Scurvy in A Malnourished Child: Atypical Imaging Findings

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ABSTRACT

Scurvy, a disease caused by a severe lack of vitamin C in the diet, is most often associated with 17th-century sailors. Its 21st-century manifestation is a disease of the poor, sick, and those living in remote rural neighborhoods in which fresh, nutritious food is hard to come by. It is caused by a deficiency of Vitamin C and is rare in the United States. We describe the radiographic and MRI findings of a case of scurvy in a child with Noonan syndrome who is a "picky eater". MRI is rarely performed in scurvy as its radiographic findings are generally well known and sufficient for a diagnosis. However, due to the rarity of the disease in the US, MRI features of scurvy have been described in only a few case reports, to date. The rarity of this disease also causes scurvy to be kept lower, if at all in the differential diagnosis list.

CASE REPORT

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A 6-year-old boy with Noonan syndrome and short stature, presented with 3 weeks of pain in both lower extremities, particularly in the hip and knee joints. He had a G-tube for feeding but was not using it. He was known to be a "picky eater". He had learning disabilities and intellectual disabilities. No visual or hearing defects were present. Due to the pain in the legs, the child had a limping gait. Initial concern in the Emergency Room was trauma (with the possibility of non-accidental trauma) or leukemia. Clinical examination did not show any bruises or bleeds. No swelling of the legs or rash was seen. No history of bleeding from gums was provided by the mother of the child. Scurvy was not on the list of differential diagnoses. Radiographs of the hip and knee were performed which showed metaphyseal lucent bands at the bilateral hips and knees. Generalized reduced bone density was also seen. No fractures or soft tissue swelling was seen. Leukemia and metastatic neuroblastoma were kept high on the differential diagnosis. Due to the absence of fractures trauma was ruled out. MRI of the knees was then performed for further evaluation. It showed metaphyseal hypointensities on T1W images and hyperintensities on STIR images. No periosseous edema was seen. No changes of myositis were seen. A complete blood count was performed, which came back normal. At this point, scurvy was considered. The classical features of scurvy are generalized osteopenia, cortical thinning, periosteal reaction due to subperiosteal hemorrhage, hemarthrosis, circular opaque shadow around epiphyseal ossification center (wimberger sign), dense epiphyseal calcification (Frankel line), Lucent metaphyseal band (trummerfeld zone), metaphyseal spurs (pelkin spur) resulting in cupping of the metaphysis, and metaphyseal corner fractures. In this case, only metaphyseal lucent bands were present and thus there was a delay in diagnosis. Ascorbic acid levels were determined to be low (<0.1 mg/dl). The normal range of Ascorbic acid is 0.6 to 2 mg/dL.

The child's symptoms gradually resolved after 2 weeks, of treatment with Vitamin C. Further multidisciplinary care is continuing for his nutritional, physical, and behavioral needs.

DISCUSSION

Etiology & Demographics:

Vitamin C is required for collagen formation, bone matrix, tendon, and ligament synthesis [1,3]. Scurvy is mostly seen in countries with low socioeconomic status and is rare in the developed world. Alcoholics and elderly individuals living alone have a higher incidence of developing scurvy. Malnourishment due to pre-existing conditions can lead to vitamin C deficiency and scurvy. It is becoming increasingly common in low-income households and rural remote America. According to a Centers for Disease Control and Prevention study published in 2009, 10 to 17 percent of low-income people suffer from scurvy-level vitamin C deficiency. It can cause fatigue and neurological symptoms. It needs to be recognized to be treated. Although easy to diagnose clinically and radiographically if classical features are present, it is not primarily suspected in developed nations.

Clinical & Imaging findings:

Scurvy results from inadequate intake of vitamin C (ascorbic acid)—a nutrient abundant in citrus fruits [4]. Scurvy can cause fatigue and neurological symptoms. It needs to be recognized to be treated. Although easy to diagnose clinically and radiographically if classical features are present, it is not primarily suspected in developed nations.

Plain radiographs classically show diffuse demineralization and subperiosteal hemorrhage [5,7]. A sclerotic epiphyseal rim (Wimberger ring), dense provisional metaphyseal calcification (Frankel line), and metaphyseal spurs leading to cupping of the metaphysis (Pelkan spurs) are classically described. However, these signs might not be present in every patient and thus the diagnosis can be missed unless suspicion is kept high.

