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Original Article

A short series of congenital pseudoarthrosis tibia

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ABSTRACT

Background: We have done retrospective study of 12 cases. Congenital pseudoarthrosis tibia is one of the most challenging subject as far as treatment is concerned. In all the treatments the basic biological considerations are the same that is pseudoarthrosis resection and bone grafting of the defect by stable fixation and correction of angular deformity without leg length discrepancy.

Methods: Various treatment modalities are described in literature up till now out of which only few modalities got success which are intramedullary roding and graft after correction of deformity, live fibular graft after excision of pseudoarthrotic part and bone transport by ilizarov method after removal of pseudoarthrotic part (acute or gradual transport as per the length of the defect). Our method is ilizarov and bone transport.

Conclusion: We have treated 12 cases of pseudoarthrosis by correction of deformity, excision of pseudoarthrotic part and bone transport by ilizarov technique. We got excellent results in 8 cases, good in 3 cases and poor in 1 case. One poor result is because of uncontrolled infection as well as pin track infection and recurrence of deformity after removal of frame.

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1. Introduction

Congenital pseudarthrosis of the tibia (CPT) is a rare pathology, which is usually associated with neurofibromatosis type I. The natural history of the disease is extremely unfavourable and once a fracture occurs, there is a little or no tendency for the lesion to heal spontaneously. It is challenging to treat effectively this difficult condition and its possible complications.

Treatment of CPT, it is a challenge, even to an experienced orthopaedic surgeon, various modalities are available but the useful treatments are very few. These are live fibular grafting,

intramedullary roding with bone graft and excision of pseudoarthrosis with bone transport by Ilizarov. Treatment is mainly surgical and it aims to obtain a longterm bone union, to prevent limb length discrepancies, to avoid mechanical axis deviation, soft tissue lesions, nearby joint stiffness, and pathological fracture. The key to get primary union is to excise hamartomatous tissue, pathological periosteum and to correct angular deformity. Age at surgery, status of fibula, associated shortening, and deformities of leg and ankle play significant role in primary union and residual challenges after primary healing. The treatment of Ilizarov will include healing as well as treating limb length discrepancy and this technique

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also increases the vascularity of bone and soft tissue surrounding the bone, so the regenerate which is formed is of good quality as in this technique upper metaphyseal corticotomy is performed.

In this study we are putting short series of CPT. We have treated 12 cases by excision and bone transport by Ilizarov. We got excellent results in 8 cases, good in 3 cases and poor in 1 case. The poor result was because of recurrence of infection and deformity in spite of routine line of treatment.

2. Material and method

Total no of 12 cases taken. All cases treated by excision of pseudoarthrosis and bone transport by Ilizarov. Number of surgery performed in each case ranges from 1 to 4 before Ilizarov treatment and follow up of 5 years taken with age ranging from 6 to 15 years.

First we correct the deformity then excise the pseudoarthrotic part and do compression at the pseudoarthrotic site if gap is less than 3 cm, fixation of frame in anticipation with upper metaphyseal corticotomy and lengthening at the site of corticotomy. If the gap is more than 3 cm then we do corticotomy and gradual transport to cover the pseudoarthrotic gap.

There are main three criteria to remove the frame.

- (1) complete healing of pseudoarthrosis
- (2) X-ray findings suggestive of formation of cortex in all views (minimum three cortices should appear).
- (3) patient is able to stand on his affected limb. (single leg weight bearing)
- (4) when long gap is covered after removal of frame then plaster is to be given for 1–2 month followed by corset for at least 6 months to strengthen the pseudoarthrotic part as well as consolidation of the regenerate.

3. Aetiology

Exact cause is undetermined, many theories concerning its pathogenesis.

- Intra Uterine trauma (Birth Fracture)
- Generalized metabolic disturbances
- Vascular malformations but all hypothesis are discarded
- CPT and Neuro-fibrometosis seem to be related
- Ducroquet who reported café-all-bit spot positive in 9 out of 11 cases of CPT
- Barber in 1939 – 5 patients of CPT had pigmental spots whose immediate families had evidence of Neuro-fibrometosis.
- Green and Rudo established intraosseous Neuro-fibrometosis

4. Pathogenesis

In CPT, the affected tibia exhibits insufficient mechanical strength and osteogenetic capability. An abnormal highly cellular fibrovascular tissue grows at expense of the bony cortex. As it encroaches on the bony cortex it promotes incoordinate

osteoclastic bone resorption that does not have a normal bone modelling purpose. Reactive changes simultaneously occur at medullary aspect leading to deposition of excess trabecular bone and hence medullary sclerosis. Excessive bone resorption accounts for intracortical cysts. Impaired vascularization can result in decreased osteogenic capabilities. The similarity of ultrastructural findings in the abnormal periosteum and in skin neurofibromas of neurofibromatosis patients may indicate a pathogenetic association of both diseases.

