

MR Imaging in a Child with Scurvy: a Case Report

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Scurvy is very rare disease in industrialized societies. Nevertheless, it still exists in higher risk groups including economically disadvantaged populations with poor nutrition, such as the elderly and chronic alcoholics. The incidence of scurvy in the pediatric population is very low. This study reports a case of scurvy in a 5-year-old girl with cerebral palsy and developmental delay based on MRI findings.

Scurvy results from a deficiency of ascorbic acid (vitamin C). A marked reduction in the occurrence of scurvy has occurred over the last 100 years due to improved knowledge about the pathophysiology and treatment of scurvy. However, the disease still exists even in industrialized countries (1, 2). Because scurvy is rare in modern society, reported magnetic resonance imaging (MRI) findings associated with scurvy are very rare (3). We report a case of scurvy in a 5-year-old girl with associated MRI findings.

Index terms :

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CASE REPORT

A 5-year-old girl was presented with swelling of the left thigh, general weakness, poor oral intake, and a mild fever for two weeks. There was no evidence of petechiae, bruising, or history of trauma. The patient was diagnosed with cerebral palsy, severe developmental delay, generalized tonic clonic seizures, and thus was treated with long-term anticonvulsant medication. The patient had a history of poor oral intake and vomiting due to swallowing difficulty for more than 1 month.

Upon physical examination, the patient's weight was below the third percentile for her age group. Moreover, the patient's left thigh was swollen and warm without erythema. No other remarkable findings were found. The laboratory data results were as follows: white blood cell count, 17,500/ μ L, neutrophil count, 79%, lymphocyte count, 14%, and platelet count, 528,000/ μ L. Furthermore, the erythrocyte sedimentation rate was 11 mm/hr, the C-reactive protein level was 6.05 mg/dL, and the hemoglobin (9.2 g/dL) was within the normal range.

The radiographs of both knees showed osteopenia, which is a thick sclerotic metaphyseal line above a widened physis, and small beak-like excrescences at the metaphysis of both femora. In addition, a disruption of the alignment of the distal physis of the left femur was also noted (Fig. 1).

An MRI of the left thigh performed on the first hospital day revealed diffuse bone marrow signal changes in the femoral shaft with large subperiosteal fluid collection and displacement of the distal epiphysis. The marrow changes appeared as heterogeneous high and low signal intensities on T1-weighted images and heterogeneous high

and intermediate signal intensity on T2-weighted images. The subperiosteal fluid collection had low signal intensity on T1-weighted images and high signal intensity at fluid-fluid levels on T2-weighted images. The soft tissue of the left thigh, including the vastus and hamstring muscles, showed high signal intensity lesions on the T2-weighted images. The periosteum and surrounding muscles of the thigh were moderately enhanced on contrast enhanced fat suppressed T1-weighted images (Fig. 2). A bone scintigraphy performed on the seventh hospital day showed no definite hot uptake in the left femur and a decreased uptake in the epiphyseal plate of the left distal femur. We

began antibiotic therapy for the impression of osteomyelitis with subperiosteal abscess.

An operation was performed on the third hospital day to irrigate and drain the periosteal fluid, which was collecting in the left femur. A relatively healthy color and consistency was noted in the vastus lateralis muscle along the incision tract. The dark serous hematoma was drained from the periosteal incision site and the pathologic findings revealed an osteonecrosis of the cortical bone and a mild chronic inflammation with hemorrhage of the periosteum.

On the 14th hospital day, there was newly developed swelling of the right thigh and recurrent swelling of the left

