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(CASE REPORT)

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Ribbing disease of bilateral tibial diaphyses: A case report and literature review

El Bachir Mghabar *, Mouad Miloudi, Rachid Alaoui Hachimi, mohammed Anas Benamer, Hatim Abid and Mohammed El Idrissi and Abdelmajid El Mrini

Department of Orthopedics and Traumatology, University Hospital Center Hassan 2 of Fez, Morocco.

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Abstract

Ribbing disease is a rare sclerosing bone dysplasia characterized by benign endosteal and periosteal bone growth confined to the diaphyses of long bones, typically affecting the lower extremities. We present a case of a 31-year-old female with bilateral tibial diaphyseal involvement, highlighting the diagnostic challenges, clinical presentation, and management of this rare condition. The patient presented with progressive pain in both tibiae, which was initially misdiagnosed as chronic sclerosing osteomyelitis. Radiographic imaging, including X-rays and MRI, revealed characteristic diaphyseal sclerosis with medullary canal obliteration. Histopathological examination confirmed the absence of malignancy or infection. The patient underwent surgical fenestration of the sclerotic bone, resulting in significant pain relief. This case underscores the importance of considering Ribbing disease in the differential diagnosis of unilateral or bilateral lower extremity pain with diaphyseal sclerosis, and highlights the role of surgical intervention in managing refractory pain.

Keywords: Ribbing disease; Pain; Sclerosing bone dysplasia; Bone fenestration

1. Introduction

Ribbing disease, first described by Ribbing in 1949 [1], is a rare sclerosing bone dysplasia characterized by benign endosteal and periosteal bone growth, primarily affecting the diaphyses of long bones, particularly the tibiae and femora. The condition typically presents in young adults, with a female predominance, and is often associated with progressive pain localized to the affected bones. Due to its rarity and nonspecific clinical presentation, Ribbing disease is frequently misdiagnosed as chronic sclerosing osteomyelitis, stress fractures, or other sclerosing bone dysplasias such as Camurati-Engelmann disease [2].

This case report describes a 31-year-old female with bilateral tibial diaphyseal involvement, emphasizing the diagnostic process, imaging findings, and surgical management.

2. Case Report

A 31-year-old female presented with a 3-year history of progressive, dull aching pain in both tibiae, exacerbated by physical activity and unresponsive to nonsteroidal anti-inflammatory drugs (NSAIDs). There was no history of trauma, infection, or systemic illness. Physical examination revealed tenderness over the midshaft of both tibiae, with no signs of inflammation, erythema, or warmth.

Plain radiographs demonstrated increased diaphyseal density and cortical thickening in both tibiae, with obliteration of the medullary canal (Figure 1). Computed tomography (CT) scans confirmed periosteal and endosteal sclerosis, while magnetic resonance imaging (MRI) revealed bone marrow edema in the affected diaphyses (Figure 2).

