A case of congenital pseudoarthrosis tibia treated by four in one procedure and review of literature

Jain S, Ajmera A, Jain M

Study performed at Department of Orthopaedics, Mahatma Gandhi Memorial Medical College, Indore (M.P.)

Abstract

Case report: Standard methods of treatment for congenital pseudoarthrosis of tibia (CPT) including internal fixation with intramedullary rodding, external fixation or ilizarov fixation, vascularized fibula transfer or combinations have varied results with high rates of recurrence. We report such a rare case of congenital pseudoarthrosis tibia which failed to unite with primary surgery of ilizarov and then was successfully treated and united with four in one procedure which involves creating a synostosis between proximal and distal ends of both tibia and fibula. The procedure is reliable and effective in preventing re-fractures.

Keywords: Congenital pseudoarthrosis tibia, Four in one procedure, Tibia fibula synostosis

Address of correspondence:
DrMitul Jain,
Senior Resident, Department of Orthopaedics, Mahatma Gandhi Memorial Medical College, Indore

Email –mitul.jain007@gmail.com

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Introduction

Congenital pseudoarthrosis of tibia (CPT) is a very rare condition, with incidence of about 1 in 25000 live birth [1]. It is most commonly associated with neurofibromatosis type 1 in more than 50 % of the cases [1]. It has also been associated to fibrous dysplasia or Campanacci's osteofibrous dysplasia [2].

Loss of neurofibromin protein and presence of hamartomas are the primary pathologies in neurofibromatosis that prevents osteoblastic differentiation, bone remodeling or fracture healing. The soft tissue at the pseudarthrotic site is composed of variable admixture of fibrous tissue, fibrocartilage, and hyaline cartilage with evidence of enchondral ossification [3].

Standard methods of treatment for CPT including internal fixation with intramedullary rodding, external fixation or ilizarov fixation, vascularized fibula transfer or combinations have varied results with high rate of recurrence [4-9]. 4 in 1 osteosynthesis used to treat CPT, involves creating a synostosis between proximal and distal ends of both tibia and fibula [10, 11]. We report such a rare case of congenital pseudoarthrosis tibia which failed to unite with primary surgery of ilizarov and then was successfully treated and united with four in one procedure and also reviewed the literature on the topic.

Case report

A 5-year-old female child, presented in our outpatient department, with chief complain of deformity, shortening of right leg and inability to bear weight on affected right leg since birth. The child was delivered at full term with normal vaginal delivery. She had history of full immunization as per the national immunization schedule. Parents and child denied any other significant past medical history. Familial history was unremarkable for any other congenital disorder. As per the parents, some deformity of right leg was noticed during the neonatal period but no treatment was taken. Child achieved all milestones at appropriate age except for ambulation, which was delayed. Child had a

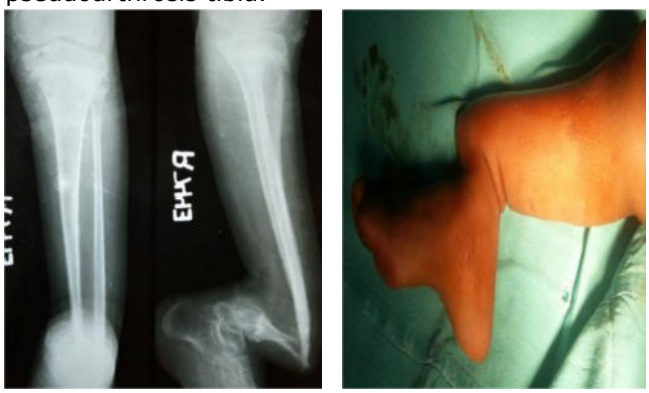
history of trivial trauma due to fall while playing 2 months back and had been non ambulatory since then.

On clinical examination, distal fourth right leg was deformed with severe bowing and procurvatum deformity of about 90^0 (fig 1). There were clear signs of non-union of fracture of tibia and fibula with frank painless abnormal mobility present in all the planes at lower third leg. The leg musculature was severely hypotrophied. Proximal end of bone was dimpling the skin which appeared smooth and shiny. On palpation local temperature was comparable on both sides. Distal pulsations of dorsalis pedis artery and posterior tibial arteries were palpable. Ipsilateral hip and knee examination were unremarkable but active movements of ipsilateral ankle were restricted. Shortening of about 5 cm was present on the right side. No other deformities were seen anywhere else in the body.

Routine radiographs of the limb including AP and lateral views were done which showed pencil thin severely hypotrophied tibia fibula with clear fracture at level of lower third leg. The bone ends were atrophic and thinned out. Severe angular deformity was present at the fracture site and ankle seen in severe calcaneus position. The knee joint appeared normal.

