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A 5-Year-Old Boy with Scurvy, Severe Anemia, and Severe Malnutrition: A Challenge

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ABSTRACT

Background: Scurvy is a rare disease caused by a severe vitamin C deficiency in the diet. Due to its rarity compared to other dietary deficiencies, it is seldom recognized, resulting in a typical delay in diagnosis. This issue is more common in children with physical disabilities or poor eating habits. This study aims to examine the medical history of a healthy young individual who was unexpectedly diagnosed with scurvy.

Case description: We report a five-year-old male with hearing impairment presented to our clinic with a movement limitation in lower limbs in one week. The patient's dietary history indicated that meat, fruits, and vegetables were strictly limited. Based on the X-ray results of the right and left genu, there is evidence of spur formation accompanied by sclerosis and irregular metaphysis in the femur and tibia of both sides (known as Pelken's spur). Additionally, there are dense areas with calcification on the surface of the spur (referred to as the Frankel line), as well as reduced bone trabeculation and thinning of the outer layer of the bone (referred to as pencil thin cortex). The presence of radiographic characteristics indicated osteopenia raises suspicions of scurvy. According to the complete blood count examination, the patient had severe anemia. The patient is severely malnourished. Based on the dietary history, clinical symptoms, and radiographic evidence, scurvy was diagnosed.

Conclusion: Our case demonstrates the importance of emphasizing the need for dietary screening to reduce over-examination in patients with non-specific complaints.

Keywords: Anemia, malnutrition, scurvy, vitamin C deficiency.
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INTRODUCTION

Vitamin C is a water-soluble antioxidant that serves as a crucial co-factor in producing collagen, carnitine, and catecholamines metabolism and dietary iron absorption. Since humans cannot produce vitamin C internally, it must be acquired exclusively through consuming fruits and vegetables, including citrus fruits, berries, tomatoes, potatoes, and green leafy vegetables. Vitamin C deficiency leads to scurvy, bleeding, thickened skin, and blood-related irregularities.[1]

Scurvy, often known as a vitamin C deficiency, is an exceedingly ancient illness that has existed for over three thousand years. The onset of scurvy usually occurs within a period of 1 to 3 months following the initiation of a diet lacking in vitamin C. Patients may present with symptoms such as lethargy, weariness, malaise, emotional instability, joint pain, weight loss, loss of appetite, and diarrhea. Additionally, individuals may encounter a heightened susceptibility to bleeding, increased incidence of bruises, and worse wound healing. The dermatological presentations associated with scurvy encompass phrynoderma, corkscrew hairs, perifollicular hemorrhage and purpura, lower extremity edema, and splinter haemorrhages.[2,3]

The incidence of scurvy has significantly decreased over time, leading to occasional oversight by medical professionals in considering it as a potential diagnosis. Consequently, patients often require comprehensive laboratory and radiographic examinations, leading to a delay in both diagnosis and treatment. Scurvy should be considered when a dietary history reveals insufficient vitamin C intake for 1-3 months, accompanied by the aforementioned clinical manifestations. However, it is essential to note that a combination of clinical and radiographic evidence typically establishes the diagnosis of scurvy.[4] This study aims to examine the medical history of a healthy young individual who was unexpectedly diagnosed with scurvy.

CASE DESCRIPTION

A 5-year-old boy came to the pediatric neurology department with bilateral limb weakness. The patient did not experience complete immobility. Instead, the patient demonstrated an inability to bear weight and stand due to discomfort and fear of potential falls. This complaint lasted for one week. The patient refused to stand and walk and reported discomfort in both lower extremities on extension. The weakness experienced was not severe and was localized to the lower extremities. There was a good range of motion in both hands and general body mobility. The patient did not show symptoms such as fever, rash, rhinorrhoea, cough,

sore throat, vomiting, diarrhea, loss of consciousness, or seizures. There is no information about a history of trauma.