The MRI findings of scurvy have been described as altered marrow signal, subperiosteal fluid collections, and periosteal reaction however, in this particular case, no periosteal reaction or subperiosteal fluid collections were seen. These findings on MRI are nonspecific, the differential for which would include high morbidity conditions, such as hematological malignancies, osteomyelitis, or metastatic disease. As a result, such findings on MRI may lead to additional invasive diagnostic procedures or scintigraphic imaging [6]. Gelatinous transformation is a sign of severe malnutrition characterized by the replacement of fat and hematopoietic cells in bone marrow with hyaluronic acid. It is seen in cachexia and can be seen in scurvy. As hyaluronic acid is a mucopolysaccharide made of residues of monosaccharides containing hydroxyl groups, it easily forms hydrogen bonds with water molecules, hence the T2-prolongation on MRI. Radiologists should increase the field of view when abnormal bone marrow signal is noted around a joint. This permits evaluation for symmetry and the diffuse or multifocal nature of the disease.

Knowledge about the normal pattern of red marrow regression and resurgence is important for the assessment of the bone marrow. The epiphysis converts from red to yellow marrow within six months of age. Then marrow conversion initiates in the diaphysis and progresses to the metaphysis, faster distally than proximally. Generally, residual red marrow is only found within the proximal metaphysis of the humerus and femur till early adulthood. These physiological changes need to be known by the radiologist to prevent misdiagnosis.

Treatment & Prognosis:

Scurvy is treated by simply adding Vitamin C to the diet. Lemons, oranges, and other citrus fruits are high in vitamin C. Prognosis is very good with most patients improving in 48 hours and complete recovery occurs in 2 weeks.

Differential Diagnoses:

Leukemia is the first differential. However, in Leukemia blood counts will be abnormal. Also, the clinical presentation will be of an anemic child. This child had no such symptoms or lab reports.

Metastatic neuroblastoma is the second differential. However, for metastatic neuroblastoma, primary neuroblastoma will be present. In this child, no primaries were detected on body imaging.

Conclusion:

As we increasingly see complex cases with multiple comorbidities, Scurvy, a disease of yesteryear is making a comeback [2,8]. It must be considered particularly in the undernourished. It needs to be diagnosed early and treated. Biochemistry-proven scurvy might present as nonclassical findings on radiographs and MRI. This makes this case important so that radiologists keep this in mind while seeing a similar case in the future.

TEACHING POINT

Scurvy can occur even in the developed world due to vitamin C deficiency among the malnourished and chronically ill. A sclerotic epiphyseal rim (Wimberger ring), dense provisional metaphyseal calcification (Frankel line), and metaphyseal spurs leading to cupping of the metaphysis (Pelkan spurs) are the classic radiological findings.

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FIGURES

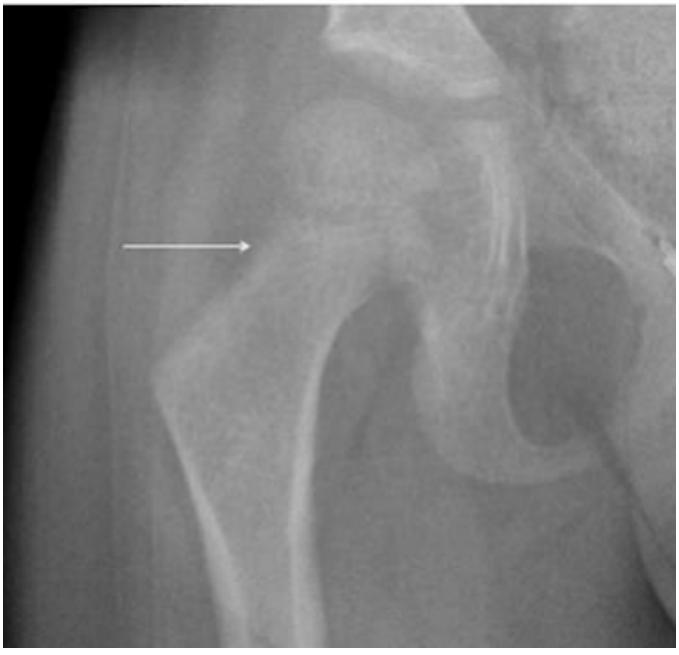

Figure 1: 6-year-old male with Scurvy.

