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Case Report

A case report of a childhood scurvy musculoskeletal manifestation: Radiologic findings and diagnostic implications ☆☆☆

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ABSTRACT

Scurvy is an infrequent pathological condition resulting from a sustained dietary vitamin C deficiency. Radiology becomes pivotal because the diagnostic process for scurvy can be intricate, given its resemblance to bone neoplasms. A 6-year-old boy, reported persistent pain and swelling in the right thigh for 2 months prior to hospitalization. Clinical examination revealed a mass localized in the right thigh and anemia. A radiograph of the right femur demonstrated extensive osteopenic changes, “Trümmerfeld zone”, “Frankel line”, “Pelkin fracture”, “Wimberger ring sign”, and para-epiphyseal subperiosteal hematoma. The absence of any such cases in our institution over the preceding decade emphasizes the uniqueness of this presentation. Histopathological evaluation yielded atypical results, prompting further radiographic assessment of the left femur and thorax. The subsequent findings corroborated the classic “scorbutic rosary” presentation, indicative of scurvy. The patient’s symptoms gradually resolved with high-dose supplementation of vitamin C. Scurvy predominantly presents with musculoskeletal manifestations. Plasma vitamin C level assessment is the gold standard for the diagnosis, but it is currently inaccessible in our nation. Consequently, radiographic evaluation reveals pathognomonic features of the disorder. In thoracic radiographs, the “scorbutic rosary” presentation is evident. In contrast, long bones exhibit hallmarks of scurvy: diffuse osteopenia, “Frankel line”, “Trümmerfeld zone”, “Pelkin fracture”, “Wimberger ring sign”, and para-epiphyseal subperiosteal hematoma. Prompt intervention with vitamin C thwarts the progression to severe complications. Radiology is an indispensable tool in diagnosing pediatric scurvy, especially in developmental countries where the assessment of vitamin C serum levels is inaccessible.

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Introduction

Scurvy, resulting from a sustained deficiency of dietary vitamin C, is nowadays an infrequent pathological condition. Nonetheless, intermittent cases surface, predominantly among socioeconomically disadvantaged populations. Prompt diagnosis and intervention are imperative to avert complications and mitigate the manifestations of scurvy. The diagnostic process for scurvy becomes intricate due to its vague clinical presentations, which can be reminiscent of bone neoplasms. In these situations, radiological assessments emerge as vital tools for recognizing scurvy and determining its repercussions on evolving bone structures and articulations [1,2].

Case report

A 6-year-old boy, manifested with persistent pain and an escalating swelling in the right thigh over a span of 2 months prior to hospitalization. The pain described was of a persistent, throbbing nature, intensifying during nocturnal periods and phases of inactivity. Preceding these symptoms, ambulatory challenges were evident for several weeks. The child's guardians endeavored to alleviate measures, administering frequent massages to the edematous thigh; however, the pain and edema were exacerbated. These symptoms were concomitant with generalized lethargy and a non-specific, low-grade pyrexia, persisting for a fortnight before clinical presentation. Prior to this, the child had not undergone any medical evaluation or interventions. There were evident hemorrhagic tendencies in the child's gingiva and labia during dental hygiene practices. Traumatic events were not reported in the patient's anamnesis. Notably, there was no familial predilection towards neoplastic conditions. The child's dietary recall highlighted a significant deficit in the consumption of fruits and vegetables. Subsequent to hospital admission, the child was subjected to an array of laboratory and radiographic evaluations.

Upon clinical assessment, the child displayed heightened irritability. The anthropometric measurement indicated a weight below the first percentile for his age group. Hemorrhagic manifestations were observable on both the gingiva and labia. The articulation of the child's coxal and femoropatellar joints was observed to be in a semi-flexed position, with pain elicited upon attempting full extension. The right thigh presented with pronounced edema, devoid of erythematous changes, petechiae, or ecchymoses. On palpation, a firm, tender mass was discernible spanning the entirety of the right thigh (Fig. 1).

