Congenital forearm pseudarthrosis associated with dislocation of the radial head: surgical treatment with one-bone forearm

Victoria Allende, J. Javier Masquijo

Pediatric Orthopedics Department, Sanatorio Allende, Córdoba

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Abstract

Congenital forerarm non-union (pseudarthrosis) is a rare entity usually associated with neurofibromatosis type 1. The injury of both bones of the forearm associated with dislocation of the radial head is extremely infrequent. We present the case of an 8-year girl with neurofibromatotis type 1 and non-union in both bones of the forearm associated with dislocation of the radial head, who was subject to resection of the ulnar head, wide resection of the non-union and reconstruction by the one-bone forearm technique with plate. The first surgery failed and it was necessary to carry out revision with removal of the implant, autologous bone graft and fixation with intramedullary nail. After the second surgery, the girl had stable one-bone forearm with shortening of 7 cm. At last follow-up, 4 years later, she does not have symptoms and uses her upper limb with minimal limitation in wrist flexion-extension. Reconstruction by one-arm forearm is a reasonable option for non-union in both bones of the forearm with dislocation of the radial head in children. It is a rescue procedure which brings about a stable upper limb with satisfactory function in wrist and elbow.

Key words: Congenital; non-union (pseudarthrosis); children; one-bone forearm.
Level of evidence: IV

Seudoartrosis congénita de antebrazo asociada a luxación de radio proximal: tratamiento quirúrgico mediante antebrazo de un solo hueso

Resumen

La seudoartrosis congénita de antebrazo es una entidad clínica rara que se asocia a habitualmente a neurofibromatosis de tipo 1. La lesión de ambos huesos del antebrazo asociada con luxación de la cabeza del radio es extremadamente inusual. Se presenta el caso de una niña de 8 años con neurofibromatosis de tipo 1 y seudoartrosis de ambos huesos del antebrazo asociada a luxación de la cabeza del radio, que fue sometida a resección del cúbito distal, escisión amplia de la seudoartrosis y reconstrucción mediante antebrazo de un hueso con placa. La primera cirugía fracasó y fue necesaria una revisión con retiro del implante, injerto óseo autólogo y fijación con enclavado endomedular. Tras la segunda cirugía, la niña tenía un antebrazo de un hueso estable, con un acortamiento de 7 cm. En el último control, 4 años después, no tiene síntomas y utiliza el miembro superior con una mínima limitación de la flexo-extensión de la muñeca. La reconstrucción mediante

Conflict of interests: The authors have reported none.

antebrazo de un hueso es una opción razonable para la seudoartrosis de ambos huesos del antebrazo con luxación de la cabeza radial en niños. El procedimiento de salvataje produce un miembro superior estable, con una función satisfactoria de la muñeca y el codo.

Palabras clave: Congénita; seudoartrosis; niños; antebrazo de un hueso.
Nivel de Evidencia: IV

Case

We report the case of an 8-year old girl with diagnosis of neurofibromatosis type 1. The patient suffered a forearm fracture at 6 years of age and was treated with brachio-palmar cast (Figure 1). Two years later, she was referred to us for non-union (pseudarthrosis). At physical examination, we detected severe mental retardation and ulnar-deviated forearm with limitations in elbow flexion-extension. In X-rays we verified ulnar and radial non-union, with anterior dislocation of the radial head and considerable atrophy in the distal end of the ulna (Figure 2).

Due to the permanent dislocation of the radial head and the remarkable atrophy in the distal end of the ulna, we decided as initial treatment reconstruction by the one-bone forearm technique, fixing the distal radius to the proximal ulna, using a radial dyaphisis fragment (whose medical and radiographic looks were normal) as free bone graft, and stabilizing with a 3.5mm-locking plate (Figure 3).

