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Case Reports and Series

Retrograde Intramedullary Nailing for Recurrent Fracture in Congenital Pseudarthrosis of the Tibia

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ABSTRACT

We present the case of a 23-year-old male with congenital pseudarthrosis of the tibia, who had undergone treatment with Ilizarov ring fixation and had experienced 4 episodes of repeat fracture. He had associated type 1 neurofibromatosis, and his radiographs confirmed a type 6 Boyd's congenital pseudarthrosis of the left tibia, with concomitant arthritic ankle and subtalar joints. He was treated successfully with retrograde intramedullary nailing of the tibia and autologous bone grafting. At his final follow-up visit at 3 years postoperatively, he displayed complete union with no repeat fractures.

Congenital pseudarthrosis of the tibia (CPT) is a difficult and rare condition to treat, with a reported incidence ranging from 1 in 28,000 in Finland to 1 in 190,000 in the United Kingdom (1). The final outcome of treatment of CPT is complicated by recalcitrant nonunion, repeat fracture, limb length discrepancy, and residual angular and rotational deformities in the involved leg and ankle (2). CPT was first described by Paget in 1891 and has been shown to be associated with neurofibromatosis type 1 in approximately 50% of cases (3).

The preferred techniques for the surgical treatment of CPT in the pediatric age group are intramedullary rods or nails, vascularized fibular grafting (4), and Ilizarov ring fixation (5), or a combination of Ilizarov and intramedullary rod fixation (6). Ilizarov ring fixation has been shown to be the most effective treatment option (6,7). The incidence of union has ranged from 61% to 100% using Ilizarov ring fixation, and the overall incidence of complications has ranged from 31% to 100% (7). In recalcitrant cases of CPT, many investigators have advocated amputation of the involved limb, which occurs in 9% to 30% of cases (8).

Few investigators have discussed the functional outcome of these patients as they grow into adulthood. Patients in whom the fracture site has united often have residual deformities of the tibia and ankle, as well as shortening. Many of these patients develop progression of the deformity, leading to degenerative arthritis of the ankle and knee joints (9,10). Although the incidence of repeat fracture has been shown to be as high as 47% (11), few published data are available regarding the incidence and management of repeat fracture in adulthood. In 1 case report, the investigators described the use of an antegrade intramedullary nail for a patient with CPT who had fractured his leg in adulthood (12).

In the present report, we describe the case of a patient with CPT treated using Ilizarov ring fixation from the age of 10 years. His left tibia had united and refractured 4 times, the last time in adulthood. We treated this patient successfully using a retrograde humeral intramedullary nail. We present our rationale for this treatment and discuss the treatment options we considered.

Case Report

A 23-year-old male presented with a history of recurrent fractures of his left tibia in June 2011. He had been treated from the age of 10 years with an Ilizarov ring fixator for congenital pseudarthrosis of the left tibia. He also had a history of neurofibromatosis type 1 with multiple cutaneous neurofibromas and café au lait patches and axillary freckling (Fig. 1). He had no other significant medical history. He had undergone 4 applications of ring fixation, once to lengthen his left tibia and 3 times for repair of the pseudarthrosis, which healed each time, only to subsequently refracture. On examination, he displayed multiple cutaneous nodules indicative of classic neurofibromatosis, and he displayed skeletal involvement localized to his left tibia. The left leg was hypoplastic with the scars of previous surgery seen on the distal third of his leg, and abnormal mobility was evident in the distal third of tibia, with a stiff ankle and subtalar joint. Radiographs revealed an atrophic type of nonunion of the distal fourth of tibia with a tapering, dysplastic fibula. The ankle joint space and subtalar joint space were diminished (Fig. 2). Type 6 Boyd's CPT was diagnosed (13).

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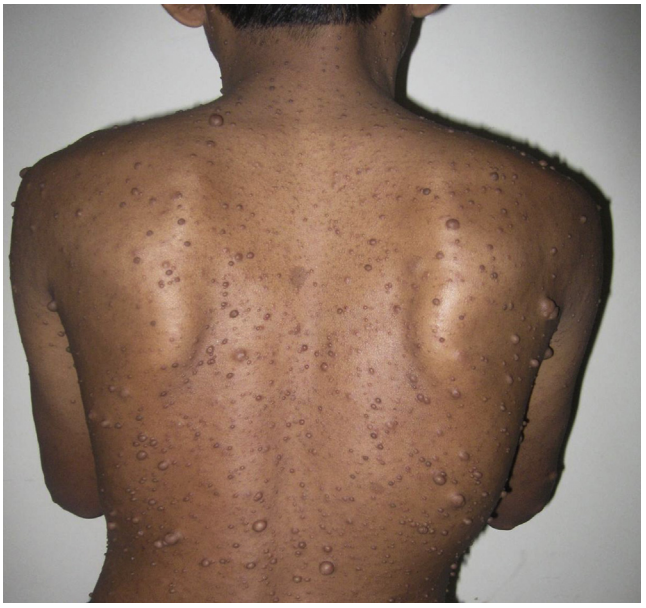

Fig. 1. View of the patient showing multiple cutaneous neurofibromas.

