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Case Report

RESIDUAL NON-UNION IN A CASE OF TWO AND HALF YEAR OLD CHILD CONGENITAL PSEUDOARTHROSIS OF IPSILATERAL TIBIA AND FIBULA TREATED BY INTRAMEDULLARY FIXATION WITH K-WIRE AND ALLOGENIC CANCELLOUS STRUT GRAFT: A CASE REPORT.

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ABSTRACT

The pseudarthrosis usually develops during the first two years of life; however, there are reports of cases in which fractures developed before birth and reports of late-onset pseudarthrosis. There is a strong association between Congenital pseudoarthrosis and Type I neurofibromatosis. CPT develops in about 5.7% of patients with NF-I and 40% of patients with pseudoarthrosis were found to have NF-I. Main pathology is hyperplasia of fibroblasts with the formation of dense fibrous tissue at fracture site causing osteolysis, and persistence of pseudarthrosis. The difficulty in treating this condition occurs because of two factors. 1) Biologic: poor healing ability of the dysplastic segment of bone and 2) Mechanical: technical difficulty to fix small and osteopenic bone fragments in children without damaging the distal physis. A multitude of treatment protocols focusing on stimulating the healing process by using different bone grafting techniques were documented⁶. The graft materials most commonly used included osteoperiosteal graft, massive only graft, autogenous iliac crest bone graft, and vascularised bone graft.

Keywords: Congenital Pseudarthrosis, Tibia, Fibula, Nonunion bone grafting, Vascularised bone graft

INTRODUCTION

Congenital Pseudarthrosis of the tibia and fibula is Congenital Pseudarthrosis of the tibia and fibula is one of the most most perplexing and challenging problems in paediatric orthopaedics because of the difficulty in achieving and maintaining union. It is an uncommon entity with a reported incidence of 1:140,000 to 1:250,000 neonates¹. It refers to nonunion of a fracture that develops spontaneously or after trivial trauma in a dysplastic bone segment of the lower third of

tibial and fibular diaphysis. Congenital pseudarthrosis of the fibula with or without ankle deformity often precedes or accompanies the same condition in the ipsilateral tibia². Sometimes it even develops between the time of successful bone grafting of a pseudarthrosis of the tibia and skeletal maturity². The pseudarthrosis usually develops during the first two years of life; however, there are reports of cases in which fractures developed before birth and reports of

late-onset pseudarthrosis³. There is a strong association between Congenital pseudoarthrosis and Type I neurofibromatosis⁴. CPT develops in about 5.7% of patients with NF-I and 40% of patients with pseudoarthrosis were found to have NF-I^{4,5}. Main pathology is hyperplasia of fibroblasts with the formation of dense fibrous tissue at fracture site causing osteolysis, and persistence of pseudarthrosis⁵.

The difficulty in treating this condition occurs because of two factors. 1) Biologic: poor healing ability of the dysplastic segment of bone and 2) Mechanical: technical difficulty to fix small and osteopenic bone fragments in children without damaging the distal physis. A multitude of treatment protocols focusing on stimulating the healing process by using different bone grafting techniques were documented⁶. The graft materials most commonly used included osteoperiosteal graft, massive onlay graft, autogenous iliac crest bone graft, and vascularised bone graft. The fixation methods also varied widely between cortical fixation using bone or metal plates, intramedullary rods of different types, external skeletal fixation, and lastly combination of external fixation augmented by intramedullary rods⁷. Although Bone morphogenetic proteins

have shown clinical efficacy in the treatment of adult tibial nonunions and in spinal fusion, its logical application in cases of pseudoarthrosis along with corticocancellous allograft has not resulted in enhanced bone healing⁸.

CASE REPORT

We report a two and a half year old female child with history of bowing of right leg since 2 months of age that was gradually increasing and deformity of the lower third of right leg after trivial trauma at the age of 20 months. Child, who used to walk with support from 16 months of age was unable to stand or walk after trauma. Father is a known case of neurofibromatosis I and has multiple neurocutaneous lesions and cafe au lait spots. Child has multiple Caffe au latte spots with no neurocutaneous lesions. Anterior angulation of lower third leg with shortening of the tibial component by 3cm and abnormal mobility in both anteroposterior and mediolateral planes with no stiffness of affected ankle joint were significant findings. [Fig 1]. X ray findings : Atrophic fracture ends of lower third tibia and fibula with obliteration of medullary canal and anterior angulation.



