Open reduction intramedullary rod with synthetic bone graft for the treatment of radius ulna pseudoarthrosis with neurofibromatosis type 1: a case report

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ABSTRACT

Background: Pseudoarthrosis is a false joint that occurs in cases of non-union and is commonly associated with neurofibromatosis. Treatment of refracture and non-union remains a challenge, and many methods have been attempted with varying results.

Case Presentation: A 7-year-old, left-handed male sought consult at the Philippine Orthopedic Center (POC) last February 2016 with a complaint of left forearm deformity. This patient was diagnosed with neurofibromatosis type 1 with pseudoarthrosis of the radius and ulna. The pseudoarthrosis was treated with an application of an intramedullary rod and synthetic bone graft. Union and a full range of motion of the forearm were achieved at 1.5 years post-op.

Conclusion: Pseudoarthrosis is a rare case of non-union that occurs spontaneously or after a minor trauma, most commonly associated with Neurofibromatosis Type 1. Since the condition is so rare, many methods have been attempted with varying results. Successful treatment of most atrophic non-union is achieved by optimizing the mechanical and biological environment of the pseudoarthrosis site.

Keywords: neurofibromatosis type 1, pseudoarthrosis, radius, ulna.


INTRODUCTION

Pseudoarthrosis is a rare case of non-union that occurs spontaneously or after minor trauma. The reported incidence varies between 1:140,000 and 1:250,000 for congenital pseudoarthrosis of the Tibia (CPT).¹¹ Five-five percent of which is associated with Neurofibromatosis (NF) Type 1, fifteen percent with idiopathic.¹¹,¹² The osseous dysplasia leads to a tibial non-union because of bowing and reduced growth on tibial epiphysis. Shortening of the limb usually occurs.⁷

The pathologic tissue which is found in the non-union site has osteoblast and osteoclast cells, but the osteolytic component was found to be more predominant. The recurrence of this tissue was reported to cause non-union and refracture. These cells are abundant in the young child due to a greater growth rate and metabolism. This incidence disappears at skeletal maturity.¹¹,¹²

Limb salvage operations of CPT are the most challenging treatment in Orthopedics because of the risk of refracture and non-union. There is also associated difficulty in stabilizing bone fragments in children. Due to the risk of multiple failures, amputation becomes the recommendation after a third failed surgery.⁵,⁶

The forearm is a common site of pseudoarthrosis in NF. It commonly involves the ulna, followed by the radius and both bones. The difficulty in either location is the varying growth potential of each bone. Unequal growth of these bones can develop deformity. The radial head subluxated dorsolaterally as a consequence of non-union of the ulna.²

There are various approaches described by many authors to achieve bone union and prevent progressive deformity and angulation. A study using an intramedullary rod and autogenous bone graft in six patients, showed bony union in all patients.⁷ In another study done by Charles et al., a vascularized bone graft was used in pseudoarthrosis of the forearm using a vascularized bone graft. They argued that conventional bone grafts rarely showed complete union.⁷

In our case, we have a pseudoarthrosis of both the radius and ulna with severe deformity. We performed an open reduction and internal fixation using an intramedullary rod with synthetic bone graft augmentation. The procedure achieved fracture union with a full range of motion. Herein, we report this case with a literature review.

CASE PRESENTATION

A 7-year-old, left-handed male sought consult at Philippine Orthopedic
Center (POC) last February 2016 with a complaint of left forearm deformity. He was previously diagnosed with a closed fracture of the radius-ulna, left, which was treated with closed reduction and casting last April 2012. At 1-year post injury (March 2013), due to non-union, the patient underwent open osteoclasis with an intramedullary rod on both the radius and ulna. Removal of the implant was done at 3 months post-operation.

At 1 year post-operation (October 2014), with the persistence of left forearm deformity, the patient came back for a follow-up. At this time, the patient was diagnosed to have a non-union of the third middle radius ulna left, secondary to neurofibromatosis. The patient was advised for surgery at this time; however, the patient was lost to follow-up. Three years post-operation (January 2016), upon thorough physical examination, there was a note of hyperpigmented patches (café au lait spots) over his face, trunk, axilla, and extremities. A shortening of 6 cm and varus deformity was noted on his affected forearm. On X-rays, we noted a fracture non-union of both the radius and ulna left. The patient was diagnosed to have congenital pseudoarthrosis of the third middle radius ulna secondary to NF type I.

