



A case report: Congenital pseudoarthrosis of tibia treated by intramedullary nail and bone grafting

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Abstract

Aim and Objective: To evaluate functional, clinical and radiological outcome achieved by intramedullary nailing and bone grafting in case of congenital pseudoarthrosis of tibia.

Introduction: Congenital pseudoarthrosis of the tibia is a rare disease with reported incidence of approximately 1 in 250000 live births. It is characterized by segmental osseous weakness resulting in deformities of the bone and spontaneous fractures which progresses to a tibial non – union. The etiology is unknown. The goals of surgery are to restore the tibial alignment, achieve bone union and re-establish the length of the tibia.

Materials and Methods: We present a case of a 24 year old female with congenital pseudoarthrosis of left distal third of tibia. Patient was treated with excision of the pseudoarthrosis followed by reduction and fixation using intramedullary expert tibia nail and bone grafting from contralateral ASIS. Radiograph showed union of the tibia at three months follow up. Patient had limb length discrepancy of 3 cm and was ambulating with a shoe raised. The patient was satisfied with her current functional status and had adapted well.

Discussion: Congenital Pseudoarthrosis of the Tibia (CPT) is a rare condition which is characterized by segmental dysplasia of tibial diaphysis resulting in anterolateral bowing of tibia, tibial non-union and reduced growth in the distal tibial epiphysis. The management of CPT still remains a challenging problem. The main goal of surgery is

to achieve bone union, restoring the leg alignment and to preserve function and bone growth. Various methods of treatment include excision of the lesion, followed by stable internal or external fixation with vascularized or non-vascularized graft. The three surgical techniques: intramedullary nailing with a bone graft; vascularized fibular transfer; and the Ilizarov technique were reported with varying results and complications.

Conclusion: Intramedullary nail in combination with bone graft improves the management of complex congenital pseudoarthrosis of tibia. Limb lengthening procedure is feasible at a later stage once union of pseudoarthrosis is achieved. This may obviate the need for treatment using complex external fixator in a highly specialized tertiary centre. However, this method needs more implementation and long term follow up to study the effectiveness and possible complications.

Keywords: Congenital pseudoarthrosis tibia, Ilizarov procedure, osteosynthesis, Intramedullary nailing.

Introduction

An estimated 1 in 250 000 live newborns are found to have congenital pseudoarthrosis of the tibia¹. It is characterised by segmental osseous weakness that leads to skeletal abnormalities and spontaneous fractures, progressing to a tibia that does not fuse. The cause is not known. The objectives of surgery are to accomplish bone union, correct tibia alignment, and restore tibia length.

Case report

In this case study, a female patient, age 24, is shown to have congenital pseudoarthrosis of the left distal third shaft of the tibia. Patient received intramedullary expert tibia nail and bone grafting from the contralateral ASIS after resection of the pseudoarthrosis, reduction, and fixing. At the 3-month follow-up, a radiograph revealed tibial union. Patient was walking with one shoe up and had a 3 cm difference in limb length. The patient had adjusted well and was content with her present functional state.



Preop X-ray



1. Old operated with illizarov 2. One month post OP 3. Two months post OP



4. three months post op

5, 6 post op clinical pictures

Discussion

The rare disorder known as congenital pseudoarthrosis of the tibia (CPT) is characterised by segmental dysplasia of the tibial diaphysis, which leads to tibial non-union, anterolateral bowing of the tibia, and decreased growth in the distal tibial epiphysis. About 50% of CPT cases have neurofibromatosis as their primary diagnosis². A fibrous hamartoma is the term used to describe the aberrant tissue at the location of pseudoarthrosis³. The primary pathological feature in pseudoarthrosis is abnormal periosteal growth, which causes this lesion⁵. This lesion's histological analysis reveals tissue that resembles fibromatosis and is continuous with the bone's periosteum.

The strong osteoclastogenicity and low osteogenicity characteristics of the fibrous hamartoma cells also contribute to the recurrence of pseudoarthrosis. The management of CPT is still a difficult issue. Surgery's primary objectives are to accomplish bone union, correct leg alignment, and maintain function and bone growth.

Excision of the lesion and stable internal or exterior fixation with vascularized or non-vascularized graft are a few of the therapeutic options. The three surgical procedures-vascularized fibular transfer, the Ilizarov technique, and intramedullary nailing with a bone graft-were described with a range of outcomes and problems. Ilizarov procedure

is a useful, safe, and efficient therapy option for atrophic-type CPT management. The Ilizarov procedure, in contrast to other treatment techniques, enables the realisation of numerous aims, including osteosynthesis, ankle stabilisation, and leg-length equality. However, in order to obtain and maintain union while minimising a deformity, to maximise the cross-sectional area of the healing mass, and to stabilise the ankle mortise by fibular fixation, the best surgical intervention based on the fundamental treatment principles is required.

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Three surgical procedures-vascularized free fibular transfer with follow-up through skeletal maturity, the Ilizarov technique, and intramedullary nailing with bone grafting-were observed to have equal results. No one surgical procedure is preferable. During surgery, removing fibromatous tissue and diseased periosteum speeds recovery. It is necessary to plan the amount of soft tissue that will be removed because drastic removal will make it difficult to restore a significant defect and could endanger nearby neurovascular structures. The Ilizarov procedure is the most popular way to treat congenital pseudarthrosis of the tibia, especially when it comes to the dysplastic variety, which necessitates substantial pseudarthrosis excision⁷.

The Ilizarov method uses the callus distraction approach, which has advantages in treating severe bone defects and shortening, and it enables superior bone regeneration. Additionally, with this technique, deformity can be corrected during a single surgery. The Ilizarov procedure, however, has a lengthy recovery period, is somewhat difficult, and runs the risk of wire-tract infections. Another option is a vascularized fibular graft, which enables early tibial consolidation⁸. To obtain adequate tibial alignment, stability, and to prevent refracture, this procedure is commonly used in conjunction with an Ilizarov external fixator or an intramedullary rod. The use of a vascularized fibular graft for CPT treatment was advised in a prior study, which also predicted great long-term results. Refracture and misalignment of the hip (such as anterior bending or valgus deformity) are complications of this procedure.

Conclusion

The treatment of complex congenital pseudarthrosis of the tibia is improved when an intramedullary nail is used in conjunction with a bone graft. The patient's clinical condition had improved after 12 months thanks to intramedullary nailing and bone grafting. Once union of the pseudoarthrosis is achieved, limb lengthening surgery is

possible. This could eliminate the need for complicated external fixator therapy in a tertiary facility with a high level of specialisation. To examine the efficacy and potential drawbacks of this approach, more application and long-term monitoring are required.

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