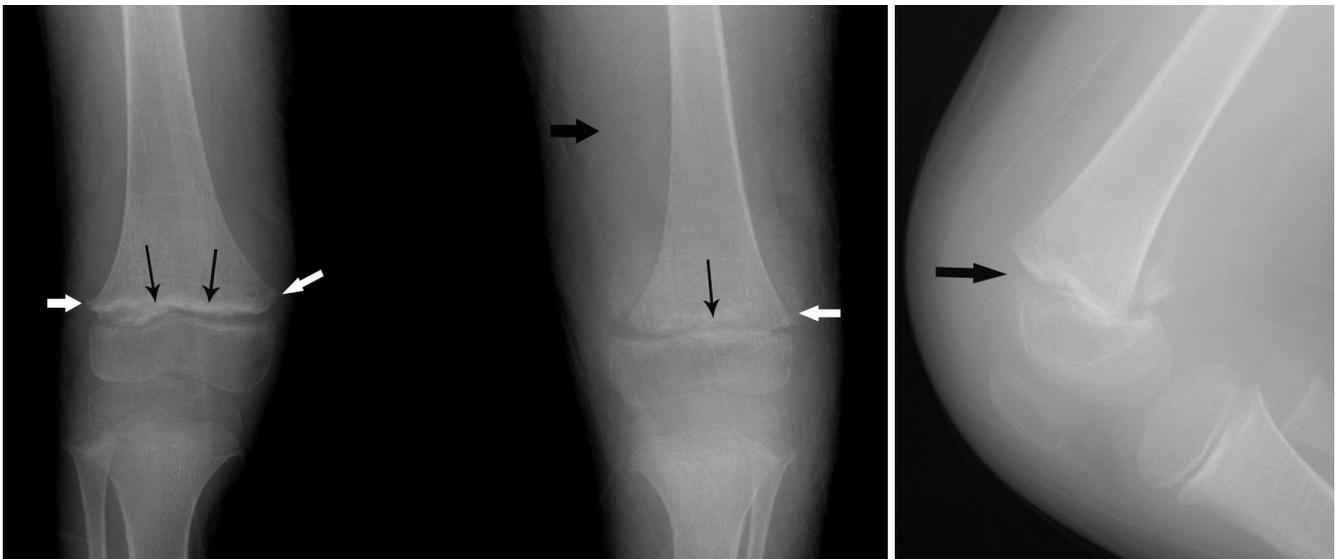


Fig. 1. A. Anteroposterior radiograph of both knees shows a thick sclerotic metaphyseal line (thin black arrows) above a widened physis and small beaklike excrescences (white arrows) at the metaphysis in both femora. Soft tissue bulging is noted (thick black arrow). B. Lateral radiograph of the left knee shows a disruption of the alignment of the distal femoral physis (arrow).

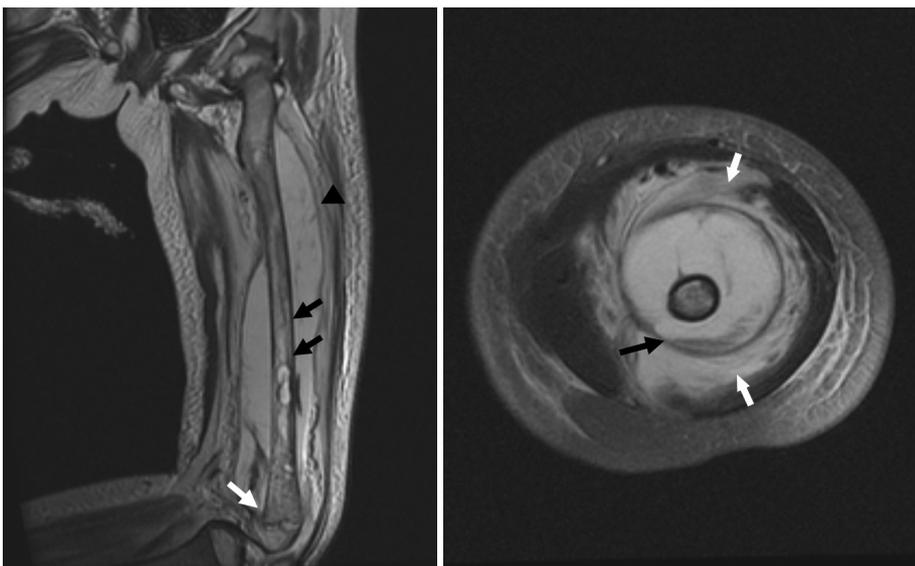


Fig. 2. MRI of the left thigh performed at the first hospital day. A. Coronal T2-weighted image shows a diffuse bone marrow signal change (black arrows) of the femur shaft with a large amount of subperiosteal fluid collection (arrowhead) and displacement of the distal epiphysis (white arrow). B. Axial T2-weighted image shows a fluid-fluid level in the subperiosteal fluid collection (black arrow). The surrounding vastus and hamstring muscles also show high intensity signal lesions (white arrows).

A B

thigh with fever. A follow-up MRI performed 16 days after the initial MRI, showed the reappearance of massive diaphyseal subperiosteal hematoma in the left thigh with similar MRI findings in the right thigh (Fig. 3). The lesions appeared to have progressed or aggravated without response to antibiotic therapy.