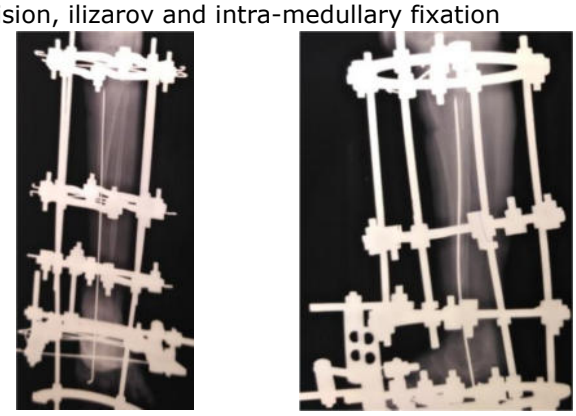
After routine hematological work up and pre anesthetic check-up patient was primarily treated by excision of hamartomatous tissue along with ilizarov application, corticotomy and intramedullary rod fixation to achieve union and prevent refracture (fig 2). After 4 months of bone transport and compression at fracture site, the pseudoarthrosis failed to show signs of union, although good regenerate bone was formed at corticotomy site. Illizarov was removed and patient was planned for re-surgery with four in one osteosynthesis technique to achieve a tibio fibular synostosis (fig 3). Copious amount of cortico-cancellous bone graft was obtained from the ipsilateral iliac crest. Utilizing the previous incision, the pseudoarthrosis site was exposed and remnants of fibrous periosteal and hamartomatous tissue were removed (fig 4). Both ends i.e. proximal and distal ends of the both the bones tibia and fibula were freshened and the copious amount of cortico-cancellous auto-graft was placed between the tibia fibula to achieve a synostosis between tibia and fibula both proximally and distally. An external fixator was applied and trans-calcaneal intramedullary rush nail was also fixed to protect the union. Postoperatively, regular pin tract dressings were done. Patient was followed regularly to confirm an uneventful postoperative period. External fixator was removed after 6 months' post operatively. The Child achieved union at pseudoarthrosis site at 9 months, but had limb length discrepancy and is explained and planned for limb lengthening once she attains maturity (fig 5).

Fig-1. Antero-posterior (a) and lateral (b) X rays and pre-operative clinical photograph (c) of the patient showing typical features of congenital pseudoarthrosis tibia.



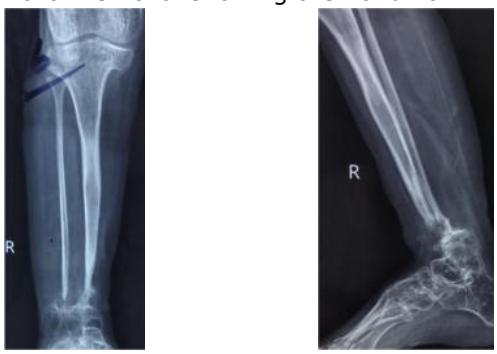
a b c

Fig-2. Antero-posterior (a) and lateral (b) X rays after primary surgery by hamartomatous tissue excision, ilizarov and intra-medullary fixation



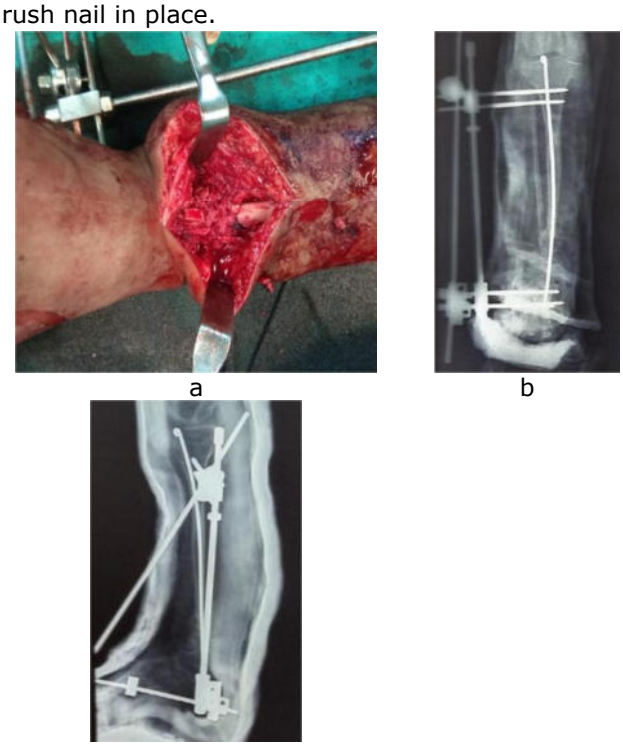
a b

Fig-3. Antero-posterior (a) and lateral (b) X rays after ilizarov removal showing the nonunion.



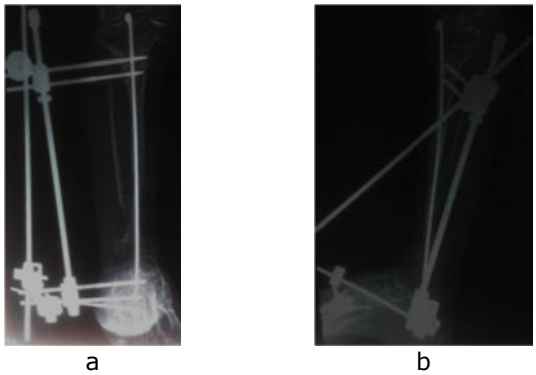
a b

Fig-4. Intra-operative photographs (a), antero-posterior (b) and lateral (c) X rays of 4 in 1 procedure showing cortico-cancellous auto-graft between the tibia fibula to achieve a synostosisand external fixator with trans-calcaneal intramedullary rush nail in place.