Fig.1: Clinica profile of a two and half year old female child with congenital pseudoartbosis tibia and fibula. **1a:** Clinical Photograph of left leg bowing **1b:**Pre op X ray Left leg with dysplastic fracture ends and obliterated medullary canal of tibia fibula **1c:** MRI showing dysplastic ends and fibrous tissue at fractured ends with decreased signal intensity in the medullary canal at the pseudoarthrosis site suggesting obliteration of the medullary canal **1d & 1e:** showing multiple coffee latte spots over the trunk and abdomen.



Fig.2: Surgical Procedure of resection of pseudoarthrosis and intramedullary fixation with allogeneous cancellous bone graft. **2a.** Intraoperative finding of fibrous tissue around dyplastic tibial ends at the site of pseudoarthrosis of the tibia. **2b.** Allogeneous cancellous bone graft from child's father's Iliac crest. **2c.** Retrograde fixation of graft to resected tibia with a k-wire. **2d.** Intraoperative picture of allogeneous graft fixation with k-wires passed retrogradely. **2e.** C-arm picture showing correction of deformity with fixation of graft in situ. **2f.** Postoperative picture showing deformity correction without limb length discrepancy by ensuring allograft sizing the resected specimen of tibia. **2g.** Postoperative splinting in above knee cast. **2h.** Clinical picture of parent with neurocutaneous lesions over the body showing surgical scar from iliac crest over allograft donation site. **2i.** Postoperative healed surgical scar with k-wires in situ. **2j** –resected gross specimen of sclerotic and dysplastic tibial ends over pseudoarthrosis site. **2k.** Histopathology slide of excised tissue show cellular proliferating fibrous tissue with immature bone is observed (40X)

The child was initially planned for excision of fibrosis and dysplastic ends of the tibia and fixation. The abnormal part of tibia – dysplastic bony fractured ends and fibrous tissue was resected leaving a gap of 5cm. A thick allogeneous

cancellous strut graft from the iliac crest of the child's father after due major cross matching the blood group was taken. The bone gap was reconstructed by passing a 1.5mm K-wire of 30cm length in a retrograde manner under c- arm

guidance from distal tibia through talus, calcaneum to heel. A 1mm k wire is passed to fix the fibula after resecting the fibrous tissue and freshening the edges to provide internal splintage to the leg. The deformity was clinically corrected and there was no limb length discrepancy without neurovascular compromise. Post operatively child was immobilised in an above knee plaster cast.

Suture removal was done after healing of surgical wounds and there was no signs of infection. [Fig 2] Child was followed for up to 3 months, with check radiographs taken every month and there were no clinical signs of infection on inspection after removing the cast. At the end of 3 months, there was no signs of bony union and the allogeneous cancells graft was sclerotic. [Fig 3]