Fluid drainage and wire fixation of the epiphyseal disruption were performed on the 21st hospital day.

The laboratory test results obtained on the 22nd hospital, revealed that the Vitamin C level was 0.06 mg/dL (reference range: 0.6–2 mg/dL) and the vitaminic D3 (1-alpha, 25 [OH]2) level was 46 pg/ml (reference range: 20.0-60.0 pg/ml). After the immediate addition of vitamin C to the diet, the general condition of the patient improved. Moreover, the serum vitamin C level was normalized (0.98 mg/dL) at two weeks after the administration of vitamin C and the other laboratory findings also improved.

A follow-up radiograph at six weeks after vitamin C supplementation therapy showed prominent diaphyseal periosteal calcification (Fig. 4) as well as a slight improvement in the metaphyseal abnormalities seen on the initial radiographs.



Fig. 4. The radiograph performed six weeks after vitamin C supplementation. Large shells of periosteal bone (arrows) are present at the left femur. There was improvement of the metaphyseal sclerotic line and widened physis compared to the initial radiograph.

DISCUSSION

Although the incidence of scurvy is extremely rare in industrialized countries (4), it is still present in economi-

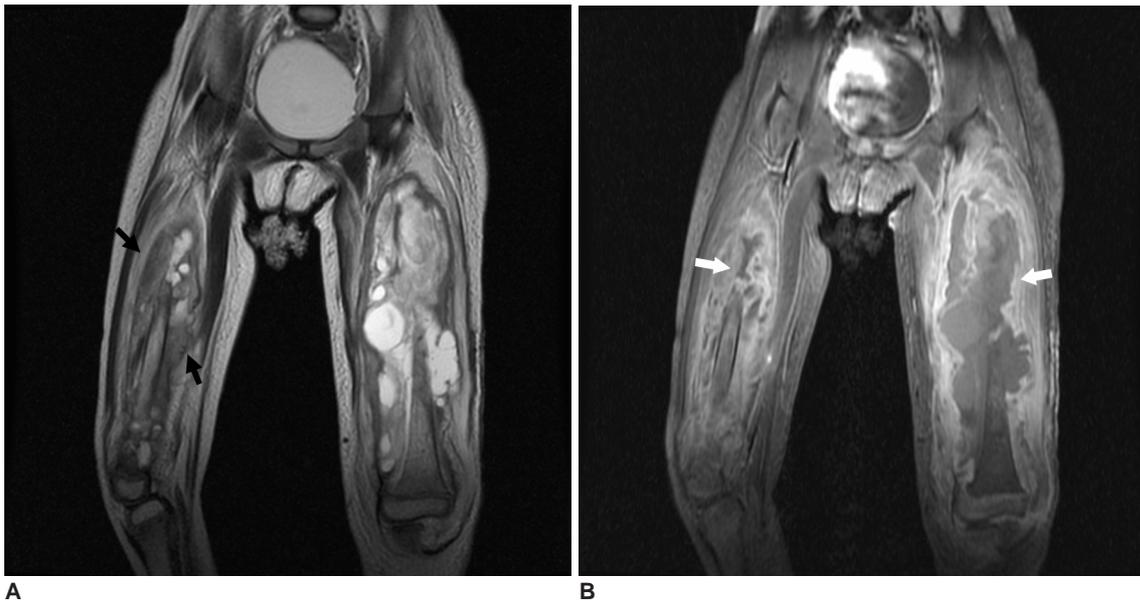


Fig. 3. Follow up MR images of both thighs. **A.** A coronal T2-weighted image shows a larger subperiosteal hematoma at the left femur and a new subperiosteal hematoma (arrows) with signal change of bone marrow of the right femur. **B.** A coronal contrast enhanced fat suppression T1-weighted image shows moderate enhancement of the periosteum and adjacent soft tissue (white arrows).

cally disadvantaged populations with poor nutrition including elderly persons living alone and alcoholics (5). Scurvy has historically been less frequent in the pediatric population. However, infants who are fed evaporated or boiled milk, in which ascorbic acid is easily destroyed by heat as well as children with poor diet as a result of psychiatric or developmental disorders, are at risk (3). In our patient, scurvy resulted from poor oral intake during one month due to a difficulty in swallowing. We also suspected the possibility of rickets due to the patient's poor oral intake, however the patient's serum vitamin D level was shown to be normal.