5. Pathology

The main histopathologic change found in the pseudoarthrosis is highly cellular fibrovascular tissue accompanied by paucity of vascular ingrowth. Some investigators have suggested that the primary pathologic lesion is in the periosteal tissue around the tibia rather than the bone itself.

The soft tissue at the pseudarthrotic site is composed of variable admixture of fibrous tissue, fibrocartilage, and hyaline cartilage with evidence of enchondral ossification.

6. Numerus theories

- Vascular insufficiency – Codvilla → 1906
- Genetic defects Gaenslen → 1915
- Nutritional deficiency Henderson → 1928
- Foetal damage or compression by amniotic bands Inglis → 1928
- Disturbance of neural tissue and function Moore → 1944
- Endocrine – Imbalance Duraiswami → 1950
- Embryonic defect in Neuro-vascular supply Badgeley → 1952
- Infection during pregnancy Pagella → 1978
- 50% CPT associated Neuro-fibrometosis Moore → 1957, Morrissey → 1982
- Only 5–11% of Neuro-fibrometosis has CPT Mc Carroll → 1950
- Six Café-all lait Spots of .5 cm in diameter is considered diagnostic Crowe → 1956
- Lloyd Roberts in 1969 stigmata after absent at birth but it will appear later → 1969

HYPOTHESIS

COMPANACCI → 1973

Mechanical force which accentuated the problem

MC FARLAND → 1951

Established fracture

Atrophic non-union

Stress Fracture

Buttressing I/M callus on concave side

Thinning of cortex on convex side

Transverse radiolucent line at the apex (Bassett → 1981)

7. Classification

Mainly two types of CPT

- 1) Atrophic
- 2) Cystic

7.1. Atrophic

Atrophic more common

Diameter of tibia is narrowed

Sclerosis or complete obliteration of marrow

Tibia bowed anteriorly

Fracture at birth or occur later in the 1st few years

Following fracture bone ends taper. Periosteum thickened,

Hamartomatous proliferation of fibrous tissue at fracture site

Fibula may also fractured

Pseudoarthrosis type nonunion and refractures are common

7.2. Cystic

There is a cyst at lower 3rd tibia

Content resemble that of fibrous dysplasia

Diameter not affected

Pseudo arthrosis of fibula may be present

Other sites of pseudo arthrosis are first rib, ulna, clavicle, humerus and femur

Various classification systems have been suggested for CPT, including those of Boyd, Anderson, and Crawford. These systems generally describe the radiographic appearance of the untreated bone at the pseudoarthrosis site, the presence of fracture at birth, and involvement of the fibula. However, none of the classification provides specific guidance for management or is predictive of outcome.

8. Andersen's

Andersen's classification of CPT (1973) differentiates the morphology of the pseudoarthrosis as dysplastic, cystic, or sclerotic types, in addition to a clubfoot type that arises because of accompanying abnormalities. According to

Anderson, neurofibromatosis is not associated with the cystic form. However, Morrissy et al reported three cases of cystic pseudoarthrosis and neurofibromatosis.

9. Boyd's

The Boyd's classification (1982) identifies six types of CPT. In type I there is anterior bowing with tibial defect. Type II and III are characterized by pseudoarthrosis with hour-glass constrictions and bony cyst, respectively. In type IV, pseudoarthrosis is present with sclerotic segments and march fracture. Type V is characterized by a complementary dysplastic fibula. In type VI, an intraosseous neurofibroma or schwannoma is evident.

Type 1	Anterior bowing of tibial defect
Type 2	Hour-glass constriction
Type 3	Hour-glass constriction with bone cyst
Type 4	Pseudoarthrosis is present with sclerotic segments and march fracture
Type 5	Characterized by a complementary dysplastic fibula.
Type 6	An intraosseous neurofibroma or schwannoma is evident.

10. Crawford's

Crawford's classification of CPT (1986) identifies four types of CPT all with anterolateral bowing of the tibia. In type I, the medullary canal is preserved and cortical thickening at the apex of the deformity might be observed; patients with this type usually have a good prognosis; some may not even have a fracture. Type II is defined by presence of thinned medullary canal, cortical thickening, and trabeculation defect. The dominant finding in type III is a cystic lesion, which may be fractured; patients with this type of pseudoarthrosis tend to experience early fracture and, therefore, require early treatment. In type IV, pseudoarthrosis is present with tibial and possibly fibular nonunion.

Type 1	The medullary canal is preserved and cortical thickening at the apex of the deformity might be observed
Type 2	Presence of thinned medullary canal, cortical thickening, and trabeculation defect.
Type 3	Cystic lesion, which may be fractured
Type 4	Pseudoarthrosis is present with tibial and possibly fibular nonunion.