* Corresponding author: El Bachir Mghabar

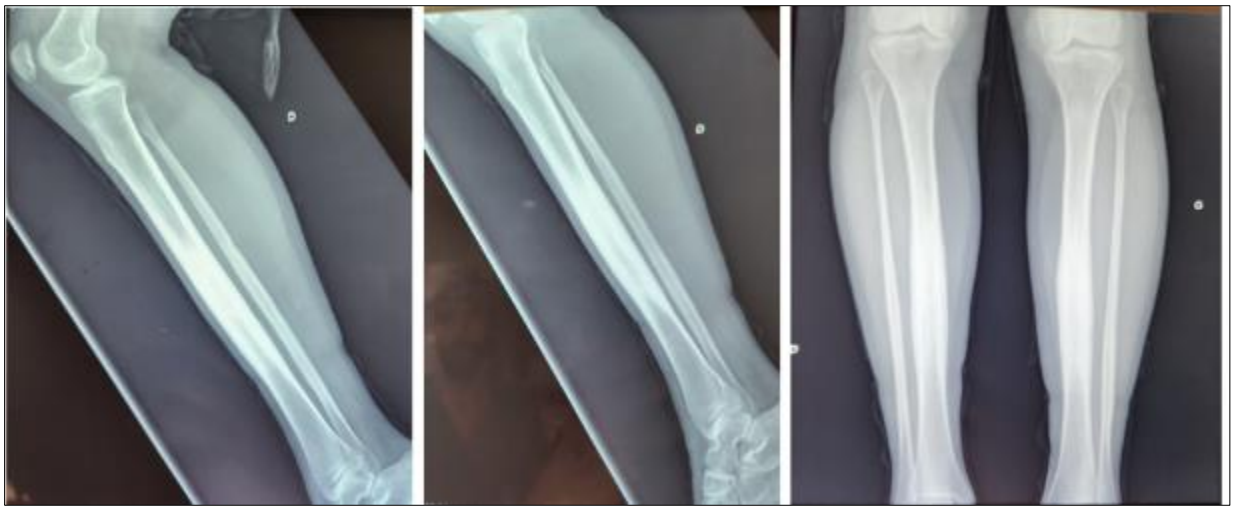

Figure 1 Anteroposterior and lateral radiograph of both legs showing midshaft sclerosis of tibia with obliteration of medullary cavity

Laboratory investigations, including complete blood count, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and serum calcium, phosphorus, and alkaline phosphatase levels, were within normal limits.

An open biopsy of the affected tibial diaphysis was performed, revealing thickened trabeculae with increased osteocyte density and no evidence of infection or malignancy . Cultures of the biopsy specimen were sterile.

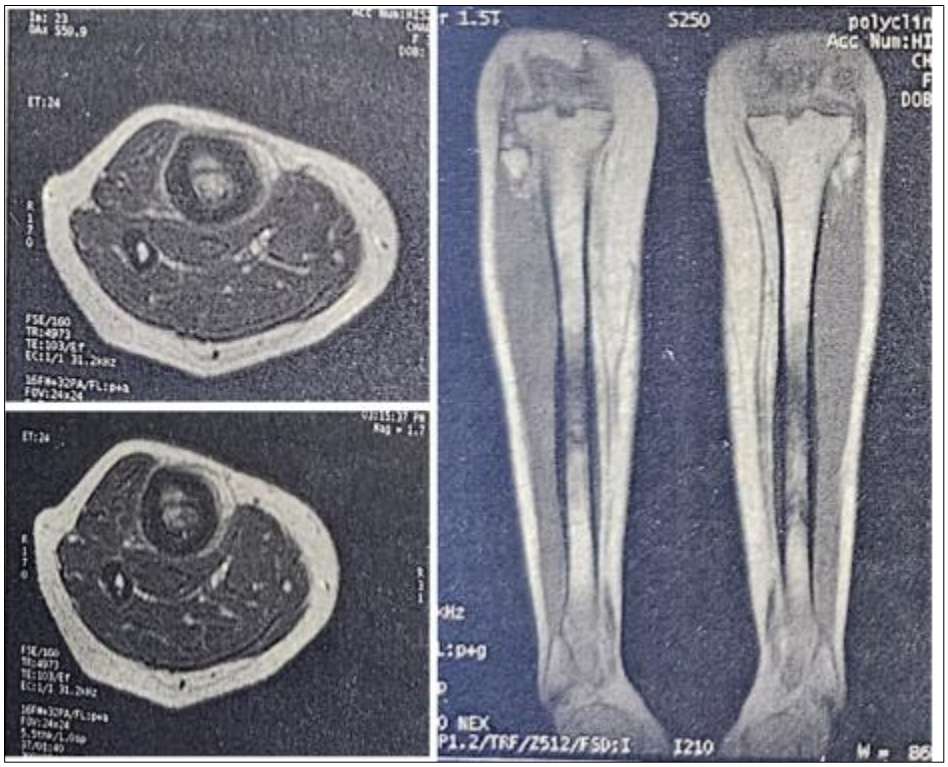

Figure 2 Coronal (a) and axial (b) images of magnetic resonance imaging (MRI) showing massive endosteal sclerosis, cortical thickening with bone marrow edema in the diaphysis of both tibias

Given the refractory nature of the pain and the radiographic findings, the patient underwent surgical fenestration of the sclerotic bone in both tibiae. The procedure involved creating a cortical window and removing the medullary sclerotic bone using a bone drill(Figure 3). Postoperatively, the patient reported significant pain relief, with a reduction in pain

score from 8 to 2 on the visual analog scale (VAS). At the 2-year follow-up, the patient remained pain-free, with no evidence of sclerotic bone regeneration on repeat imaging .

