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Free Periosteal Graft with Bone Morphogenic Protein 2 and Internal Fixation combination as Treatment of Congenital Pseudoarthrosis of the tibia

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Abdulwahab Ahmed Alzahrani*

ABSTRACT

Pediatric orthopedics contains various musculoskeletal deformities. Functional distortion, biological and structural abnormalities are the common denominator among congenital or acquired pediatric deformities. Leg bowing, shortening, and catastrophic fracture representing the predominant characteristics of congenital pseudoarthrosis of the tibia in this case report. Plate and screw internal fixation with bone graft augmentation, and complementary pharmacological therapy by zoledronic acid peri-operation are used in the treatment plan after failure of conservative and rigid external fixation trial. An accepted healing, restoration of the mechanical alignment, and minor shortening were the final results of 4 years follow up.

Keywords: Congenital pseudoarthrosis, periosteal graft, Paley X-Union protocol, 2 Bone Morphogenic Protein 2, BMP2.

1. INTRODUCTION

Congenital pseudoarthrosis of the tibia (CPT), a particularly rare pathology, still constitutes one of the most difficult management problems encountering an orthopedic surgeon (Paley, 2019). The documented feature of CPT is stubborn bone atrophy and nonunion, leg deformity, tibial bowing in anterolateral direction, and repeated refracture even following the union is achieved in surgery (Shannon CE et al., 2021; Paley, 2012). Moreover, many linked deformities of length and angulation should be considered in the complete management of CPT. the risk of refracture in these cases is present and extends beyond skeletal maturation and may be presented in adults. The bad prognostic factors include younger treatment age, associated neurofibromatosis, and failure for previous treatment (Paley, 2012). Many management techniques are offered, but the challenge of achieving union and prevention of refracture is still unclear. These techniques range from simple techniques as wearing protective brace and electrostimulation to induce union up to more complicated surgical techniques such as internal and external

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fixation, grafting, and induced membrane techniques (Paley, 2012 ; Vander Have, et al., 2008).

2. CASE PRESENTATION

We report a case of a 2-year-old boy child characterized by Tibia congenital pseudoarthrosis. He had a history of leg bowing since birth and a recent fracture following trivial trauma. Radiologically, it was anterolateral bowing with nonunited fracture at the junction of distal 1/3 and proximal 2/3. Clinically, there were no signs suggesting neurofibromatosis or other congenital diseases. The condition was treated multiple times with different methods over a period of several months, including conservative management, open reduction with bone grafting and external fixation. Finally, A Certified pediatric orthopedic surgeon use Paly protocol treatment with some modification, he treated this patient surgically by a combination of autogenous free periosteal graft, cancellous bone graft, bone morphogenic protein 2 and internal fixation with intramedullary rods, was adopted followed by postoperative bisphosphonate. The fracture healed uneventfully in six months.

Tibia pseudoarthrosis is a difficult disease to manage and treat. With wide options of management approaches and with varying rates of union and refracture. Our patient was treated with this combination and successful union was achieved in six months and satisfactory follow up to 4 years.

3. CASE SUMMARY

A 2-year-old boy referred to our outpatient orthopedic clinic having a fracture right tibia three months earlier that failed to unite with conservative management [Figure 1].