The patient did not exhibit any corkscrew hair manifestations. Laboratory evaluations delineated the following haemato-pathological findings: a leukocytic surge evidenced



Fig. 1 – Swelling of the right thigh. The patient is more comfortable in a semi-flexed hip and knee position.

by a white blood cell concentration of $11,740/\mu\text{L}$, a neutrophil predominance at 83.5%, lymphocytic presence at 9.9%, and a monocyte fraction of 6.2%. Profound anemia was discerned with a hemoglobin concentration measuring 4.8 g/dL and a hematocrit value registering at 16.3 vol%, with normal MCV, MCH, and MCHC values of 76.4 fL, 24.1 pg, and 31.5 g/dL, respectively. A reactive thrombocytotic surge was identified, with platelet concentrations at $751,000/\mu\text{L}$. Parameters including the coagulation profile, glycaemic indices, renal, and hepatic functional markers, along with electrolytic values encompassing sodium, potassium, chloride, and calcium ion levels, were all within accepted physiological parameters. Given these clinical manifestations, the orthopedic consultant postulated a primary bone neoplasm as a differential diagnosis. A radiographic assessment of the right femur and a fine-needle aspiration biopsy (FNAB) was requisitioned to substantiate this.

Radiographic imaging of the right femur shows diffuse osteopenic alterations, coupled with distinct features such as the “Trümmerfeld zone”, “White line of Frankel”, “Pelkin fracture”, “Wimberger ring sign”, and para-epiphyseal subperiosteal hemorrhagic presentations (Figs. 2 and 3). Collectively, these radiological manifestations were suggestive of scurvy. Cytological evaluation via FNAB of the right femur re-

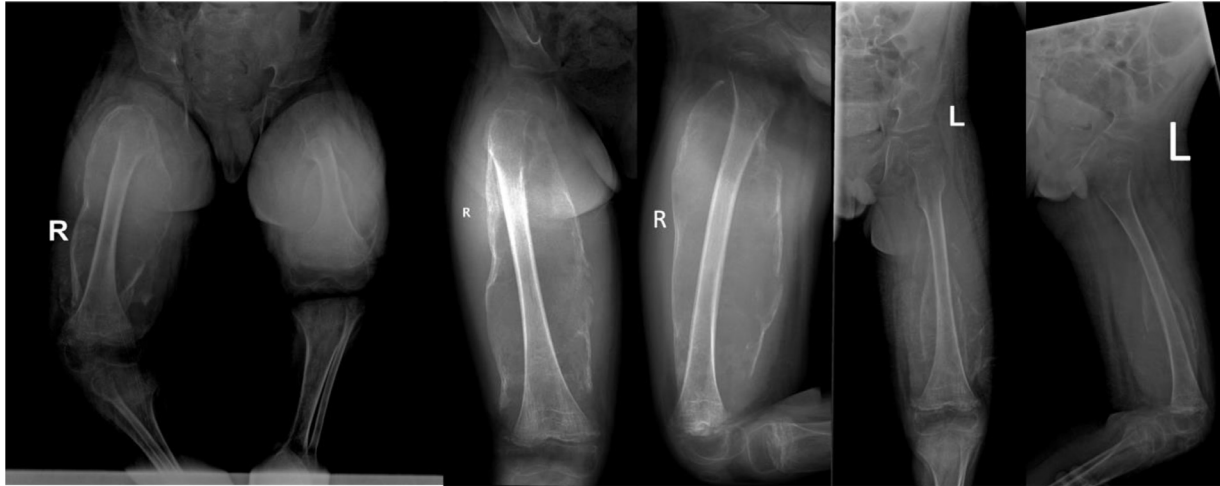


Fig. 2 – The imaging appearance of both femurs of the patient. Note the scorbutic features of both femurs, elaborated in Figure 3.