Both the radius and ulna have radially deviated, with 110 degrees for the radius and 120 degrees for the ulna. Apex volar was also noted to be 85 degrees for the radius and 130 degrees for the ulna. Limited range of motion with 0 degrees of supination and 20 degrees of pronation (Figures 1 and 2).

Management and Outcome

On March 23, 2016, the patient underwent open reduction intramedullary nailing with synthetic bone grafting. The volar incision along the previous scar was made. Intraoperatively, there was a note of abundant fibrous tissue over the fracture gap of both bones. Fracture edges were freshened, reduced, and stabilized with an intramedullary rod. The synthetic bone graft was added between the fracture gap (Figure 3). The patient was discharged 3 days post-operation. Sutures were removed at 2 weeks post-operation. The patient was allowed to do flexion and extension of the elbow but no rotation of the forearm during this time.

At 2.5 months post operation, removal of the ulnar rod was done due to the signs of pin tract infection. Only the radial nail was retained. At 1-year post-operation, the removal of the radial rod was done. The patient then followed up every 3 months. At 1.5 years post operation, fracture union was noted at the radius; however, ulnar fracture showed signs of non-union. The patient can do a full range of motion, although the radial head remained dislocated during forearm rotation due to the deformity of the ulna. The limb length discrepancy is 3cm (18/15cm) (Figures 4 and 5).

**DISCUSSION**

Pseudoarthrosis is defined as a “false joint”. In CPT, the affected tibia exhibits insufficient mechanical strength and osteogenetic capability. Some authors found that pseudoarthrosis was associated with neurofibromatosis. The prevalence ranges from 40-80%, but only 1-4% of pseudoarthrosis is associated with neurofibromatosis. Others reported cases that did not find any microscopic evidence of neurofibromatosis at the non-union site.1

Fibrous hamartoma cells were found to be the main pathology of CPT, which occupies a non-union site. These cells fill...
the space between the fragment and do not undergo osteoblastic differentiation.\textsuperscript{1,9,10} Lee found that deficiency of local Mnt ligands caused the disturbance in osteoblastic differentiation.\textsuperscript{10} Cho reported that BMP treatment did not enhance osteoblastic differentiation, which many authors claimed to be effective.\textsuperscript{9,11} Excision of hamartomatous fibrous tissue around the pseudoarthrosis site during surgery is usually emphasized.

Shah \textit{et al.} reported a long term follow-up in CPT, which was treated with intramedullary rod and cortical bone grafting. This report showed that bony union was achieved in 9 out of 11 patients.\textsuperscript{5} A study on congenital pseudoarthrosis of the forearm, reported by Charles \textit{et al.}, showed a complete fusion with vascularized graft.\textsuperscript{2} On the other hand, a study by Ohnishi \textit{et al.} showed that bone grafting is rarely used as long as good bone contact is achieved during surgery.\textsuperscript{11}

CONCLUSION

Pseudoarthrosis is a rare case of non-union that occurs spontaneously or after a minor trauma, most commonly associated with Neurofibromatosis Type 1. Since the condition is so rare, many methods have been attempted with varying results. Successful treatment of most atrophic non-union is achieved by optimizing the mechanical and biological environment of the pseudoarthrosis site. Mechanical modalities include correction of angular deformity and stability of fixation using external and/or internal fixation. In contrast, biological modalities include resection of the fibrous hamartoma and bone graft of the CPT site.

ETHICAL CONSIDERATION

This patient in this study has understood and agreed to the use of patient personal data related to the writing of scientific articles. The patient also gave informed consent consciously regarding the use of the data obtained for the preparation of articles to be published in scientific journals.

CONFLICT OF INTEREST

The authors declare that there is no competing interest regarding the manuscript.

FUNDING

The authors are responsible for the funding of the study without the involvement of grants, scholarships, or any other resource of funding.

AUTHOR CONTRIBUTION

All of the authors equally contributed to the study from the conceptual framework,
data gathering, and data analysis until interpreting the study results.

REFERENCES