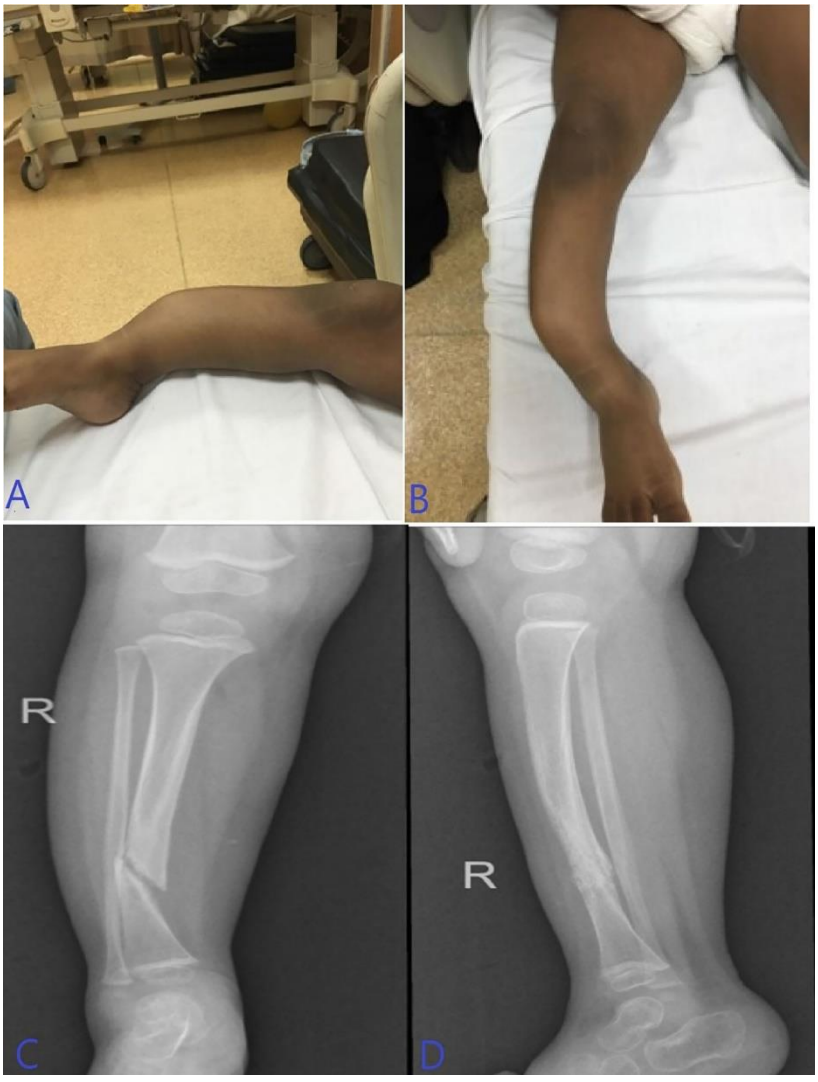

Figure 1: Anteroposterior (AP) and lateral views of the right leg showing tibial anterolateral curvature with Ununited fracture located at the junction of the proximal two thirds and distal one third.

According to the history, the child had anterolateral leg bowing since birth. There was no family history of similar conditions and no history of neurofibromatosis. The general examination was unremarkable. Local inspection revealed anterolateral bowing with an inability to bear weight on that leg. There was no tenderness or signs of infection at the site of deformity, and the deformity was mobile. Laboratory investigations, including inflammatory marks, were unremarkable. A diagnosis of CPT was suspected, the family decided to continue conservative management with above knee serial castings. Follow up in outpatient clinic for ten months showed no improvement, the deformity progressed, and the fracture never healed [Figure 2].