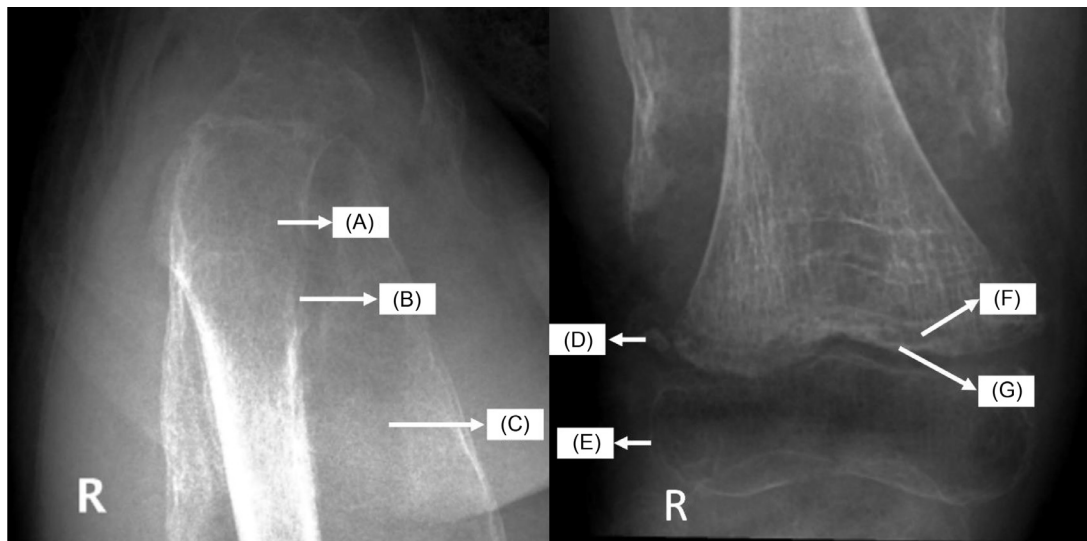


Fig. 3 – The right femur x-ray with its scorbutic features. The findings were consistent with the left femur. (A) Diffuse osteopenia; (B) Pencil-thin cortex; (C) Para epiphyseal subperiosteal haemorrhage; (D) “Pelkin fracture”; (E) “Wimberger ring sign”; (F) “Trümmerfeld zone”; (G) “White line of Frankel”.

vealed large nuclear structures with basophilic cytoplasmic features, a surrounding inflammatory cell, osteoid matrix deposition, necrotic fragments, and erythrocytes; the deduction was indicative of an anomalous osteoblastic response with chronic inflammatory undertones (Fig. 4). Given the paucity of scurvy diagnoses in the preceding decade within the institution and the unconventional histopathological findings, further radiographic assessments of the left femur and thorax were commissioned. The ensuing results resonated with typical radiographic scurvy representations, depicting scorbutic alterations on the left femur and a distinct “scorbutic rosary” delineation on the thoracic radiograph (Figs. 2 and 5).

Subsequently, a comprehensive diagnosis of scurvy was formulated, predicated upon a thorough history taking, physical examination, laboratory findings, hallmark radiological

findings, and histopathological evaluation. The therapeutic regimen entailed analgesics and a high-dose intravenous vitamin C supplementation of 150 mg administered twice a day. After 2 weeks of intensive vitamin C therapy, the clinical manifestations witnessed significant improvements, restoring the patient’s ambulatory capabilities. A maintenance dose of vitamin C, gauged at 100 mg daily, was subsequently prescribed.

Discussion

Scurvy is a relatively rare medical condition arising from a sustained deficiency of vitamin C in the diet. This deficiency is often observed in areas with constrained economic resources.

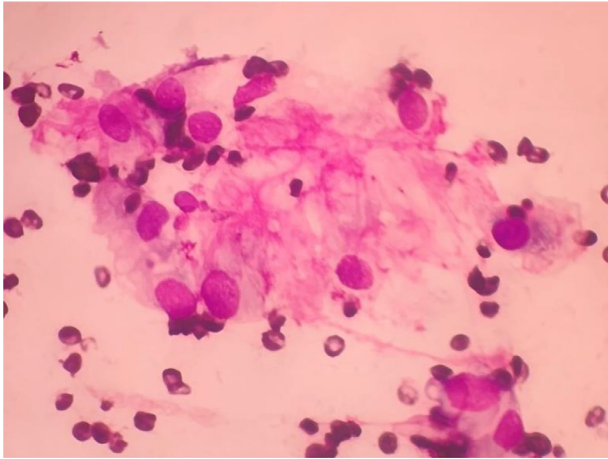


Fig. 4 – The histopathologic evaluation showed an atypical osteoblastic reaction with chronic inflammation.

Collagen, an essential protein integral to the structural integrity of connective tissues—including skin, blood vessels, bones, and gums—depends significantly on vitamin C for its synthesis. Given that humans cannot endogenously produce vitamin C, it is imperative to source it externally from fresh produce or dietary supplements. A shortage of this vital nutrient manifests in a spectrum of symptoms and complications characteristic of scurvy [2,3].

