Callus distraction in Ollier's disease

A case report

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An 8-year-old girl complained of increasing difficulty in walking. She had a 20 percent limb length discrepancy, and was reliant on a large shoe raise and crutches. Radiographs showed multiple enchondromata throughout the right femur and tibia (Figure 1), producing 7.5 cm of femoral and 4 cm of tibial shortening. The femoral neck was in valgus and the distal shaft in varus. There was varus and external torsion of the tibial shaft. Ollier's disease (enchondromatosis, dyschondroplasia) is characterized by multiple enchondromata.

Surgical correction began with the application of a circular external skeletal fixator to the femur, and corticotomy of the distal metaphysis. 1 week later, a similar frame was applied to the tibia and a proximal metaphyseal corticotomy performed (in all, 12 pins with a diameter of 1.8 mm each were used). Peroperative correction of the 30 degree tibial torsion was achieved, but the other deformities were corrected gradually.

Lengthening of each bone was begun after 14 days, at a rate of 1 mm per day in 4 divided increments. After 1 cm of axial distraction, coronal plane deformity was corrected, using appropriately-sited hinges and differential rates of distraction on the inner and outer threaded rods.

Several complications were encountered. A radiolucent lesion seen in the femur radiographically was dealt with by slowing down the distraction for 7 days. Thereafter, rapid bone proliferation was noted radiographically (Figure 2) and ultrasonographically in both the femur and tibia, despite a return to distraction at 1 mm per day in both limb segments.

A severe cavovarus deformity developed in the ankle, together with paresthesias of the dorsum of the foot. Tibial lengthening was therefore stopped after distraction had reached 2.5 cm (10 percent lengthening). Nerve recovery and hindfoot mobility rapidly recovered. Femoral lengthening was continued beyond the original discrepancy to 9.5 cm (33 percent lengthening) in anticipation of future retarded growth.

Pin-tract infections occurred around many of the fine wires, but were more severe around the proximal femoral half-pins. They were treated by antibiotics and topical skin care and no pin required removal for infection or breakage.

The tibial frame was removed at 4 months and the femoral frame at 6 months, without subsequent bending or fracture. Correction of the limb length discrepancy was achieved, albeit with undercorrection of the tibial shortening and overcorrection of the femoral length. Minimal external rotational malalignment of the tibia persisted during lengthening, but resolved subsequently. Knee stiffness became severe, but gradually improved and 2 years after lengthening the range of movement was 0–120 degrees.

Discussion

Osteogenesis under tension may occur by membranous rather than by endochondral ossification (Ilizarov 1989, Aronson et al. 1990). The osteotomies in our patient were made through defective bone in which cartilage predominated, as seen on the radiographs, and it is interesting that the response during distraction was replacement not with an extension of the cartilaginous tissue but with new bone, which rapidly corticalized (Figure 3). In this way, the inherent bone weakness due to enchondromata has probably been improved in those segments. Radiographic evidence of radiolucent spots being replaced by new bone has been shown earlier in a case of enchondromatosis in which lengthening was performed by interposition of allogeneic bone grafts (Urist 1989).

Despite the formation of a radiolucent lesion in the region of distraction, the formation of bone was unusually rapid, and nerve and tendon stress were the limiting factors rather than the osteogenic capacity. External torsion of the limb persisted during lengthening, possibly due to tension in the iliotibial tract; the torsion returned to normal within 6 months after

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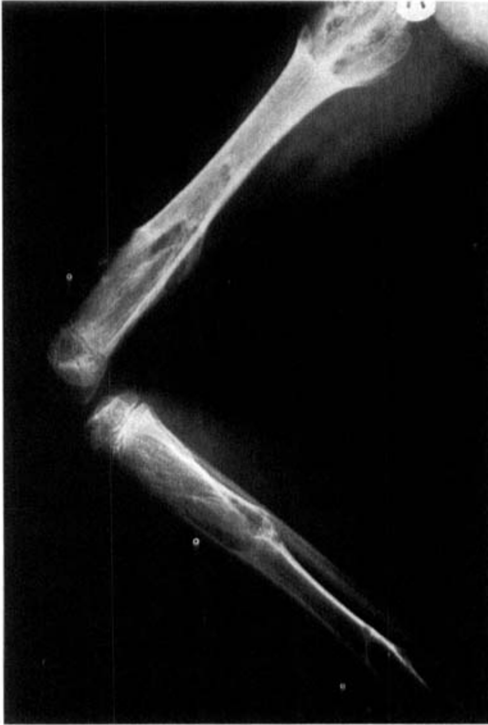