Findings: An arrow points to the metaphyseal dense band in the proximal femur. It appears radiodense in the metaphysis of the proximal femur. A generalized decrease in bone mineral density is also seen. No fractures are seen. Joint spaces are normal. Visualized soft tissues are normal. Similar findings were seen in the opposite hip (image not included).

Technique: Right Hip Frontal Radiograph.

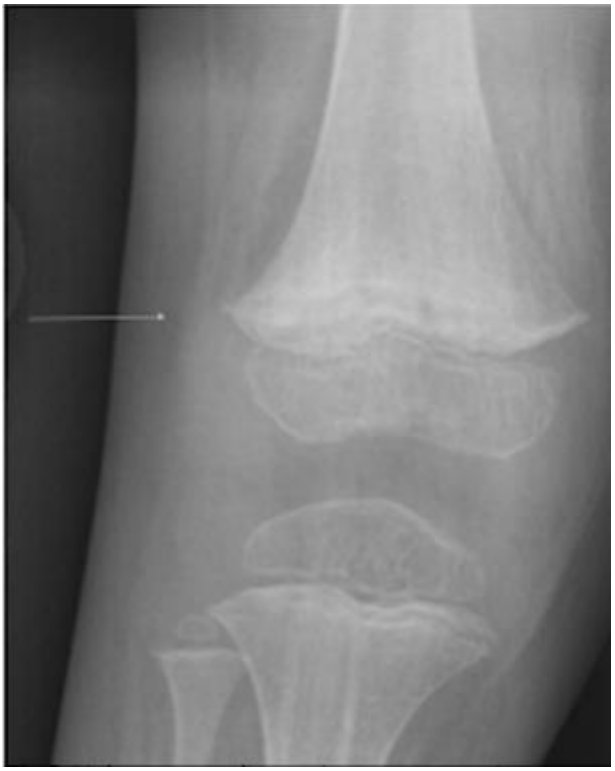

Figure 2: 6-year-old male with Scurvy.

Findings: Arrow points to metaphyseal lucent bands. It appears radiodense in the metaphysis of the distal femur. A generalized decrease in bone mineral density is also seen. No fractures are seen. Joint spaces are normal. Visualized soft tissues are normal. Similar findings were seen in the left knee.

Technique: Right Knee Frontal Radiograph.

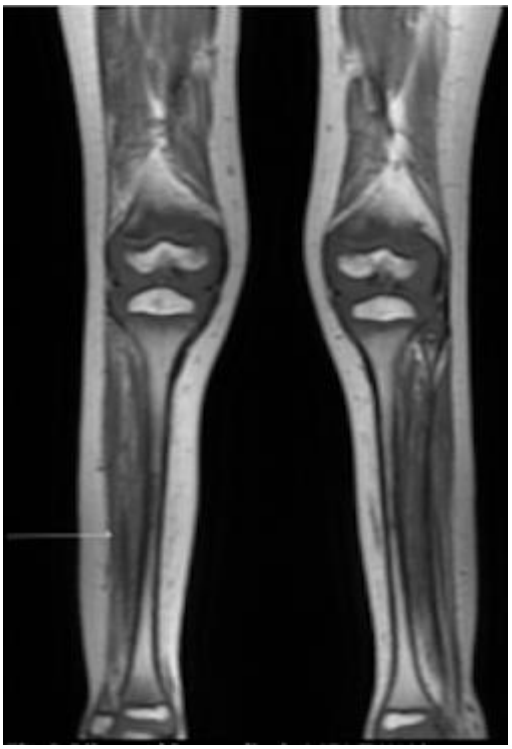

Figure 3: 6-year-old male with Scurvy.

Findings: Arrow points to metaphyseal hypointensities in the tibia and femur. The rest of the visualized bones are normal. Joint spaces are normal. Soft tissues are normal.

Technique: Bilateral lower limb MRI 1.5 Tesla magnet, coronal T1W images.

