

Late Onset of a Congenital Pseudarthrosis of Both the Forearm Bones in an 8-Year-Old Girl

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Abstract

Introduction: Congenital pseudarthrosis of the radius or ulna is rare and less common than congenital pseudarthrosis of the tibia. It may lead to deformity, pain and functional impairment.

Objectives: Remind that conventional fracture treatment as non-surgical treatment or open reduction and internal fixation is not successful in congenital pseudarthrosis.

Case: A missed diagnosis of an 8-year-old girl with late onset pseudarthrosis of both radius and ulna associated with neurofibromatosis is presented. In this case, inadequate initial treatment led to poor results. The use of free vascularized fibular grafts is known to be effective to treat this condition but has some local and lower limb morbidity. Final success was obtained with large resection and bone transport.

Conclusion: It is important to recognize the entity to avoid failure of treatment.

Keywords: Congenital pseudarthrosis; Neurofibromatosis; Ulnar pseudarthrosis; Radius pseudarthrosis

Introduction

Congenital pseudarthrosis may be present at birth but in most cases, they are developmental conditions, and are often seen in association with neurofibromatosis. Indeed, about half of the patients with congenital pseudarthrosis have neurofibromatosis [1-4]. At birth, a deformity is recognized in less than one third of the patient. Most deformities are discovered after a relatively trivial traumatic episode. The term "congenital" may be therefore inappropriate and the term "dysplasia" may be preferred. There is however no question that the underlying dysplasia process is usually present at birth. Before the first fracture occurs, there are sometimes no clinical signs nor symptoms and the disease may be overlooked. In case of misdiagnosis, inadequate treatment of the fracture could lead to failure. The treatment of congenital pseudarthrosis is one of the most challenging problems in all of orthopedics because of the difficulty of achieving and maintaining union.

Congenital pseudarthrosis of tibia is much more frequent and easier to treat than pseudarthrosis of forearm [1,5,6]. Only 72 cases of congenital pseudarthrosis of forearm bones have been reported in the English literature. In the leg, the pseudarthrosis is more commonly found in the tibia, while in the forearm, the radius and the ulna seem to be affected at similar rates [5]. Involvement of both bones is extremely rare [7]. Many treatments had been used in the past, often with poor results, until the advent of free vascularized fibular grafting first described in 1981 [8] and becoming more and more popular [1,5,9-13].

The aim of this case report is to remind to the reader to wonder if an apparent simple fracture in childhood does not heal properly, because when the diagnosis of congenital dysplasia is initially missed, inadequate treatment could lead to poor results.

Case Report

An 8-year-old girl was admitted in an emergency room of a level 2 center (center number 2) for an apparently simple fracture of the distal metaphysis of both right radius and ulna (Figure 1). An orthopedic treatment was first applied with a 6-week long arm cast immobilization after what apparent consolidation of the radius only was obtained with 30   dorsal angulation (Figure 2) and cast was removed with restricted

activity consigns. Ten weeks after the fracture, the patient came back with a painful deformed wrist without traumatism, the radiographs showed a secondary displacement of 60   of both bones (Figure 3). Elastic stable intramedullary nailing was then performed (Figure 4). The fracture evolved in 3 months to an atrophic pseudarthrosis of ulna whereas the radius seemed to be united (Figure 5). Seven month after the initial fracture, a plate osteosynthesis of ulna was then performed



Figure 1: 8-year-old girl admitted in an emergency room for fracture of the distal metaphysis of both the forearm bones.

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with an iliac bone autograft, but the pseudarthrosis persisted on the ulna and moreover, bone progressively became atrophic and sclerotic around radial fracture (Figure 6).

The patient was transferred to the university hospital (level 1



Figure 2: 6 weeks after the fracture, consolidation of the radius with 30° dorsal angulation.



Figure 3: 10 weeks after the fracture, secondary displacement of 60° without traumatism.



Figure 4: Elastic stable intramedullary nailing was performed 10 weeks after the fracture.



Figure 5: 5 months after the fracture, consolidation of the radius and atrophic pseudarthrosis of the ulna.