A lack of vitamin C intake for a duration spanning 8-12 weeks can usher in the clinical symptoms of scurvy. Early manifestations are non-specific, encompassing fatigue, predisposition to bruising, and a low-grade fever. An increased fragility in capillaries and impaired platelet aggregation, often seen in scurvy patients, leads to an increased bleeding tendency. Furthermore, a distinct feature of scurvy is its association with various anemias [3,4].

Vitamin C deficiency hinders osteoid bone maturation. The function of vitamin C is to maintain intracellular substances of mesenchymal derivation, such as connective tissue, osteoid tissue in bones, and dentin in the teeth. A deficiency of vitamin C can cause hemorrhagic tendency, leading to subperiosteal bleeding and abnormal function of osteoblasts and chondroblasts. Predominantly, musculoskeletal complications arise from scurvy, more commonly affecting pediatric populations. These complications manifest as edema, arthralgia, myalgia, and muscular hematomas, primarily resulting from fractures at growth plates and hemorrhages occurring subperiosteally and within joints. The mineralization of bones remains intact, as evidenced by the hypercalci-fied provisional cartilage zone. However, the resultant bones are structurally compromised, making them susceptible to pathological fractures [3–5].

Early identification and intervention are paramount to avoid complications and ameliorate the symptoms of scurvy. Given the vague nature of scurvy signs, diagnosis can pose challenges. Comprehensive diagnostic protocols encompass an in-depth dietary history, thorough physical examination, blood investigations, and notably, radiological evaluations. Although assessing plasma vitamin C concentrations is considered the diagnostic gold standard for scurvy, it's not universally available. Furthermore, recent dietary intake of vitamin C can render this metric unreliable [5,6].

Traditional radiographic features play a pivotal role in diagnosing scurvy, with primary alterations observable at the distal ends of long bones, particularly around the knee and ankle joints.

The characteristic bone lesions of scurvy are caused by the cessation of endochondral bone ossification caused by the failure of the osteoblasts to form osteoid tissue. Continuing osteoclastic resorption without adequate formation of new bone yields the appearance of osteoporosis, with generalized osteopenia and thinning of the cortices. Deposition of calcium phosphate continues in whatever osteoid tissue is formed so that an area of increased density develops adjacent to the

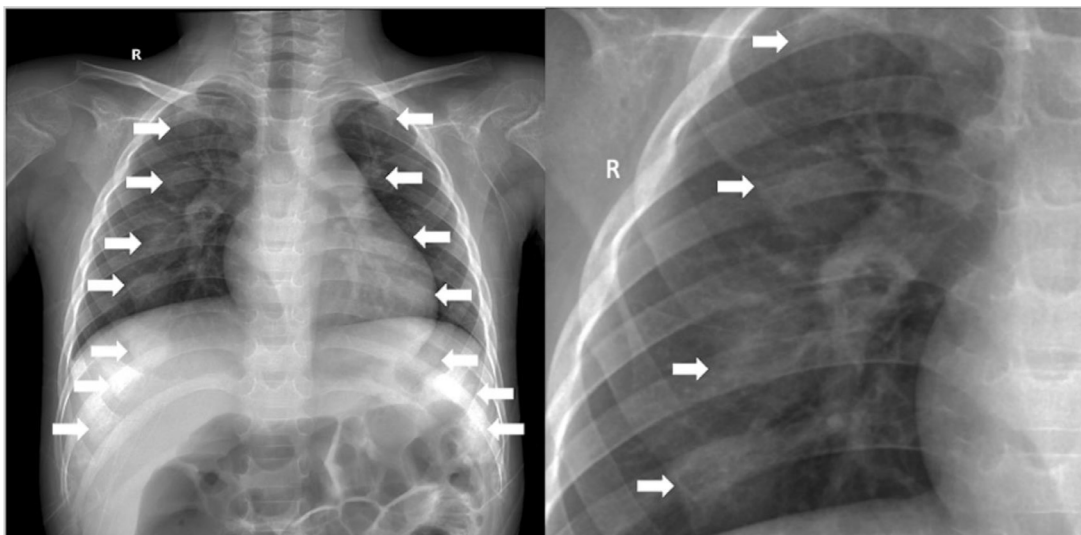


Fig. 5 – Costochondral junction expansion, or “scorbutic rosary”, was seen on the patient’s chest X-ray (white arrow).