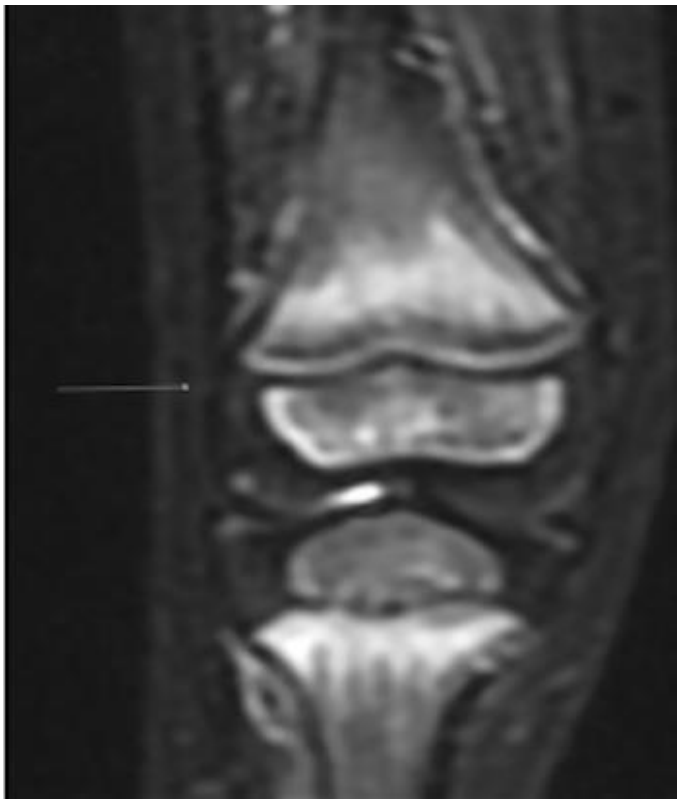

Figure 4: 6-year-old male with Scurvy.

Findings: Arrow points to hyperintensities seen within the femoral and tibial metaphyses corresponding to the metaphyseal bands on the radiograph. The rest of the visualized bones are normal. Joint spaces are normal. Soft tissues are normal. Similar findings were seen in the left knee (image not included).

Technique: Right knee MRI 1.5 Tesla magnet, coronal STIR images.

Etiology	Vitamin C deficiency
Incidence	10 to 17 percent of low-income people
Gender ratio	No gender predilection.
Age predilection	None.
Risk factors	Low income, malnourishment, chronic illness.
Treatment	Oral vitamin C, Citrus fruits.
Prognosis	Good
Findings on imaging	Sclerotic epiphyseal rim (Wimberger ring), dense provisional metaphyseal calcification (Frankel line), and metaphyseal spurs leading to cupping of the metaphysis (Pelkan spurs) on radiographs.

Table 1: Summary table of Scurvy.

	RADIOGRAPH	MRI
SCURVY	• Diffuse demineralization. • Subperiosteal Hemorrhage. • Sclerotic Epiphyseal rim (Wimberger ring) • Dense metaphyseal calcification (Frankel line) • Cupping of the metaphysis (Pelkan spur)	• Subperiosteal hematoma. • Periostitis. • Metaphyseal changes.
LEUKEMIA	• Osteopenia • Alternating radiolucent and radiodense metaphyseal bands. • Periosteal reaction. • Coarse trabeculation. • Bony lytic lesions.	• Diffuse low signal on T1W images. • Diffuse high signal on T2W images. • Post-contrast diffuse enhancement. • Intramuscular leukemic infiltrates.
METASTATIC NEUROBLASTOMA	• Ill-defined lucent with periosteal reaction.	• Heterogeneous on T1W images. • Hyperintense with cystic necrotic areas on T2W images. Signal voids may be present. • Post contrast variable and heterogeneous enhancement.
FRACTURE	• Discontinuity of bones. • Fracture fragments.	• Marrow edema. • Soft tissue inflammation.

Table 2: Differential diagnosis table for Scurvy.

ABBREVIATIONS

MRI = Magnetic Resonance Imaging
US = United States

KEYWORDS

Scurvy; Knee; Hip; Noonan syndrome; MRI; radiographs; malnourished; sick

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