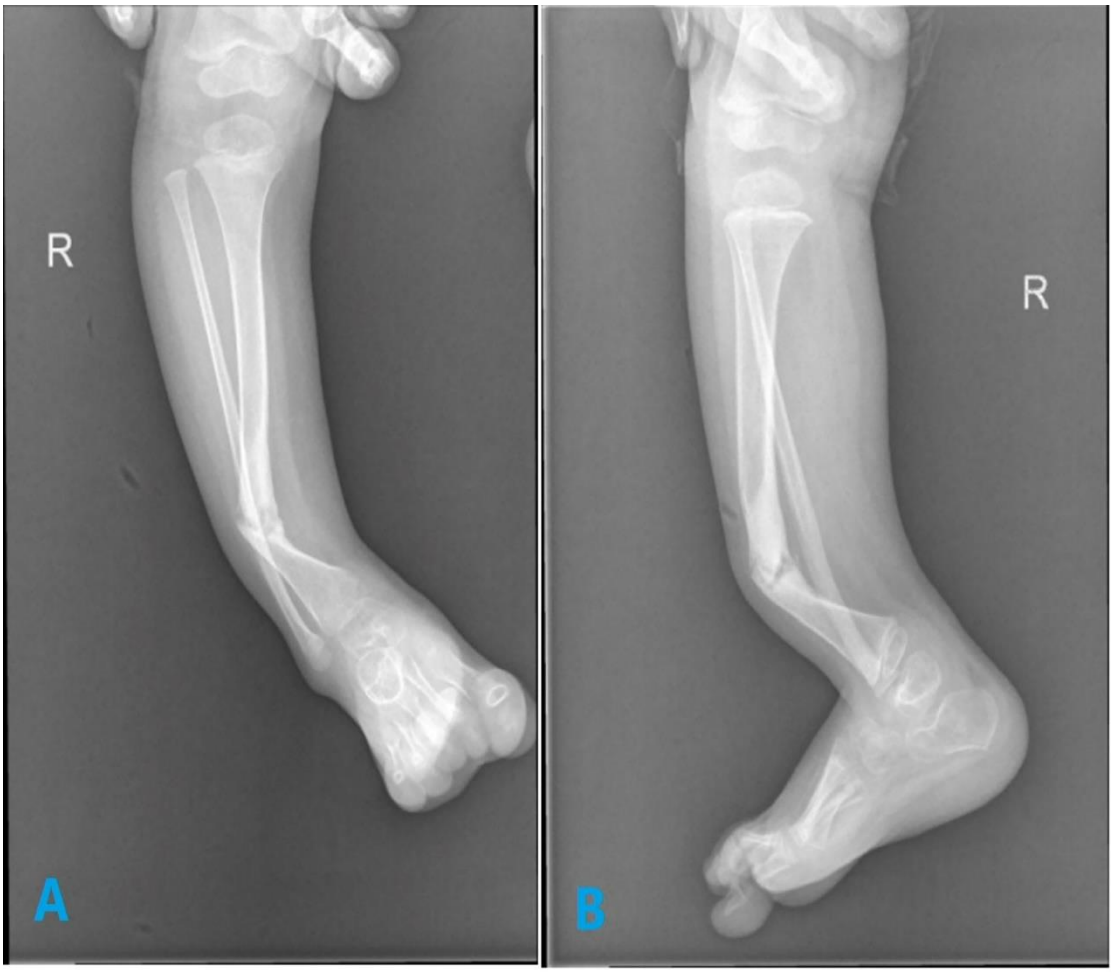

Figure 2: Anteroposterior (AP) and sideway scene of the right leg after months of diagnosis and initiation of conservative management. The deformity became worse, and the fracture was not healed.

In the next year, surgical intervention with fibular osteotomy, excision of the diseased ends, bone grafting and Ring External Fixation did not achieve union and the deformity increased [Figure 3].

At the age of 4 years, last surgical trial of treatment was done, actually, it was a combined pharmacological and surgical treatment method, which been carried by well-trained pediatric orthopedic surgeon at this time by open reduction with internal fixation by 3.5 Less Invasive Stabilization System (LISS) plate with locked screws, insertion of ante grade titanium Elastic Nail for the tibia and Retrograde Kirschner-wire (K-wire) for the fibula.

This was followed by a free periosteal graft harvested from the ipsilateral iliac bone and wrapped around the tibia and bone graft on top of it. 2 Bone Morphogenic Protein 2 (BMP2) layer was added as the last layer. The wound was closed over a drain and a below knee back slab was applied. Follow up in the outpatient visits showed nearly complete bone healing of both bones right leg. [Figure 4].

