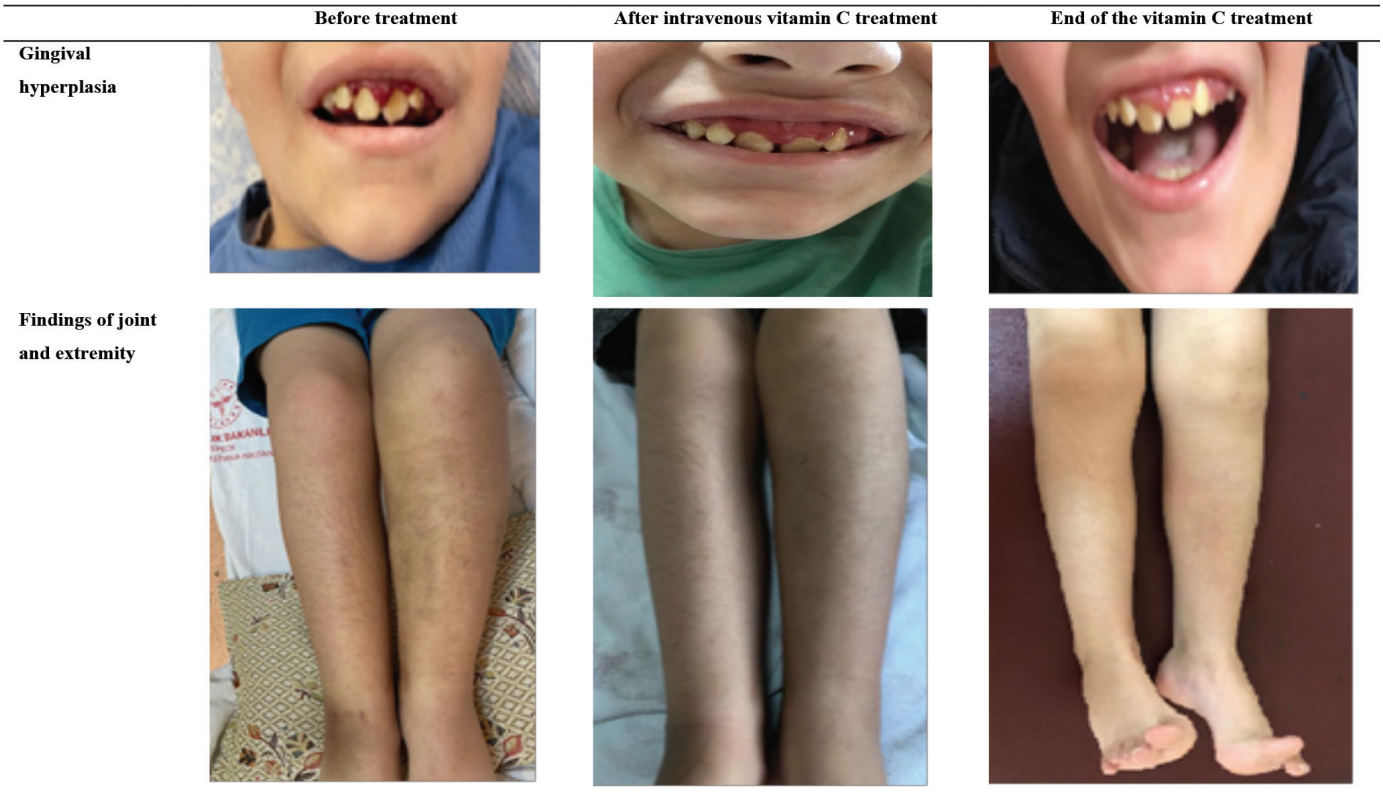

Figure 1. Clinical findings of the case before and during treatment

and creatine kinase, rheumatoid factor, and complements were within normal limits. No uveitis was detected.

For the symptoms in the joints that persisted for more than 6 weeks, nonsteroidal anti-inflammatory therapy was started for juvenile idiopathic arthritis (JIA), but no regression was observed. Bone marrow aspiration was performed for malignancy due to cytopenia, rash, and joint swelling, which was found to be normal. Radiographs of the left knee and magnetic resonance imaging of the left knee were performed, and dense lines were observed in the metaphyseal area (Figure 2). Diseases with dense line signs were evaluated in the differential diagnosis.