Eight months after the surgery, we found re-absorption of the bone graft and plate loosening; therefore, we decided to carry out revision with removal of the osteosynthesis material, free auologous fibular bone graft and

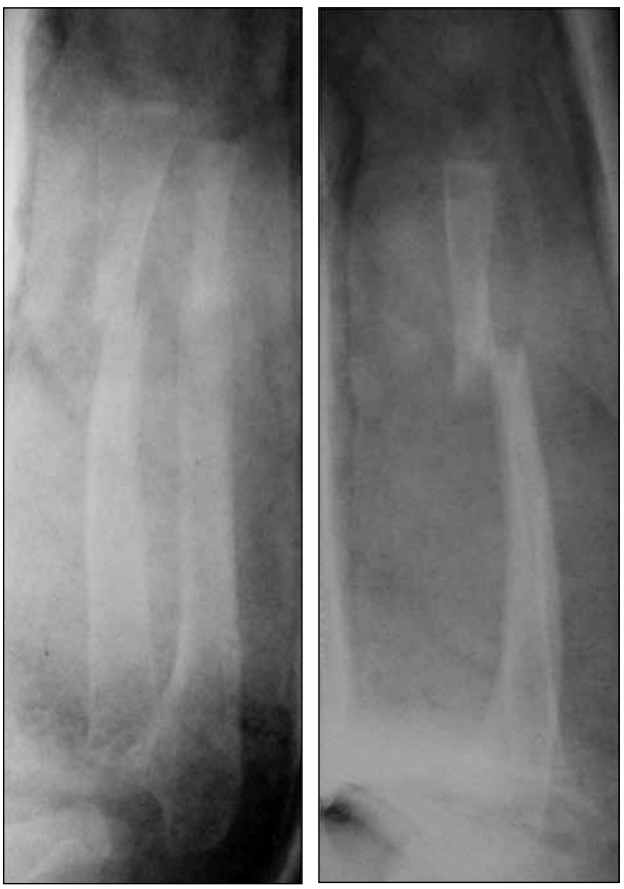

Figure 1. Initial X-rays at the time of the fracture.

Figure 2. X-ray taken in consultation with us.
It shows non-union in both forearm bones with radial head dislocation.

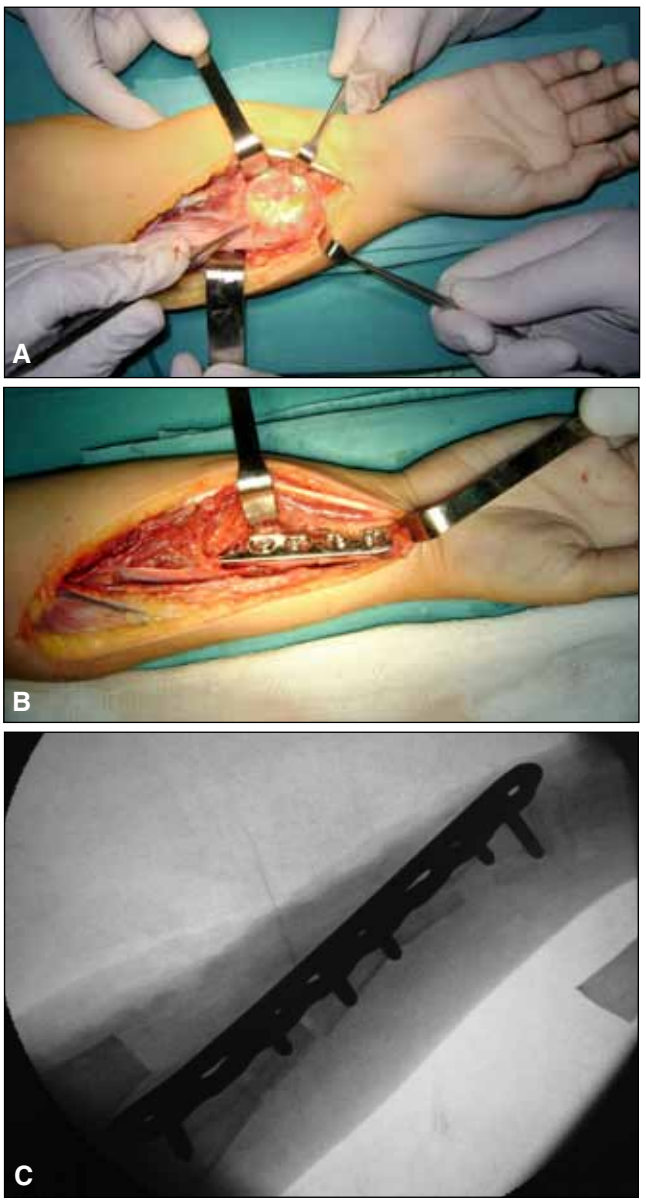

Figure 3. First surgery. **A** and **B** Resection of the injury by Henry approach. **C.** Intra-operative X-ray showing one-bone forearm and fixation with plate.

