

## Radiological “bone within a bone” appearance with atraumatic fractures

### CASE REPORT

A 6-year-old boy presented with progressive pain in the right groin for a couple of months. Physiotherapy was initially started because of no apparent trauma, but did not deliver any relief. An x-ray examination of the hip showed a Salter–Harris type 2 fracture of the femoral neck without any callus formation (fig 1). The medical history consisted of fractures in the hands and feet. On physical examination, a head circumference +2.5 standard deviations of the mean was noticed. Bone density was assessed by dual-energy x-ray absorptiometry scan and was 10 standard deviations above the mean for the patient’s age for both the lumbar spine and both hips. Radiographs confirmed the increased bone density and showed a typical “bone within a bone” appearance in both hands (fig 2) and feet.

### DIAGNOSIS

The figures show a “bone within a bone” appearance that is linear in the phalanx and arcuate beneath the iliac crests. This radiological appearance in combination with increased bone density and bone brittleness is characteristic of osteopetrosis. However, bone within a bone may also occur as a manifestation of diverse conditions.<sup>1</sup>

Osteopetrosis, also known as Albers–Schoenberg or marble bone disease, is the classic example of a primary hereditary disorder that produces a bone within a bone appearance.<sup>1</sup> Osteopetrosis is a heterogeneous group of heritable conditions in which there is a defect in bone resorption by osteoclasts. This defect affects the shape and structure of bone by altering its capacity to remodel during growth, and contributes to the brittleness of bone in osteopetrosis.<sup>2</sup> An intermittent process of normal and abnormal bone production forms the bone within a bone as a miniature inset of earlier constructed bone within the confines of the bone. Several disease-causing gene mutations of osteopetrosis have been identified. Thus far, all genes associated with osteopetrosis in humans encode proteins that participate in the functioning of the differentiated osteoclasts.<sup>3</sup>

In severe cases, the medullary cavity may be affected because of new endochondral bone, which interferes with development of haematopoietic cells, resulting in thrombocytopenia, anaemia, severe infections and hepatosplenomegaly due to extramedullary haematopoiesis.<sup>2</sup> Visual loss caused by bony encroachment of the optic nerve at the level of the optic foramina may also occur in severe cases and require surgical



**Figure 1** Radiograph of the hip showing a Salter–Harris type 2 fracture of the right femoral neck without any callus formation.



**Figure 2** Radiograph of one hand confirming increased bone density and showing a typical “bone within a bone” appearance.

intervention.<sup>4</sup> In severely affected children, haematopoietic stem-cell transplantation is described as a therapeutic possibility.<sup>2,4</sup> The transplanted marrow contains progenitor cells and normally functioning osteoclasts, which will correct the bony and haematological manifestations.<sup>2</sup> Orthopaedic management is required for the surgical treatment of fractures and their complications including malunion and post-fracture deformity.

No other manifestations of osteopetrosis were apparent in this case except for the bone brittleness and the enlarged head circumference. Considering the absence of other family members with symptoms of osteopetrosis, we diagnosed the mild form of autosomal recessive osteopetrosis. On follow-up after 5 months, the patient had broken three metatarsal bones, and x-ray examination showed minimum callus formation with an apparent fracture line in the femoral neck.

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### REFERENCES

1. Williams HJ, Davies AM, Chapman S. Bone within a bone. *Clin Radiol* 2004;**59**:132–44.
2. Tolar J, Teitelbaum SL, Orchard PJ. Osteopetrosis. *N Engl J Med* 2004;**351**:2839–49.
3. Balemans W, Van Wesenbeeck L, Van Hul W. A clinical and molecular overview of the human osteopetroses. *Calcif Tissue Int* 2005;**77**:263–74.
4. Wilson CJ, Vellodi A. Autosomal recessive osteopetrosis: diagnosis, management, and outcome. *Arch Dis Child* 2000;**83**:449–52.



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