When the patient's dense lines finding, myalgia, petechiae, anemia unresponsive to iron treatment, gingival

Table 1. Evaluation of the physical examination and hemoglobin value of the patient during the treatment process

	Before treatment	After intravenous vitamin C treatment	End of the vitamin C treatment
Left knee circumference	38 cm	37 cm	31 cm
Right knee circumference	34 cm	34 cm	31 cm
Left leg circumference	40 cm	38 cm	32 cm
Right leg circumference	34 cm	33 cm	32 cm
Left ankle circumference	25 cm	26 cm	24 cm
Right ankle circumference	26 cm	25 cm	24 cm
Hemoglobin value	7 gr/dL	8.8 gr/dL	11.9 gr/dL

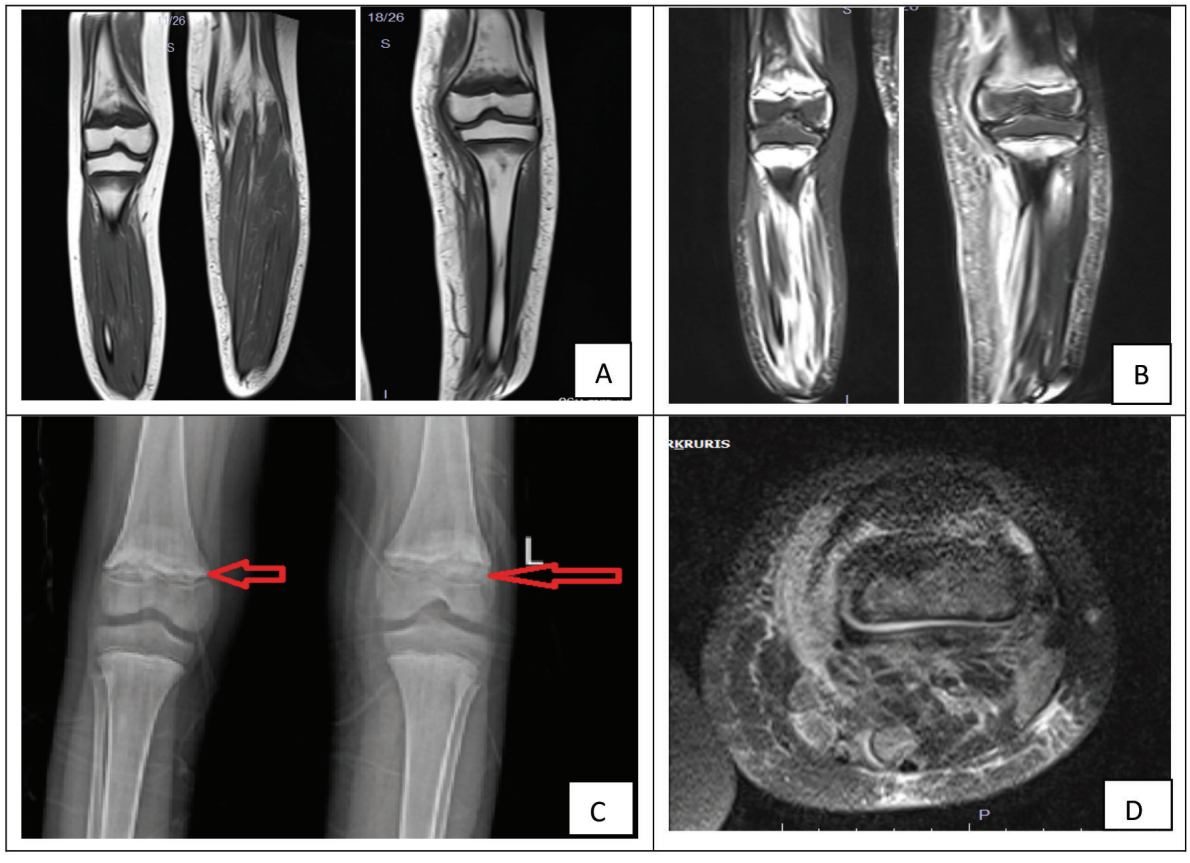

Figure 2. Extremity direct X-ray and magnetic resonance imaging of the case (A: T1 Coronal: hypointense areas adjacent to the metaphysis, without signal. B: T2 coronal: Metaphyseal dense line, edema, inflammation. C: Radiopacity increase in the metaphyseal line, linear dense matphysis line. D: Fat-suppressed axial section: signal increase compatible with intense inflammation in the muscle, cellulite in subcutaneous fatty tissue, lymphedema, increase in soft tissue thickness, linear fluid echoes between fat lobules)

