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**Case Report**

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# A rare case of ribbing disease of the proximal tibia

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## ABSTRACT

Ribbing disease is a rare disease that causes bony growths to develop in long bones mostly the lower limbs. It often develops after puberty. The disease also goes by other names like multiple diaphyseal sclerosis. The disorder is caused by discrepancy in the osteoclast regulation process. Here in we report a 28 years old male presenting with a case of ribbing disease of right proximal tibia. A 28 years old male gives history of fall when he was 6 years old i. e., 22 years back. Now patient complaints of pain in right proximal tibia since last 6 months. Pain is of mild nature. It is most likely a case of ribbing disease based on the clinical, radiological findings. Ribbing disease is a disease of exclusion, needs high degree of suspicion for its diagnosis. However, patient should be advised for regular follow ups.

**Keywords:** Ribbing, Tibia, Sclerosis, Pain

## INTRODUCTION

Ribbing disease is a rare form of sclerosing dysplasia characterized by benign endosteal and periosteal bone growth confined to the diaphysis of the long bones, usually the tibia and femur. It occurs after puberty and is more commonly seen in women.[1] The most common presenting symptom is pain that is usually self-limited; however, progression is known. The etiology and optimal treatment for the disease are as yet undefined.[2] We here are presenting a case of 28 years old male gives history of fall when he was 6 years old with clinical, radiological and bone scan manifestations of Ribbing disease.[3]

Ribbing disease goes by other names, including multiple diaphyseal sclerosis, diaphyseal sclerosis, and heredity multiple diaphyseal sclerosis.[4] It is also is a part of a group of disorders called sclerosing bone dysplasias-rare genetic diseases characterized by abnormally thick and overgrown bones.

These disorders are usually caused by defects in the osteoclast regulation process (replacement of old bone with new bone), leading to abnormal accumulation of bone.[5]

## CASE REPORT

28 years old male gives history of fall when he was 6 years old i.e., 22 years back. Now patient complaints of pain in right proximal tibia since last 6 months which is of mild nature. There is no swelling. Range of motion was terminally painful. Also, the patient did not have any neurovascular deficit. Further investigations like X-rays. Anteroposterior and lateral radiograph of both legs demonstrated sclerosis with cortical thickening involving the diaphysis of both tibias, which was more prominent on the right side. A computed tomography CT scan which was patient was advised CT scan on 12$^{th}$ august 2022 it showed ill-defined undulating sclerosis of the right tibial cortex circumferentially but asymmetrically in the proximal diaphysis. This was also seen on an MRI on 17$^{th}$ august 2022. There is no marrow involvement. The cortex shows asymmetric thickening. In two areas, one being anteromedial and the other posterolateral, serpiginous vessels are seen, which on the dynamic study show an

enhancement pattern suggestive of venous flow. No peri osseous or marrow edema was seen.