a b c

Fig-5. 8 months follow up antero-posterior (b) and lateral (c) X rays after 4 in 1 procedure showing sound union after external fixator removal.



a b

Discussion

Congenital pseudoarthrosis tibia, often associated with neurofibromatosis type 1, is a progressive disorder that has troubled orthopedicians for more than a century [3]. Many theories have been given with regards to pathogenesis of the disease. As per McElvenny's thickened, adherent periosteum which constricts tibia-fibula is the cause of atrophy, fracture and pseudarthrosis [12]. Boyd demonstrated osteolytic fibromatosis as the cause of CPT from a study of amputated specimens [13]. Codivilla was the first to recognize that the periosteum in CPT is diseased and he recommended osteo-periosteal grafting as the treatment [14].

The natural history of CPT is quite unfavorable and results are seldom optimal. The deformity progresses from anterolateral bowing at birth to eventual fracture of tibia fibula which fails to unite. Fracture of tibia lead to absence of weight bearing bone and instability. Fracture of fibula causes proximal migration and valgus at ankle joint. The anterior bow of the tibia causes the foot to assume a severe dorsi-flexed or calcaneus position of the ankle. Due to lack of loading of distal tibia physis the growth rate is reduced causing limb length discrepancy and atrophy of the bones and muscles [13]. Also there is compensatory overgrowth of femur as well as coxa valga that may even cause hip dysplasia.

Many classifications of CPT have been described. Boyd classified CPT into 6 types with type 1 being anterior bowing of tibia, type 2 is the most common type with hourglass constriction, type 3 is cystic lesions of tibia, type 4 is sclerotic lesion with stress fractures, type 5 includes dysplastic fibula and type 6 is intra-osseousneurofibroma [13].Crawford classified CPT into 4 categories. Type 1, non-dysplastic type with anterolateral bowing of tibia with cortical thickening at apex of deformity but medullary canal is preserved; type 2, dysplastic type with anterolateral bowing and narrowing of medullary canal; type 3, cystic lesions present in tibia along with deformity and type 4, pseudoarthrosis [15]. None of these classifications consider the status of the fibula, which plays a role and

also a key factor in the pathology and management of CPT. Choi classification was the first to highlight the important consideration of proximal fibular migration [16]. Although they documented the fibular migration, Choi et al did not recommend any treatment for this.

Child with CPT presents with mild deformity i.e. anterolateral bowing of tibia, which is present since birth. The deformity eventually progresses and results in moderate to severe bowing and atrophy of bone, which ends into fracture of tibia and fibula which fails to unite [1,12]. Foot deformity, leg-length discrepancy (LLD), knee malalignment and even hip dysplasia due to valgus deformity of the proximal femur are myriad of conditions that are all considered part of the natural history of CPT due to involvement of fibula causing valgus [10]. Our patient also presented with similar complains of nonunion of tibia and fibula with severe deformity and hypotrophy of leg musculature and bone along with ankle dorsiflexion deformity.

If the child presents early with just bowing of tibia, then treatment is focused on preventing the fracture using bracing. Adjunctive treatments such as bisphosphonates, Bone Morphogenic Protein (BMP 2, 7) and electric stimulation have also been used [3]. Once the bone has fractured non-union is the rule and surgery remains the only choice of treatment. Primary objective of surgery is to obtain and maintain union at fracture site and secondarily correct the bowing of tibia and prevent proximal fibular migration [10]. The standard methods of treatment for CPT include: internal fixation with intramedullary rodding [4, 5], external fixation (predominantly ilizarov apparatus), combination treatment with an ilizarov and rodding construct [6, 7] or vascularized fibula transfer [8, 9]. Excision of fibrous hamartoma is often used along with these procedures. The results of these procedures are varied with high rate of recurrence of fracture and no consensus exists, on what surgery offers best union rates and least recurrence or on the appropriate age of surgery [10]. We also considered our patients primarily for excision of the hamartomatous tissue and ilizarov, but failed to achieve desired results.

A newer modality of treatment termed as 4 in 1 osteosynthesis as suggested by Choi et al has been used in atrophic type CPT. It involves creating and achieving a synostosis between the proximal and distal ends of tibia and fibula by use of bone grafts. The procedure has yielded favorable results and has been effective in preventing re-fractures in almost 100% patients [10, 11]. By the procedure we could also achieve union and synostosis for the pseudoarthrotic site, although we still are skeptical to remove the intra-meduallary rod and are planning for limb lengthening. Amputation remains the salvage procedure when all the treatment modalities have failed and articular functions are compromised.

Conclusion

Congenital pseudoarthrosis of tibia is a rare pediatric disorder in which fibrous hamartomatous tissue between the bones ends leads to formation of pseudoarthrosis. The entity is difficult to treat with high rate of recurrence with standard treatments. 4 in 1 osteosynthesis which involves creating and achieving a synostosis between the proximal and distal ends of tibia and fibula by use of bone grafts is a reliable and effective procedure in preventing re-fractures.

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