These symptoms have been repeated three times. The initial complaint was reported two years ago, where the patient experienced a temporary inability to walk for two weeks, followed by spontaneous resolution. At the same time, the patient felt pain in both legs (Figure 1). The second complaint was reported approximately one year ago, where the parents stated concern that the patient experienced a period of immobility lasting for two months. The patient has sought medical attention and has undergone X-rays. These have revealed no abnormalities.

The patient also looked pale for two weeks before hospital admission. The patient had inadequate nutritious food intake. The patient prefers to consume snacks compared to staple foods. The patient likes to eat crackers. Additionally, the patient exhibits a dislike towards a diet consisting of both meat and vegetables. The patient has been diagnosed with hearing impairment, which was first identified by his parents when he reached 2 years old. Subsequently, he began to use hearing aids at the age of 2 years and 10 months. The patient showed a comparatively delayed rate of development compared to his peers. The patient can walk independently by the age of 24 months.

According to the examination, the patient appeared pale and consistently supported both knees with his hands. The patient uses more signs to communicate. The patient refuses to straighten his lower limbs. Upon the doctor's attempt to straighten the patient's lower limbs, the patient cried and indicated the presence of discomfort. The patient current weight was 12.8 kg (below the 3rd percentile), and his height was 109 cm (<-1 SD). The patient's nutritional state is an indication of a severe case of protein energy malnutrition.

The patient patient underwent a complete blood count (CBC) examination, which revealed the presence of severe microcytic hypochromic anemia. The test indicated a hemoglobin (Hb) level of 5.8, as well as reduced mean corpuscular hemoglobin (MCH) and mean corpuscular volume (MCV). The blood glucose and electrolyte levels were within the normal ranges. The peripheral blood smear and iron panel examination results indicate iron deficiency anemia. Therefore, suspicion may develop into immunological allergic diseases, nutritional-related diseases, and metabolic diseases.

The Department of Allergy-Immunology excluded the possibility of juvenile idiopathic arthritis (JIA) as a diagnosis due to the lack of clinical symptoms and examination findings that align with the established criteria for JIA. To exclude the possibility of chronic inflammatory demyelinating polyneuropathy (CIDP), the patient underwent nerve conduction investigations and electro-neuromyography. The results were found to be within normal parameters. Based on the X-ray results of the right and left genu, there is evidence of spur formation accompanied by sclerosis and irregular metaphysis in the femur and tibia of both sides (known as Pelken's spur). Additionally, there are dense areas with calcification on the surface of the spur (referred to as Frankel line), as well as reduced bone trabeculation and thinning of the outer layer of the bone (referred to as pencil thin cortex). The radiographic characteristics indicated osteopenia, raising suspicions of scurvy (Figure 2).

Subsequently, the patient received an oral administration of vitamin C at a dosage of up to 250 mg per 24 hours. After administering high doses of vitamin C, the patient's condition improved within three days. The lower extremities have initiated movement, and the sensation of discomfort has decreased. The patient received stabilization phase therapy, including F-75 and subsequent treatment with F-100, to manage the severe malnutrition. Upon the conclusion of the hospitalization period, the patient started consuming standard formula milk.

DISCUSSION

Scurvy is a disease caused by a persistent vitamin C deficiency. It is seldom detected, which usually results in delayed diagnosis. It is early identification and suitable treatment typically yield favorable outcomes. Bruising, arthralgias, and

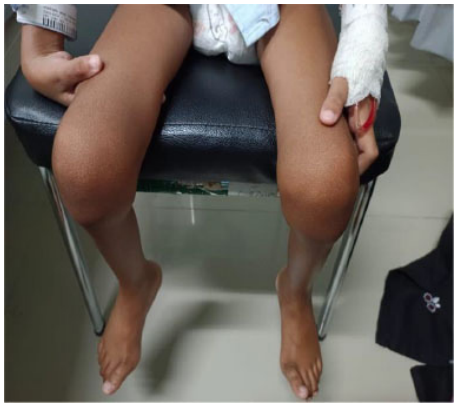

Figure 1. A 5-year-old patient is experiencing bilateral leg pain and exhibiting a reluctance to mobilize their lower limbs.