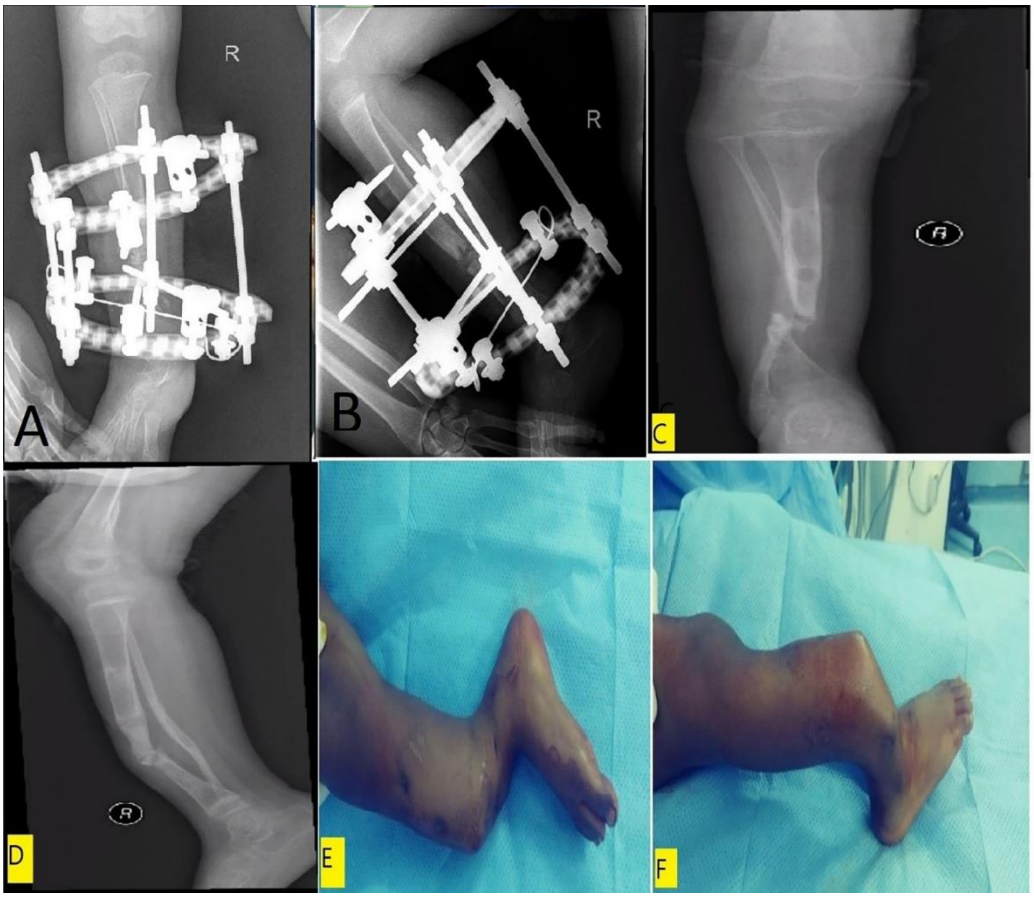

Figure 3: Fibular osteotomy, excision of the diseased ends, bone grafting and Ring External Fixation.

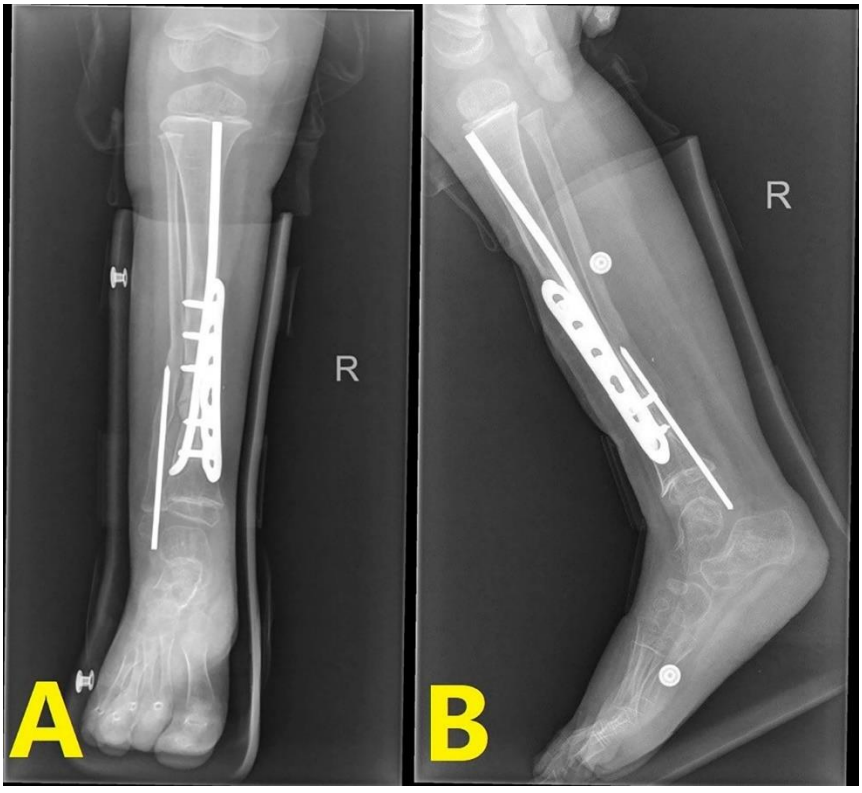

Figure 4: Anteroposterior (AP) and sideway scene of the right leg 3 months postoperative showing good union with correction of the deformity.

The patient started full weight bearing while wearing ankle Foot Orthosis (AFO) three months post operatively. The patient then started on (Zoledronic acid). Assessment of lower limb length showed shortening in his right lower limb [Figure 5].