hypertrophy, and bleeding were evaluated, it was found to be compatible with the scurvy disease. C vitamin plasma level was detected <0.1 (N=0.5-1.8) mg/dL. When the nutrition of the child was examined, it was learned that the patient had chosen only eggs, cheese, and bread for the last two years because of his rejection of food variety. C vitamin (100 mg/day) intravenous treatment was given for 1 week, and after clinical recovery in 1 week, for 3 months oral C vitamin (100 mg/day) treatment was given. A diet that provided vitamin C treatment and age-appropriate calorie and nutrient distribution was regulated.

At follow-up, the joint, gingival, and hemoglobin values improved, and the patient was discharged.

DISCUSSION

Vitamin C deficiency is a disease with a decreasing frequency in children, especially in cases of insufficient intake. Neuropsychiatric diseases are the most common causes of insufficient intake. Similarly, our case was a case with a diagnosis of cerebral palsy and food selective. In these cases, the type of nutrition is the preference of the case or the parent due to food preference or intestinal dysmotility.[4,5]

If there is no intake, symptoms begin after 8-12 weeks. The first symptoms are irritability, loss of appetite, and low-grade fever.[2,6] At approximately the fifth month, purpura, petechiae, ecchymosis, and hyperkeratosis begin, and these symptoms are similar to vasculitis.[6,7] Arthralgia, myalgia, limping, soft-tissue edema, and bone marrow suppression ranging from anemia to pancytopenia may mimic malignancies and rheumatologic diseases.[7-9] The reason for our patient's presentation was joint swelling and pain, suggestive of septic arthritis, but negative acute phase reactants and no growth in the joint fluid sample excluded the diagnosis.[10] The spread of a single joint finding to other joints in the follow-up and the addition of lower extremity soft tissue swelling suggested rheumatological diseases, especially JIA.[11] In the follow-up of the patient, other symptoms and findings excluded the diagnosis. In addition to the finding of anemia requiring transfusion and petechiae-ecchymosis, the patient's white blood cell count, platelet count, aggregation, and bleeding profile were normal, suggesting that there was no bone marrow suppression.

The diagnosis of scurvy disease starts after 3-5 months with radiological findings. Long bone radiographs show metaphyseal irregularity and dense lines.[2,3,12,13] X-ray findings may play a primary role in diagnosis, as in this study. The other diseases were ruled out by history, physical examination, laboratory, and imaging methods, and when the clinic of the case was considered as a whole, the only disease that would explain the dense line finding was evaluated as a scurvy disease.[4]

Symptoms of scurvy usually occur when the plasma concentration of ascorbic acid is 0.2 mg/dL (11 micromol/L). Plasma vitamin C levels are included in the diagnosis by clinical and physical examination.[14] Treatment of scurvy begins with the administration of vitamin C deficiency intravenously, intramuscularly, or orally. During the treatment process, the cause of the deficiency should be determined. In cases developing due to intake deficiency, the patient's diet should be arranged as a balanced diet with sufficient vitamin, protein, fat, and carbohydrate content.[7] In our patient, vitamin C levels were determined, and first intravenous and then orally vitamin C treatment was administered. Nutrition was prepared with the dietitian, and appropriate vitamin levels were adjusted.

CONCLUSION

As a result, scurvy was suspected in the case with insufficient intake, physical examination findings, and imaging methods, and a diagnosis of scurvy was made on the basis of vitamin C level. Despite easy access to food and the prevalence of healthy nutrition information, scurvy disease should be considered in patients with selectively fed autism spectrum disorder or neurodevelopmental retardation, as in our case.

Ethics

Informed Consent: Consent form was filled out by all participants.

Authorship Contributions

Surgical and Medical Practices: Ö.Ü., B.K-D., A.K., Concept: Ö.Ü., F.C.S., B.K-D., A.K., Design: Ö.Ü., İ.T.Y., F.C.S., G.İ., B.K-D., A.K., Data Collection or Processing: Ö.Ü., C.T., Analysis or Interpretation: Ö.Ü., İ.T.Y., F.C.S., B.K-D., Literature Search: Ö.Ü., C.T., G.İ., Writing: Ö.Ü., B.K-D., A.K.

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