Musculoskeletal manifestations are present in 80% of patients with scurvy (2). Moreover, bone disease is a more frequent manifestation of the condition in children than adults, as is in our patient. The radiographic findings of pediatric or infantile scurvy are as follows: a transverse metaphyseal line of increased density, a transverse metaphyseal line of decreased density (scurvy line), metaphyseal excrescences of the beaks, subepiphyseal infractions, increased density of periostitis and epiphyseal shell with a central lucency (Wimberger's sign of scurvy). The scurvy line reflects the decrease in trabeculae and detritus in the junctional area of the metaphysis. Moreover, the Wimberger's sign is a prominent thickened provisional zone of calcification with atrophy of the central spongiosa on pathology (6). Furthermore, the radiographic findings including osteopenia, thick sclerotic metaphyseal line, metaphyseal excrescences of beaks, subepiphyseal infraction and periostitis were observed on the radiograph of our patient; however the scurvy line was not prominent. The resolution of the metaphyseal abnormalities after vitamin C supplementation was also consistent with radiographic findings of the healing stage of scurvy. The large shells of periosteal bone are common radiographic findings, particularly during the healing phase of disease (6), which seem to result from periostitis as a result of a subperiosteal hematoma.

Because of the rarity and a lack of understanding of the MRI findings of scurvy, the laboratory findings suggested an inflammatory condition and we initially could not suspect the possibility of scurvy. Therefore, the subsequent antibiotic therapy and operation for subperiosteal fluid drainage were performed under the impression of osteomyelitis and a subperiosteal abscess. Due to the unresponsiveness and further progression of the disease despite antibiotic therapy, we then suspected the possibility of scurvy or another metabolic disease.

Due to the radiologic findings the meta-epiphyseal fracture and subperiosteal hematoma, another possible diagnosis could have been 'battered child syndrome'.

Fortunately, our patient's symptoms subsided after vitamin C supplementation; however, an entire skeletal survey, a check up of the patient's parental psychiatric status or anger about their child's chronic illness were necessary in the early stages of the patient's evaluation.

The bone scintigraphy showed no definite hot uptake in the left femur. Hence, we believe that the decreased radionuclide uptake in the physis of the left distal femur resulted from the epiphyseal separation of the left distal femur.

The initial MRI showed heterogeneous signal intensities along nearly the entire femoral shaft on both T1- and T2-weighted images, and a large collection of subperiosteal fluid with rim enhancement and surrounding soft tissue edema with enhancement. These MRI findings were somewhat nonspecific and may have suggested other more common conditions such as osteomyelitis, subperiosteal abscess, or leukemia. The follow-up MRI showed a much more notable increase in the amount of subperiosteal hematoma in both femoral shafts. The recurrent subperiosteal hematoma was an important clue for the diagnosis of scurvy. The MRI clearly revealed a disruption of the epiphyseal line; however, metaphyseal changes including sclerosis and typical radiographic findings in scurvy cases were not detectable, which probably resulted from poor conspicuity of sclerosis on MRI and the large field of view, which included the entire thigh. The marrow signal intensity of the femoral shaft in this case seems likely to represent edema and hemorrhaging in the marrow cavity. Due to the rarity of scurvy, the MRI findings are not well known and additional cases are needed to establish MRI findings.

A low vitamin C level in the plasma is specific for the diagnosis of scurvy; however, this is not always a reliable indicator because plasma levels may be normal with recent intake of ascorbic acid. Measuring vitamin C levels in the Buffy-coat of leukocytes better reflects the body stores; however, this method is technically more difficult. The best evidence for scurvy is the resolution of the manifestations of the disease after treatment with ascorbic acid. The dose and duration of treatment is patient specific (3).

Because of the extremely rare occurrence of scurvy in modern society, it is difficult to differentiate it from other diseases. The diagnosis of scurvy is made by clinical and radiographic findings and may be supported by additional findings such as reduced levels of vitamin C in the serum or Buffy-coat of leukocytes. The MRI findings of scurvy are not well known; however, when the MRI findings include subperiosteal hematoma with periostitis, metaphyseal changes, and heterogeneous bone marrow signal intensity, scurvy should be included in the differential

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diagnosis. Clinicians and radiologists must be aware of this extremely rare but still present condition, because it is potentially fatal and easily cured with vitamin C supplementation.

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