Figure 1. Preoperatively. Multiple enchondromata in femur and tibia.

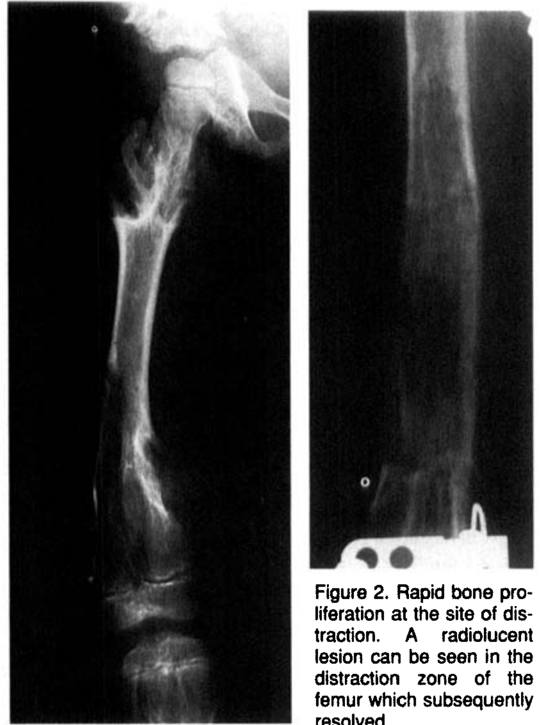

Figure 2. Rapid bone proliferation at the site of distraction. A radiolucent lesion can be seen in the distraction zone of the femur which subsequently resolved.

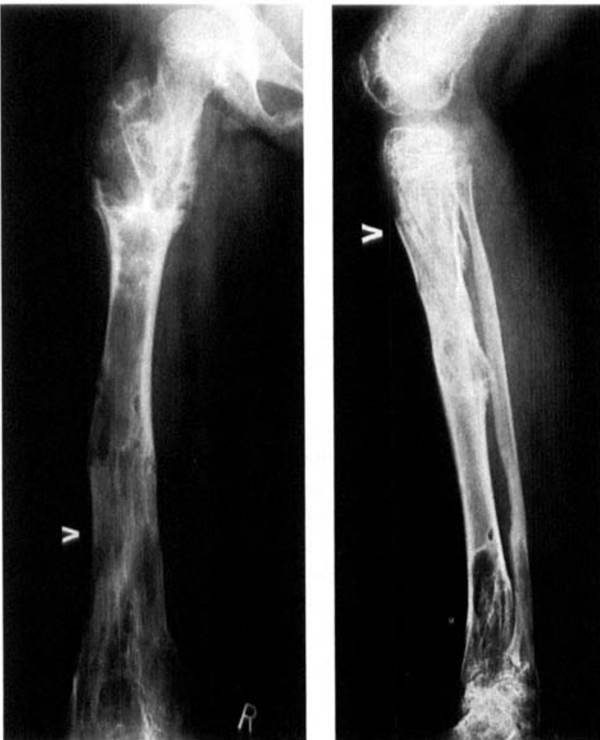

Figure 3. After 2 years, improved mineralization in the distraction region (marked with arrows).

frame removal. These problems were, no doubt, exacerbated by attempting simultaneous correction of the major deformities, but the overall result was satisfactory after 2 years.

In conclusion, our experience suggests that surgeons need not fear defective mineralization in callus distraction for Ollier's disease; rather, bone formation improved the local deficiency of bone mineralization.

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