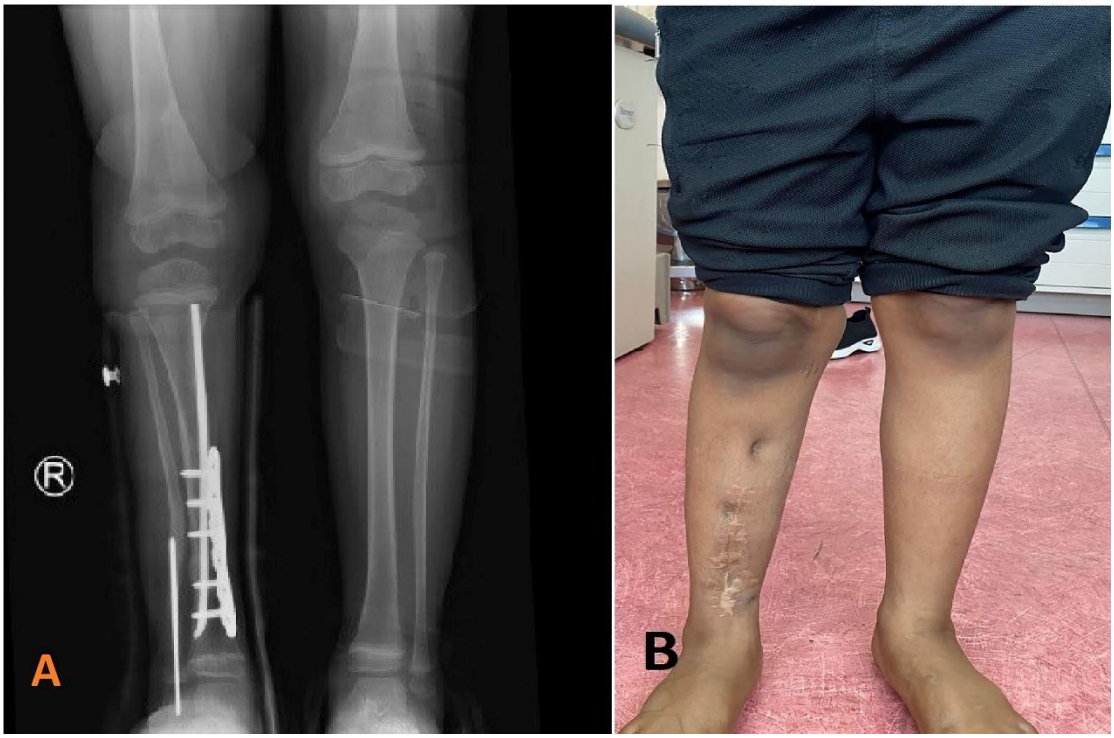

Figure 5: Anteroposterior (AP) radiograph of both lower limbs 6 months postoperative showing healed Fracture, and right lower limb shortening.

Complete healing was achieved after six months post operatively, and the fibular K-wire was removed. At the 18 months postoperative follow up, the patient was 6 years old, and was walking, but with limping. There was no leg deformity, and radiograph exhibited comprehensive restoration of the CPT with shortening of right lower limb [Figure 6].

The patient then follows up once every six months for 2 years showed complete healing and union with no refracture and shortening about 3.5 cm of the right tibia, he will be for lower limb lengthening in the future.

4. DISCUSSION

Tibia Pseudoarthrosis (CPT) is an uncommon musculoskeletal deformity with a 1: 140 000 to 1: 250,000 incidence rates (Manner HM et al., 2005; Paley, 2019). CPT is considered a primary periosteal disease as well as secondary bone disease that is associated with increased activity of osteoclast along with the lowered osteoblastic activity. In general, two goals of treatment of CPT are offered: primary and secondary. The 1st target of treatment is applied to achieve union, correct the mid-shaft deformity, and correct any fibular migration proximally, and the secondary goal of treatment is how to maintain union (Shannon CE et. al., 2021).