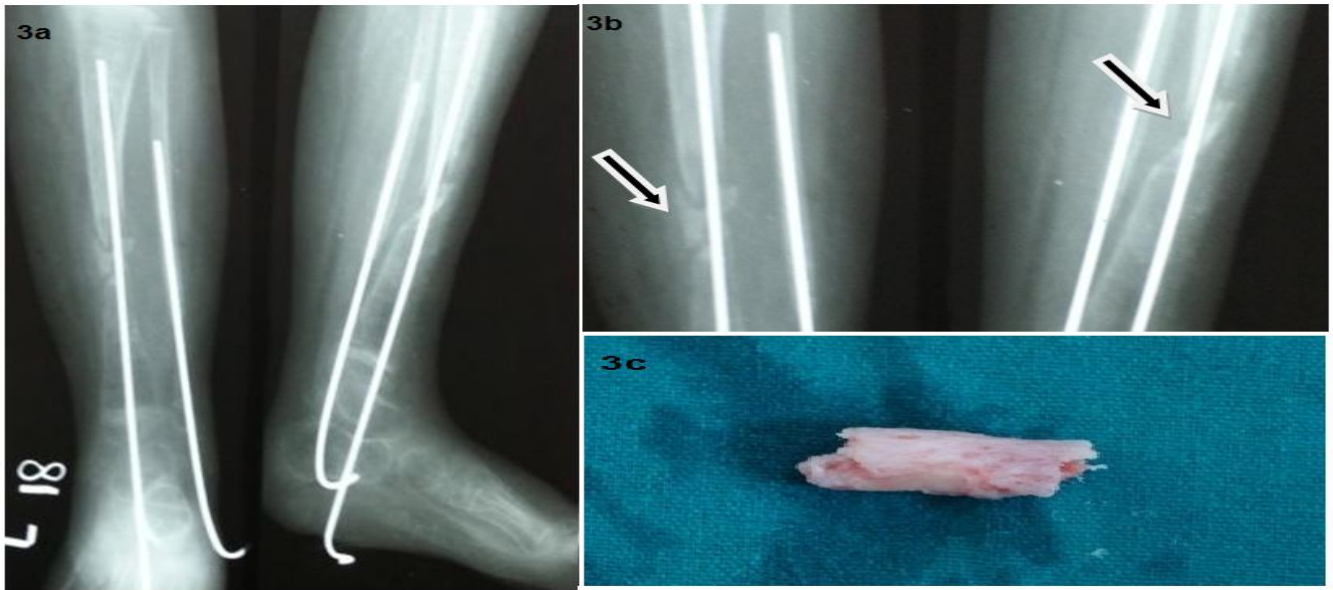


Fig.3: 3months post surgery followup scenario **3a**. X-ray showing alignment with k-wires insitu and no signs of union and sclerosis of allograft **3b**. Arrowmarks in the X-ray pointing to sclerosed and atrophied allograft **3c**. The gross specimen of allograft after 3months.

DISCUSSION

The treatment goal for congenital pseudarthrosis of the tibia is to achieve bony union and avoid recurrent fracture and limb length discrepancy. The pathologic process of the disorder is the growth of abnormal, fibromatosis-like tissue either within the periosteum (dysplastic type) or within the endosteal/marrow tissue (cystic type), or the coexistence of both (mixed type)^{9,10}. Complete excision of the abnormal tissue is necessary to treat the problem, evidenced by visualising normal bleeding and marrow cavity¹¹. Resection of the abnormal tissue, followed by fragment approximation, intramedullary nailing, and bone grafting, is a relatively easy procedure and is suggested as an initial treatment for congenital pseudarthrosis of the tibia¹². This

treatment has a union rate of approximately 86%^{13,14}.

In the present case, after resection of 4cm of dysplastic bone, reconstruction with allogeneous cancellous graft with intramedullary nail was technically challenging because of the small medullary canal of the tibia, which is only 1.5mm diameter at isthmus. In literature, documented intramedullary fixation was done with Titanium Elastic nails, Enders Nails^{7, 15}. Use of external frames such as a Taylor spatial frame, JESS, Ilizarov are reserved usually for older children. In the present case due to non availability of TENS and Enders of diameter of 1.5mm, a K-wire is used as an intramedullary device to provide stability and alignment in this child. Excision and

grafting with vascular fibular grafts or cortical fibular strut grafts are recommended in literature. Vascularised fibular grafts are technically demanding and autologous cortical strut grafts in a child less than 3 years is practically not feasible. Hence, we have tried allogeneous cancellous strut graft from the parent after ensuring major compatibility as a grafting choice coupled with intramedullary k-wire fixation in treating a pseudoarthrosis of the tibia in a child less than 3 years old. Attention to internal fixation and fibular continuity are crucial for maintaining alignment and consequently union¹. Hence, after resection of fibrous tissue around fibular pseudoarthrosis and the excision of periosteum, fibula was fixed with 1 mm k-wire to act as an internal splint and maintain fibular continuity. Toward these goals, immediate contact of bone ends with compression and ample bone graft and biological enhancers are likely to achieve union most frequently. Although similar principles were followed in this case, it resulted in non union.

CONCLUSION

Pseudoarthrosis is both a biological and mechanical issue. Mechanically although stabilised, biological pathology predominated in this case and resulted in failure. In conclusion, there was no predictor identified for nonunion in this study. Hopefully, in the future, combined surgical and medical treatments that effectively modulate or block the biochemical pathways responsible for congenital pseudoarthrosis will downgrade its recalcitrant status.

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