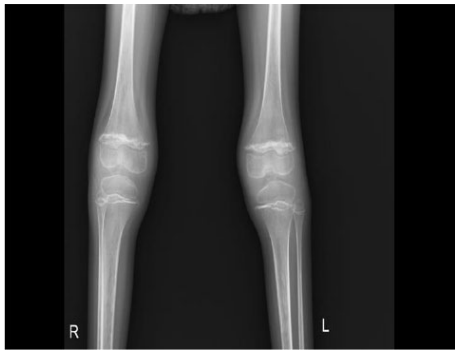

Figure 2. The radiological examination revealed the presence of Pelken's spur, along with the observation of Frankel lines and a pencil-thin cortex, which provides evidence establishing the diagnosis of scurvy.

bone disease are among the main clinical symptoms of this illness, which causes poor tissue healing.[5] Vitamin C is one of the essential vitamins that has a role in collagen synthesis. Collagen is crucial for preserving the strength and integrity of the body's connective tissues. In addition, vitamin C also acts as a cofactor that promotes procollagen transcription into collagen and allows crosslinking to help proline and lysine hydroxylases stabilize collagen type I and VI. Therefore, inadequate taking of vitamin C can lead to scurvy.[6]

In this case report, the patient was a 5-year-old boy who came to the pediatric neurology department with bilateral limb weakness that led to limitations in standing or walking. He is the family's first child from low to middle income. Every

time he refused to eat, the family would let him do so because they did not want the patient to cry and agitate. They try to give him food, but he only wants to eat tempeh, tofu, egg, or fish. He does not like poultry, meat, fruits, and vegetables. The parents also gave the patient milk at home but it was usually more diluted.

The patient was not showing any symptoms such as fevers, rashes, rhinorrhea, cough, sore throat, vomiting, diarrhea, loss of consciousness, or seizures. No history of trauma was reported. The patient's complaint has been repeated three times from two years ago. On examination, the patient has a pale appearance and has a severe case of protein energy malnutrition. The result of the CBC examination was microcytic hypochromic anemia with Hb 5.8. In this patient, he has severe anemia but without hemorrhagic manifestation. Radiological examination demonstrated that Pelken's spur, Frankel line, and pencil thin cortex impressing osteopenia with radiological features suspicious for scurvy.

The clinical presentation observed in this patient differed from that described in other case reports, which typically include symptoms such as fever, purpura, ecchymoses, and perifollicular hemorrhages. However, all of the case reports indicated the presence of limited movement in the lower extremities. The diagnostic tests included a complete blood count (CBC), renal and hepatic function assessment, blood culture analysis, and radiologic examination. Most of these examinations have been documented in this report to confirm the diagnosis of scurvy.[7-10]

In this case, the diagnosis of vitamin C deficiency was established by evaluating clinical manifestations and assessing the individual's nutritional background. To address the deficiencies, we implemented a therapeutic regimen consisting of a daily administration of 250 mg of vitamin C orally alongside a nutritionally balanced diet. After administering high doses of vitamin C, the patient's condition improved within three days. Other literature indicates a range of oral dosages, varying from 100 mg to 300 mg up to 4000 mg per day, administered in divided doses. The duration of treatment exhibits variability and should last until complete clearance of clinical symptoms, with a range spanning from a minimum of two weeks to a maximum of seven months.[11]

CONCLUSION

Scurvy is a rare disease, and its clinical symptoms are similar to those of other diseases, so misdiagnosis is common. The presence makes the diagnosis of scurvy of symptoms of unexplained rash, discomfort, reluctance to eat, limb weakness, and history of inadequate food intake, with radiological examination revealing the presence of Pelken's spurs, together with the observation of Frankel's lines and pencil-thin cortex, providing evidence to confirm the diagnosis of scurvy.

CONFLICT OF INTEREST

The author declared that there is no conflict of interest.

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None.

AUTHOR CONTRIBUTION

All of the authors contribute in all step manuscript preparation.

INFORMED CONSENT

The patient's parents have agreed to the publication of this case. Written informed consent has also been obtained from the patient's parents.

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