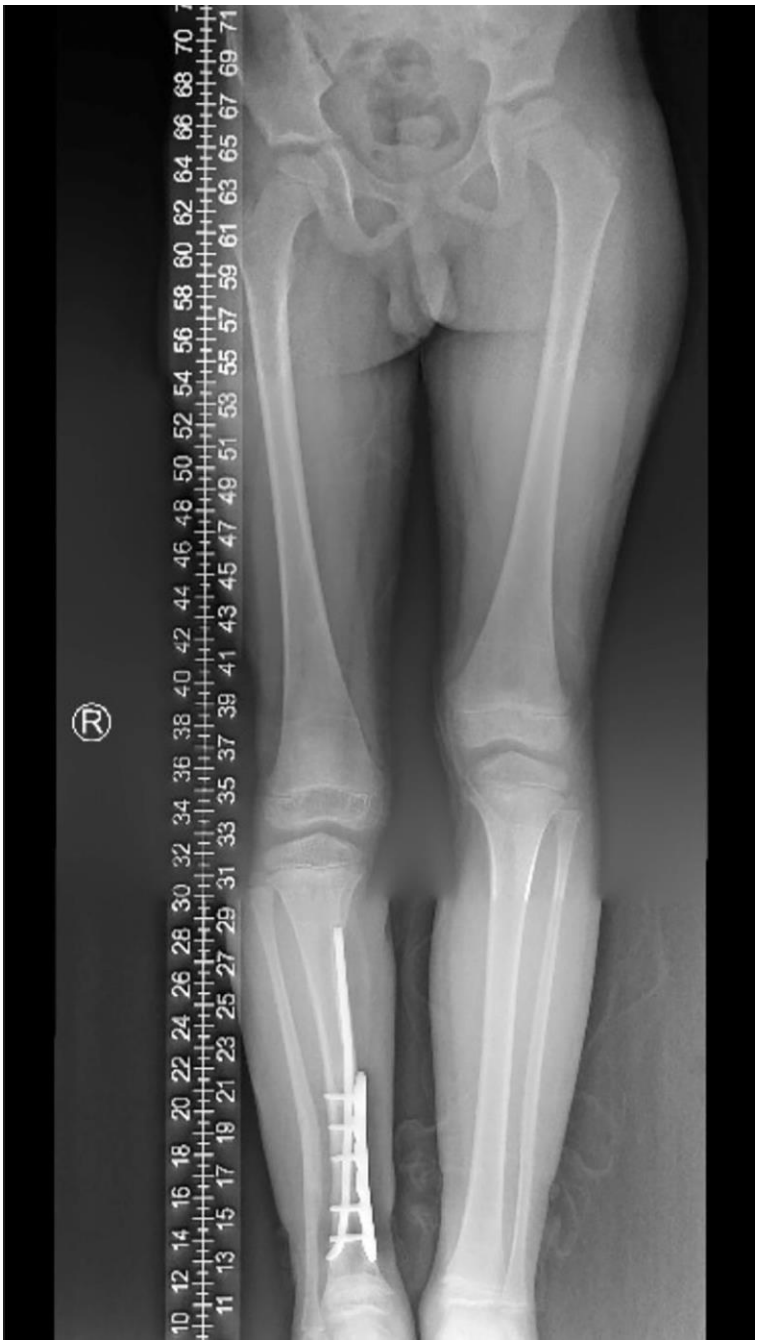

Figure 6: Radiograph showing complete healing of the CPT with shortening of about 3.5 centimeters of both right Tibia and fibula.

Tibia Pseudoarthrosis was classified by Paley into; Type 1 in which no fracture is present. Type 2, which is further split into (i) no tibia fracture, fibula fracture with fibula at station, and (ii) proximal migration. Type 3, which revealed tibial fracture without fracture fibula. And lastly type 4, which is further subdivided into (a), (b) and (c). In Type 4a, the tibia and fibula fractures are present with fibula at station in contrast to Type 4b in which a proximal migration is found, lastly, Type 4c in which the tibial defect besides fibular proximal migration is found (Paley, 2019). The pathogenesis of CPT revealed the presence of increased osteoclasts through ambient fibrous hamartoma and a reduction in the osteogenesis process and Bone Morphogenetic Protein (BMPs) by the bone and the net results are loss of remodeling potential, gradual anterolateral bowing, bone atrophy and tibia and/or fibula fractures on occasion (Manner HM et al, 2005). The challenges of genetically inherited pseudarthrosis management are in the form of several pathobiological, and pathomechanical concerns from the pathobiological concerns, the healthy periosteum is replaced by fibrous hamartoma, which leads to marked osteolysis and vascular abnormality. However, despite these changes, the bone is not altered completely and most of the bony mass is still viable with resultant bony sclerosis, which leads to obliteration of medullary canal. In addition, the biochemical alteration occurs in the osteocytes that resulted in decrease production of BMP and on the other hand, the activity of osteoclasts is increased (Paley, 2012). The presence of angular deformity of the diaphysis that gives the