Figure 6: 12 months after initial fracture, persisting pseudarthrosis of the ulna despite iliac bone autograft, concomitant evolution to pseudarthrosis of the radius.

center). The diagnosis of type 1-neurofibromatosis was given. Six brown “café au lait” spots were present as well as axillary and groin lentigines. Small neurofibromas were also present in the back. She presented also a scoliotic attitude. The girl’s father was also affected by type 1-neurofibromatosis associated with a epidermodysplasia verruciformis. A new surgery was performed 17-month after the inaugural fracture. Surgery consisted of large resection of the ulnar pseudarthrosis area and a bone transport was performed from the proximal ulna with an external fixator. The nail was removed from the radius and a plate-osteosynthesis was performed (Figure 7). Two month later (after the completion of ulnar bone transport), osteosynthesis was performed with a plate on the ulna with a tricortical iliac bone graft. Finally union was achieved six month after the last procedure (Figure 8).

Discussion

Congenital pseudarthrosis may be localized anywhere within the involved bone (proximal, midshaft or distal) [7]. Ulna pseudarthrosis may cause bowing of the forearm, dislocation of the radial head, distal fragment hypoplasia, and lead to functional limitations [1]. The treatment of the congenital pseudarthrosis of ulna has three goals [1]. The first one is to establish bony union. Different treatments are



Figure 7: 17 months after the fracture, large resection of the ulnar pseudarthrosis, bone transport of the proximal ulna and plating of the distal radius.



Figure 8: 6 month after the plating (25 month after the fracture) healing is obtained.

frequently used in congenital pseudarthrosis: cast immobilization, internal fixation, pulse electromagnetic field stimulation, simple pseudarthrosis excision, allograft or autogenous bone grafting. Union is very difficult to obtain and to maintain after a fracture, only one case has been reported to heal by cast immobilization alone [14]. Treating the pseudarthrosis like a fracture in normal bone (internal fixation with intramedullary nailing or plate and screw) leads to poor results as well as correctives osteotomy and nonvascularized bone grafting, while promising results have been reported by utilizing free vascularized fibular grafts [1,5,7,9-13]. Results obtained without vascularized graft could be primary favorable (correct alignment, even sign of callus formation), but bone around the fracture progressively resorbs, becoming atrophic and sclerotic. Hvid and et al. showed that the fibular graft appears to be more effective than the Ilizarov treatment in the congenital pseudarthrosis of both forearm bones [2]. Indeed internal fixation combined with vascularized fibular graft provides a structural stability. The graft is able to bridge large gaps created by wide excision of atrophic bone and abnormal neurofibromatous tissue. The bony healing is so more rapid and complete [1,5].

The second goal is to stabilize the adjacent Distal Radioulnar Joint (DRUJ) and ulnocarpal joint. If the pseudarthrosis was not treated

rapidly, the forearm might have been deformed, resulting in a DRUJ instability or a radial head dislocation. Thus, during free vascularized fibula grafting, the ulna alignment must be preserved to obtain a stable radioulnar and radiocapitellar articulation.

The third goal is to allow for continued skeletal growth. It is difficult to obtain stable internal fixation in proximal of distal pseudarthrosis. In these sites free vascularized fibular graft was used with an epiphyseal transfer.

Free vascularized fibular graft, particularly the step of epiphyseal transfer with its small vessels if needed, is technically difficult and has to be performed by an experienced surgeon. Furthermore the procedure has some possible complication: injury to peroneal nerve, lower extremity vascular complication, valgus deformity of the ankle even with distal tibiofibular arthrodesis, forearm growth disturbance and resorption of graft and recurrent pseudarthrosis [1,5,12,15]. Bae et al. [1] explain that wide debridement of pseudarthrosis tissue with stable internal fixation and accurate microvascular reconstruction allow to avoid this last complication [1].

Bone transport with external fixator is well described and widely used for congenital pseudarthrosis of the tibia [16-20], but it is considered to be less effective than vascularized graft for the treatment of pseudarthrosis of the forearm [2]. However, in this case, the proximity of the ulnar epiphysis led us to try bone transport in order to minimize possible local complications and avoid lower limb morbidity.

In conclusion, congenital pseudarthrosis of radius and ulna is a very rare entity that must not be treated as a simple fracture. Free vascularized fibular graft is an effective surgical procedure when it is performed by an experienced surgeon with stable internal fixation, meticulous microvascular reconstruction and postoperative immobilization. Fibular epiphyseal transfer with careful TFCC and DRUJ reconstruction in patients with skeletally immature and distal ulna pseudarthrosis allows to obtain bony healing, ligamentous stability and continued skeletal growth.

However, bone transport with external fixator is also a valid less invasive option.

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