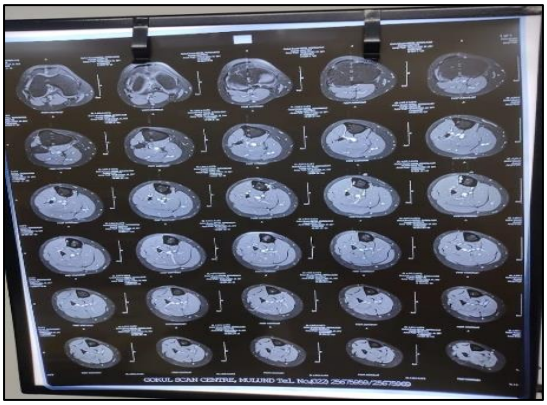

**Figure 1: CT scan of proximal tibia.**

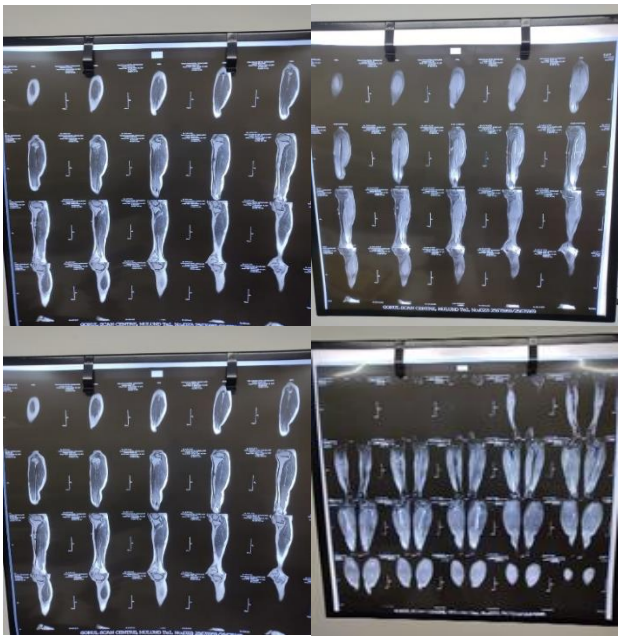

**Figure 2: MRI of proximal tibia.**

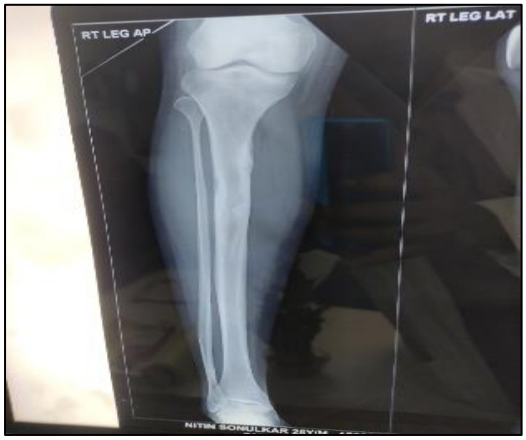


**Figure 3: X-ray tibia ap.**

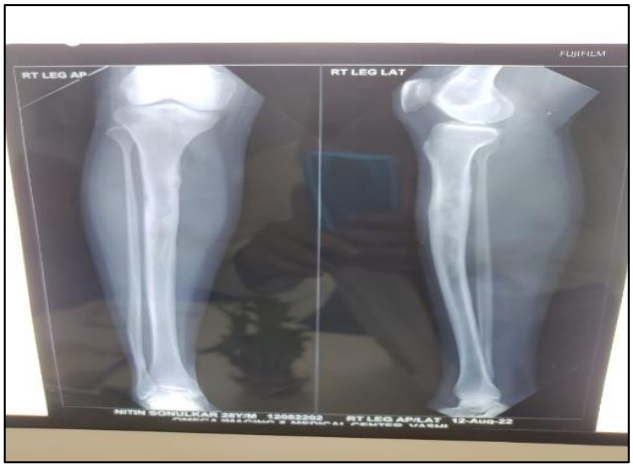


**Figure 4: X-ray tibia AP/Lat.**

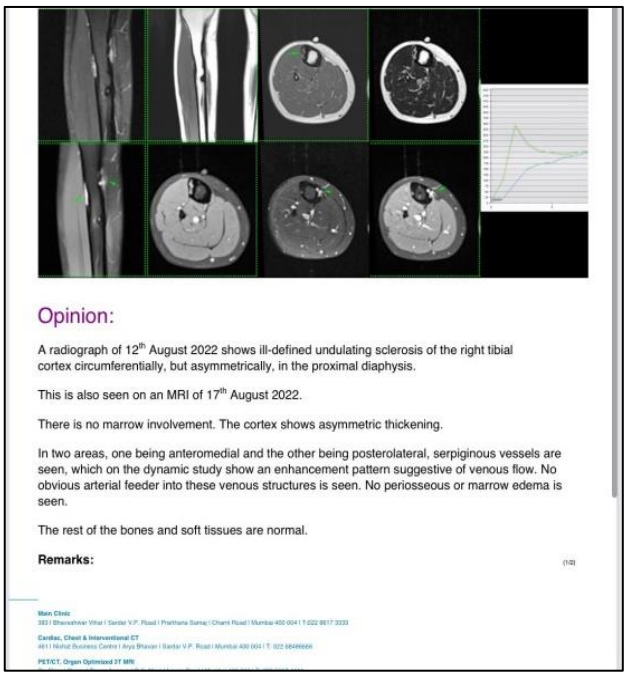


**Figure 5: Printed report.**

The rest of the bone and soft tissues are normal.

Based on the above radiological and histopathological correlation, the patient was diagnosed to have ribbing disease by exclusion method.

## DISCUSSION

The limited literature on Ribbing disease leaves researchers to speculate there have only been 20 to 30 cases reported.