After considering the treatment options, we decided to treat the fracture with a retrograde intramedullary nail. The ankle joint had fused, and the subtalar joint was in fibrous ankylosis and therefore, for all practical purposes, clinically fused. A retrograde nail, transfixing the entire limb from the heel to the proximal tibia, would stabilize the fracture, reduce the stress on the fracture site from the stiff ankle and subtalar joints, and clinically fuse these joints. Open arthrodesis of the subtalar joint was not performed, because it would have increased the morbidity of the procedure, and retrograde nailing of the subtalar joint has been shown to be an effective procedure for subtalar fusion (14).

Surgical Procedure

With the patient under spinal anesthesia, he was positioned supine on a radiolucent table. Under tourniquet control, the fracture site was opened through the previous incision. The fracture ends were mobilized, and the intervening fibrous tissue was removed. Under image intensifier guidance, through an incision made in the heel, an entry was made in the calcaneum with a 3.2 AO drill bit. We directed the drill retrograde through the calcaneum and the talus into the distal tibial fragment. This entry was then widened using a 4.5-mm Steinmann pin. An olive-tipped guide pin was then passed retrograde from the calcaneum to the distal tibial fragment and under vision through the proximal tibial fragment to the proximal fifth of the tibia. Cannulated flexible intramedullary reamers were then introduced along the guidewire, and the tibial canal was reamed up to 11 mm, after which a 10-mm × 315-mm humeral nail (Sharma Surgical & Engineering, Pvt., Ltd., Vadodara, India) was passed over the guidewire. The guidewire was then removed, and the nail was positioned such that the proximal interlocks of the nail passed through the calcaneum, talus, and distal tibial fragment. The 3 proximal

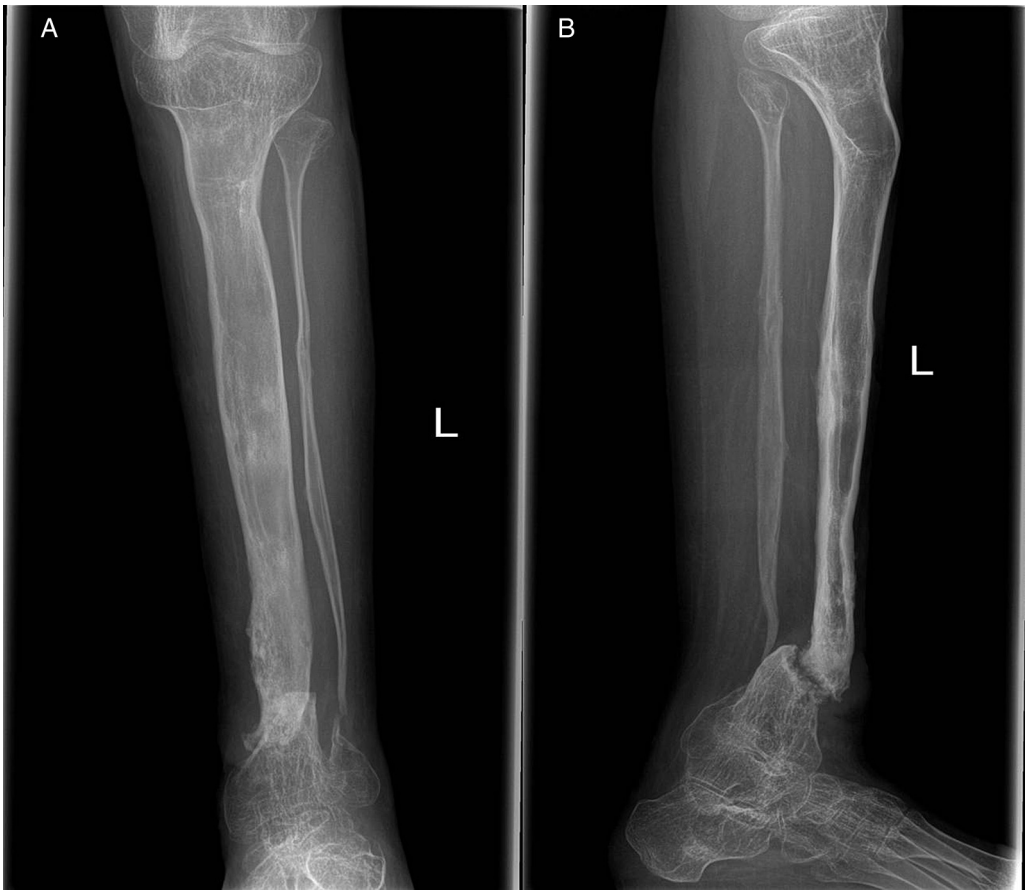

Fig. 2. (*A*) Preoperative radiograph of left lower extremity in the anteroposterior view showing atrophic nonunion of the tibia and tapering dysplastic fibula. (*B*) Preoperative lateral radiograph showing the arthritic ankle and subtalar joint.