Zebra stripe sign: osteogenesis imperfecta timeline

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INTRODUCTION

Osteogenesis imperfecta (OI), also known as brittle bone disease, is a rare and clinically heterogeneous genetic connective tissue disorder. It manifests mainly as bone fragility, with increased susceptibility to fractures, deformities, and significant growth abnormalities, although other organs can be involved¹. It is commonly caused by autosomal-dominant heterozygous mutations in one of the two genes encoding type I collagen, COLIA1 and COLIA2. Mutations in other genes have also been documented¹.

Treatment is based on symptom reduction through a multidisciplinary approach consisting of pharmacologic therapy, surgical treatment, and rehabilitation¹. The standard pharmacologic approach of children with OI relies in bisphosphonates (BP).

CASE REPORT

A 27-year-old man with OI (history of multiple fragility fractures treated with five cycles of intravenous pamidronate disodium during childhood) was given entry at the emergency department after reporting pain in the right knee, following by a direct trauma. Swelling, painful mobilization and limited range of motion were shown upon physical examination.

Radiographs of the knee revealed a proximal tibial fracture (Figure 1,*). Surgical treatment was performed.

The radiographs had also shown five metaphyseal bands of increased density paralleling the contours of the physes in the distal femur, proximal tibia, and fibula (Figure 1, arrows). This finding was also present in other metaphyses, such as the distal tibia and fibula.

These metaphyseal bands - also described as zebra stripe sign- represent a radiological manifestation of cyclical BP therapy prior to closure of epiphyseal growth plates³. They usually develop after about two months of BP therapy.³ These sclerotic lines are then the result of increased bone formation and decreased bone resorption, representing the newly restored balance between osteoblastic and osteoclastic activity³. The new bone, formed in between cycles of BP and within sclerotic lines, presents normal lucency. Each sclerotic line corresponds to a BP cycle, with the resulting distance between lines influenced by bone growth rate, frequency of BP courses and the patients’ age³.

The possible clinical implications of these lines are currently unexplored. However, they can serve as a helpful timestamp of cyclical BP therapy.⁴ With the increasing usage of BP in various pediatric skeletal disorders, physicians need to be aware of the implications in bone physiology and expected imaging findings, such as the zebra stripe sign.
FIGURE 2. Antero-posterior and lateral radiograph of right ankle - arrows: metaphyseal bands of increased density paralleling the contours of the physis.

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REFERENCES