Ribbing described a family with six siblings for the first time in 1947 in which four of them had asymmetrical diaphyseal sclerosis of long bones. He designated the term hereditary multiple diaphyseal sclerosis. This was later

found to be an uncommon cause of leg pain and in further case studies was called Ribbing disease.[6]

It is important to differentiate this entity from the more common causes of bony leg pain like stress fractures, shin splints, osteomyelitis, fibrous dysplasia, osteoid osteoma, osteosarcoma, and other rarer causes like adamantinoma, melorheostosis, hyperphosphatasia, histiocytosis, lymphoma, intramedullary sclerosis, endosteal hyperostosis and sclerosteosis.[7]

Bone dysplasia like Engelmann disease are a close differential of Ribbing disease. The distinction between Ribbing disease and Camurati-Engelmann disease (progressive diaphyseal dysplasia) has been unclear.[8] Engelmann disease has been more frequently reported in literature. It is a progressive disorder associated with pain, muscle weakness, fatigue, waddling gait, and anemia. Though Engelmann disease and Ribbing disease may appear to be identical radiographically, Seeger et al pointed to certain clinical and histologic differences.[9] Engelmann disease presents during childhood, while Ribbing disease usually presents after puberty. Engelmann disease presents with bilateral and symmetric bone involvement, whereas Ribbing disease is either unilateral or asymmetrically and asynchronously bilateral. Engelmann disease affects diaphyses of long bones and bones formed by intramembranous ossifications; hence, the skull is involved almost as frequently as the long bones. Ribbing disease has been reported only in the long bones.

While Engelmann disease features trabecular thickening, normal or enlarged haversian systems, and both osteoblastic and osteoclastic activity, implying bone formation and resorption, histologic study in Ribbing disease shows osteoblastic activity alone and progressive obstruction of the haversian systems.[10] It is thought that there may be a difference in the mode of inheritance. Engelmann disease is autosomal dominant, with considerable variation in penetrance. Autosomal recessive inheritance appears to be demonstrated in Ribbing disease. Makita et al reported a three-generation Japanese family with Engelmann disease with a wide variation in phenotype among the affected family members. Of the 12 patients, 7 had full manifestations of Engelmann disease, while the other 5 exhibited only segmental (rhizomelic and/or mesomelic) involvement and asymmetric diaphyseal sclerosis without any clinical symptoms, resembling Ribbing disease. The authors proposed that Engelmann disease and Ribbing disease represent phenotypic variations of the same disorder.[11]

Ribbing's disease presents with pain, and recent studies have demonstrated that marrow edema is often associated with, and may be responsible for, at least in part, this pain. The MRI appearance and the prominent finding of pain in Ribbing disease may be useful in helping to differentiate Ribbing disease from intramedullary osteosclerosis, another diaphyseal dysplasia disease which bears perhaps the greatest clinical and radiographic similarity to Ribbing disease. It does not appear that the MRI finding of marrow edema is characteristic of intramedullary osteosclerosis.[12] This finding leads to what has been thought to be a potential treatment for Ribbing disease, the creation of a wide surgical "window" at the site of pain. One may also consider the diagnosis of painful transient tibial edema, which clinically resembles the presentation here. However, in that condition, the radiographs are usually normal.

In Ribbing disease, markers for bone formation like alkaline phosphatase and osteocalcin are not elevated. In addition, the markers of bone resorption like N-telopeptide, pyridinoline and deoxypyridinoline cross-links are also normal. Together, these measures of bone metabolism would suggest that Ribbing disease is not a "high turnover" bone state, a finding which may help to differentiate it from Camurati-Engelmann disease.

The treatment of ribbing disease is mainly symptomatic and is done with nonsteroidal anti-inflammatory drugs in increasing dose as required. However, bisphosphonates have been used in some cases of Ribbing and Camurati-Engelmann diseases, with discordant outcome; however, the low bone turnover that seems to characterize this disease and the prevalence of osteoblastic activity over the osteoclastic action are probably the reasons why osteoclast inhibitors may have limited success.

Because it is rare and a lot of healthcare providers don't know much about it, diagnosis is often delayed and confused with other sclerosing bone dysplasias, metabolic diseases (genetic conditions that cause metabolism problems), or osteomyelitis (infection of the bone).

A diagnosis of Ribbing disease can be made after reviewing clinical history, bloodwork, and imaging, and ruling out other conditions. For your clinical history, your healthcare provider will want to know what symptoms you are experiencing, including bone pain and muscle weakness, and the location of the pain.

Bloodwork for most people with Ribbing disease will be normal.[7] Normal bloodwork rules out inflammatory diseases.

So, in conclusion Ribbing's disease should always be diagnosed only by exclusion basis and we've done the same in our case.

## CONCLUSION

In the end we conclude that these investigations points towards no specific differential. It is most likely a case of Ribbing's disease based on the clinical, radiological findings. So, we should always keep in mind these differentials and not do unnecessary investigations.

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