growth plate. Such areas have been called the white lines of scurvy or the “white line of Frankel”. A ring of increased density is also seen around the secondary centers of ossification, a finding known as a Wimberger ring sign. Lateral metaphyseal spurs secondary to infarctions will produce a “corner” sign or “Pelkin spur”. The lucent metaphyseal band underlying the Frankel line that known as the “Trümmerfeld zone”, increased capillary fragility leads to subperiosteal and soft-tissue bleeding and the formation of hematomas, which may trigger a periosteal reaction. While osteopenia is a common radiological observation in scurvy, later stages unveil specific features such as the “Frankel line,” the “Trümmerfeld zone,” the “Pelkin spur or fracture,” and the “Wimberger ring sign.” Additionally, chest radiographs might exhibit the expansion of the costochondral junctions known as “scorbutic rosary” and sternum depression [3,4,6,7].

Ultrasonography in scurvy patients can pinpoint anomalies such as bone irregularities and subperiosteal hemorrhages. Given its similarities to certain malignancies, MRI offers a deeper insight into scurvy. The MRI findings in scurvy provide insights into the underlying pathophysiology, revealing areas of hemorrhage at fracture sites within the bones and periosteum, and notably, the absence of the diffuse marrow changes typically observed in hematological malignancies. Furthermore, in the metaphysis, MRI can reveal multifocal symmetrical signal abnormalities along with enhancements in the marrow [2,4].

Prompt detection and treatment of scurvy are instrumental in preventing complications and aiding recovery. The mainstay of scurvy management revolves around vitamin C supplementation, either orally or intravenously. Emphasizing a nutritionally balanced diet that incorporates fresh produce rich in vitamin C is paramount for the long-term prevention and management of scurvy [6].

The subject of this case report presented with progressive swelling and discomfort in the right thigh, characterized by a persistent dull ache, exacerbated nocturnally, and during periods of rest. Concomitant symptoms included generalized fatigue, low-grade fever, irritability, and an evident malnourished physique. Examination revealed an immobile, palpable mass spanning the right thigh. The articulation of the child’s coxal and femoropatellar joints was observed to be in a semi-flexed position to avoid pain. Laboratory investigations indicated significant anemia and reactive thrombocytosis. Given the patient’s clinical presentation and the palpable mass, primary bone malignancy was initially suspected. However, the undernourishment and inadequate dietary intake of fresh produce and vegetables hinted at a possible metabolic bone disorder.

In our patient, the right femur X-ray showcased multiple scorbutic features, which showed diffuse osteopenia, “Trümmerfeld zone”, “white line of Frankel”, “Pelkin fracture”, “Wimberger ring sign”, and para epiphyseal subperiosteal hemorrhages. However, there have been no reported cases of scurvy at our center in the past 10 years. Therefore, to help confirm the diagnosis, additional radiographs were ordered. A subsequent radiograph of the left femur mirrored these findings, which showed the same consistent scorbutic features as the right side, which indicated that the features were found bilat-

erally. Additionally, a thoracic radiograph revealed the characteristic “scorbutic rosary.” The palpable mass was determined to be a consequence of subperiosteal hemorrhages and compromised osteoid bone formation due to vitamin C deficiency. The severe anemia and the bleeding of the gums and lips were thought to be manifestations of iron deficiency caused by impaired iron absorption in the small intestine. Afterward, the patient was given high-dose vitamin C supplementation and maintenance. Post-treatment with high-dose vitamin C supplementation, the patient exhibited a marked improvement, regaining mobility, and returning to daily routines.

Conclusion

Radiology is an indispensable tool in diagnosing pediatric scurvy, especially in developmental countries where the assessment of vitamin C serum levels is inaccessible. The discovery of typical scurvy manifestation in radiography, helps the radiologist and clinician to make the right diagnosis, enabling timely intervention with vitamin C. Differentiating scurvy from other conditions with similar musculoskeletal presentations, such as bone malignancies, is crucial. An integrative approach, optimizing the expertise of radiologists and clinicians, is vital to enhance patient care and outcomes in scurvy presentations.

Patient consent

Written informed consent for the publication of this case report was obtained from the patient.

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