appearance of anterolateral bowing is considered as one of the pathomechanical concerns (Vander Have, et al., 2008). In addition to the presence of abnormal position of the fibula that gives rise to proximal migration, also, one of these pathomechanical problems is the presence of a small cross section of bone in the fracture site. The following points should be considered prior to treatment: comprehensive resection of hamartoma, and all the bone, at the CPT site, surrounding structures and bone cells must be viable (Cho TJ et al., 2008). Furthermore, when a bone graft is required, the type of bone is critical because autogenous cancellous bone grafts contain more progenitor cells as opposed to autogenous grafts and hence produce more bone. Angular correction, intramedullary fixation and rigid fixation are needed to prevent refracture and provide greater rigidity at the CPT site. BMP may be used to promote the damaged tibia's BMP production, and periosteal grafting can aid in the restoration of healthy periosteum. Finally, the prevention of osteoclasis by zoledronic acid also prevents rapid bone graft resorption. The traditional treatment techniques of CPT are done by Ilizarov technique, internal fixation by intramedullary rods with bone grafting, and vascularized fibula transfer. These methods may be used separately or in combinations. Furthermore, adjunctive to the methods, BMB and bony graft may be used (Paley, 2012; Vander Have, et al., 2008). Other reported methods such as Thabet et al. and Paley are used as alternative methods that have both higher refractor rate and rapid union than these traditional techniques (Thabet, et al., 2008; Paley, 2019). In the presenting case of CPT, we used the combination of autogenous periosteal bone grafting with internal fixation by plate and screw and intramedullary rods of both bones and with the utilization of BMP, in addition to postoperative Zoledronic acid infusion provided good clinical and radiological results at 18 months post operatively. Thabet and colleagues used a similar combination in 20 cases of CPT with some modifications as they used Ilizarov external fixator, and without IM rodding or BMP, and the results of all 20 cases achieved union, but, on follow up, refracture took place in eight cases later (Thabet, et al., 2008). Recently, a technique offered by Paley and his team termed Paley X-Union Protocol [PXUP] used a combination of pharmacological and surgical with periosteal grafting and obtained a union percentage of 100% and no Refracture was recorded (Paley, 2012). However, two differences of the treatment of the presenting case with that of Paley, firstly; Paley used Zoledronic acid pre and postoperatively, second; to decrease the rate of Refracture, Paley created a cross union between tibia and fibula to increase a representative sample of union, which was not used in the presenting case. Kesireddy study reported the union rate is 75%, with a refracture rate of 35%, which achieved by using the traditional techniques, nevertheless, this study did not introduce an added advantage of BMP to any of the techniques (Kesireddy N, et al., 2018). In addition, Birke et al. revealed that a recipe of Bisphosphonate and BMP may give rise to lower the osteoclastic activity, and which improves the process of healing and increase its rate (Birke O et al., 2010).

5. CONCLUSION

It is True that the management of CPT is challenging and of much debate, but with the combination of autogenous periosteal bone grafting with internal fixation and intramedullary rods of both bones, utilization of BMP, pre and postoperative Zoledronic acid and free periosteal graft provided good clinical and radiological results, as seen in our case full tibia union with no Refracture after 4 years of follow up.

Ethical approval

"The study was carried out in conformity with the Declaration of Helsinki, and the Institutional Review Board gave its approval" (Ethics Committee) of King Fahad Hospital, which is Affiliated by Faculty of Medicine at Albaha University.
Approved Research Proposal. Ref number KFH/IRB/21122021/5.

Author contribution:

Author certifies that he has participated sufficiently in the work, including participation in the Conceptualization, Methodology, Investigation, design, analysis, writing, and revision of the manuscript. Author accepts full responsibility for the work and he oversaw the study's implementation, had access to the information, and took the choice to publish.

Informed Consent

The patient's parents provided written informed consent for publication of this case report and associated images.

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This study has not received any external funding.

Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

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