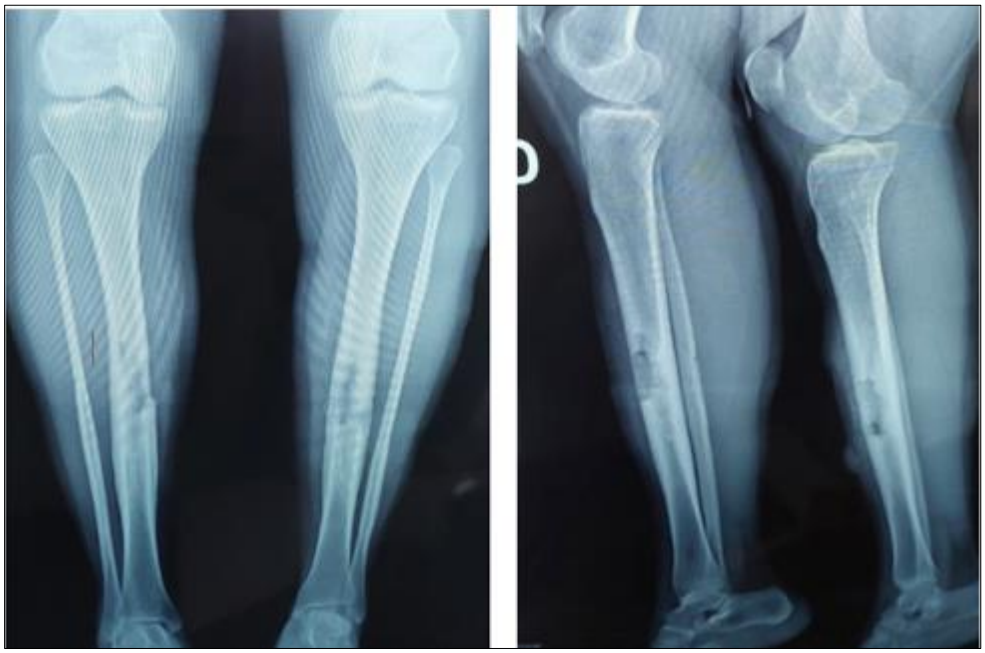

Figure 3 Final postoperative radiograph of affected legs after bone fenestration intramedullary reaming

3. Discussion

Ribbing disease is a rare condition that poses significant diagnostic challenges due to its nonspecific clinical presentation and radiographic findings. The disease is often misdiagnosed as chronic sclerosing osteomyelitis or other sclerosing bone dysplasias, such as Camurati-Engelmann disease [2]. However, Ribbing disease typically presents unilaterally or asymmetrically, with sparing of the metaphyses and epiphyses, and is not associated with systemic symptoms or laboratory abnormalities.

The etiology of Ribbing disease remains unclear, although some authors have suggested a possible genetic component, with autosomal recessive inheritance [3]. The condition is characterized by abnormal bone remodeling, with excessive endosteal and periosteal bone formation leading to medullary canal obliteration and increased intramedullary pressure, which may contribute to the pain [4].

Treatment options for Ribbing disease are limited, with no established medical therapies. NSAIDs may provide symptomatic relief in some cases, but surgical intervention, such as intramedullary reaming or fenestration of the sclerotic bone, is often required for refractory pain [5]. Surgical decompression of the medullary canal has been shown to provide significant and lasting pain relief, as demonstrated in this case [6].

4. Conclusion

Ribbing disease is a rare sclerosing bone dysplasia that should be considered in the differential diagnosis of unilateral or bilateral lower extremity pain with diaphyseal sclerosis. Accurate diagnosis requires a combination of clinical, radiographic, and histopathological findings. Surgical intervention, such as fenestration of the sclerotic bone, can provide significant pain relief and improve quality of life in patients with refractory symptoms.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of informed consent

Informed consent was obtained from the patient for publication of this case report and accompanying images.

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