stabilization with elastic intramedullary nail and wire loop (Figure 4). The patient was subject to immobilization with brachio-palmar cast due to poor collaboration. Six months after the second surgery, we verified graft consolidation, and we removed the intramedullary nail. At last follow-up, four years later, the patient has no symptoms and uses her upper limb with minimal limitation in wrist flexion-extension (Figure 5).

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Figure 4. A and **B.** Post-operative X-ray after plate removal, intramedullary nail and autologous fibular graft. **C.** X-rays at 4th year follow-up: Non-union consolidation.

Figure 5. Medical status and range of motion.

Discussion

Congenital forearm non-union is a rare entity, which in 40-55% of the cases is associated with neurofibromatosis type 1,[1,2] also known as Von Recklinghausen disease; its prevalence is 1/3500 and it shows autosomal dominant inheritance pattern.[3] It is characterized by neurofibromas, *café-au-lait* skin spots, Lisch nodules and skeletal findings such as macrocephaly, short stature, kypho-scoliosis and congenital bowing or congenital non-union.[3,4] The tibial bone is the most frequently affected in this condition, the forearm is more rarely affected.

When the forearm is affected, usually the involved bone is the ulna, followed by the radius and sometimes the two bones. Up till today there have been reports on 14 cases of non-union in the two forearm bones[5-13], and only three of them showed associated dislocation of the radial head.[9-11]

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There are multiple treatments described for this conditions: immobilization with cast, corrective osteotomy, internal fixation with intramedullary nails or plates, free cortical-spongy graft, and fibular vascularized bone graft.[14,15] Fibular vascularized bone graft has been the most frequently used type of graft in the cases with involvement of the two forearm bones. When the vascularized graft is combined with stable internal fixation, it gives immediate structural integrity to the receptor zone, and allows the surgeon to fill large defects created by the resection of abnormal tissues and atrophic bone. The immediate blood supply brings about primary endosteal bone healing and faster consolidation. Although it also causes some complications and requires a surgical team with microsurgery background, it is probably the best therapeutic alternative when the two bones in the forearm are involved.[16]

The involvement of the two bones of the forearm associated with dislocation of the radial head results in a remarkable deformity in the forearm with severe functional deficit.[9-11] The bone defect and the fibrous retraction caused by the non-union bring about progressive dorsolateral displacement of the radial head until dislocation.[12] In this hardly frequent scenario, the main goals are bone consolidation, distal radio-ulnar joint stability, and the growth of the remaining bone. Reconstruction by the procedure of one-bone forearm is a rescue therapeutic alternative that allows the surgeon to reach such goals. This procedure was described by Hey-Groves in 1921[17] and its main indications are post-traumatic sequela, tumor resection and congenital forearm deformities. This technique can be carried out with vascularized[11] or non-vascularized[9] grafts. Vascularized grafts are associated with many biological advantages, but technically they are more challenging. In this case, we first used a bone graft taken from a part of the radial diaphysis and gave stability with a 3.5mm-locking plate. We believe that the main reason for failure in our first surgery was the quality of the bone graft that, even with normal medical and radiographic looks, may have had anomalies invisible to the naked eye. On the other hand, fixation may have been too rigid, what prevented bone from consolidation. Although several authors recommend plate fixation,[16] the angular stability given by the locking screws fixed to the plate might be too much.

Since it has only one bone, the forearm does not act as a joint anymore, and the relative stability given by the intramedullary nail might be ideal, allowing micro-mobility at the level of the fusion between the graft and the bone, thus promoting the formation of the secondary bone callus. Therefore, we decided to carry out revision with free autologous fibular bone graft and give stabilization with an intramedullary nail which, at long last, brought about consolidation.

Reconstruction by one-bone forearm is a reasonable therapeutic option for non-union in both bones of the forearm with radial head dislocation in children. This rescue procedure results in a stable upper limb, with satisfactory function in wrist and elbow.

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