Johnston suggested that two criteria be considered to initially classify anterolateral bowing of tibia: (1) presence or

absence of fracture (2) the age at which the first fracture occurs (early onset before age 4 years, delayed onset after age 4 years). Tibia that fracture and go on to pseudoarthrosis are treated surgically, intact tibias are observed, with perhaps some orthotic protection.

11. Clinical features

CPT is characterized by anterolateral angulation of the tibia known as kyphotic tibia. This bowing, combined with an overall decrease in distal tibia growth, results in shortening of the limb.

Anterolateral bowing may be noted at birth. The deformity is an apical prominence in the leg laterally, with the foot inverted or medially displaced in relation to the lower leg. If fracture has occurred mobilization at the site of pseudoarthrosis is evident. When signs of neurofibromatosis are present, the diagnosis is more straightforward. Much later, mild deformity may be noted when a limp develops because of shortening or impending fracture. The foot on the involved extremity may be normal or just slightly smaller than the contralateral foot.

12. Treatment

The Management of congenital pseudoarthrosis of the tibia remains one of the most difficult problem. Rober W H & Pho. have so far found more than forty operative techniques that have been described. A review of the literature discloses many methods advocated, Boyd & Saged report. 23 variations of operations in 167 procedures in 91 patients ranging from conservative measures to radical upto Amputation.

Treatment of CPT is challenging. The primary goal is to obtain and maintain union while minimizing angular deformity.

12.1. Mc. Farland bypass graft 1957 (Inlay Graft)

Steps

Resection of sclerotic bone with rongeur

Drilling of the medullary canal

Excision of segment of fibula if tibial fragments are held apart

Retrograde insertion of Steinmann pin and impaction of tibial and fibular fragments done.

12.2. Boyd double only

15–20 degrees of posterior angulation created by manipulation

Grafts held by silk sutures

Osteoperiosteal grafts placed on lateral and posterior surfaces
Cancellous bone chips and cortical matchsticks impacted around pseudoarthrosis.

Charnleys compression clamp used where distal distal tibial fragment large

In this manner antero-posterior angulation corrected

When distal tibial fragment short intramedullary pin used

12.3. Sofield fragmentation

In this technique the bone is fragmented into many pieces, then the pieces are shuffled and resettled with IM fixation.

12.4. Intramedullary stabilization

Intramedullary stabilization of CPT was introduced by Charnley. The intramedullary nail provides stability to allow healing of the pseudoarthrosis and helps prevent refracture. Fixed length solid nails can be inserted through the heel or, less commonly, through the knee, but may require replacement as the child grows.

Several steps are common to successful IM treatment of CPT. First, abnormal tibial bone and associated abnormal periosteum must be resected. If present, a fibular pseudoarthrosis also must be resected. Next, the normal tibial ends are apposed and stabilized with the IM nail, and bone graft is placed. IM stabilization should remain in place after healing to help prevent refracture.

Damage to articular surfaces by transfixation of the ankle, combined with fibrosis and atrophy of the periarticular tissues resulting from lack of ankle motion and longterm bracing to protect the trans articular implant, uniformly produces a calcaneus gait and a stiff ankle. Recognizing the detrimental effects of the rod across the ankle, especially in patients who appear to have gained union of the pseudoarthrosis, some authors recommend surgically pushing the rod up, out of the ankle and into the tibial medullary canal, as soon as possible following apparent union and after sufficient longitudinal growth has occurred to make this feasible.

13. Free vascularized fibula graft

Surgeons generally harvest 10–12 cm length of contralateral fibula with its vascular pedicle. The fibula is then placed in the gap and fixed by a variety of techniques. Initially these techniques included plating or simply screws; more recently, IM nailing has been used. The vascular pedicle typically is attached to the anterior or the posterior tibial artery and any associated vein.

The high success rate of primary and secondary union is the distinct advantage of vascularised fibular graft.

Problems encounter included nonunion, predominantly distal. Moderate to severe angular deformities were reported as well as notable limb length discrepancies.

13.1. Former composite bone graft 1952

Canurati F/U 30 cases Union (Bony) 67% (Success Rate)

Marrissy 1982 → 55% (Success Rate)

(Many after several surgeries)

Amputation – Early Treatment → Failure to surgery

(Anderson – 1976, Masserman – 1974)

13.2. Sutcliffe. Goldberg, Patterson, Simonis Basset – 1981, 1982, 1985 → 70% success

• Weiland → 1979

• Chen → 1979

- Gilbert2 → 1980
- Pho → 1985

(Free vascularize fibular graft reported high success rate)

14. Ilizarov technique

A number of techniques have been reported, and several different frame configurations and strategies have been used. Most commonly, an Ilizarov frame is applied with two rings, one above and one below the pseudoarthrosis. When pseudoarthrosis is distal, the foot is included in the frame. The pseudoarthrosis is resected, and the gap is compressed. When there is limb length discrepancy, a proximal corticotomy is done for distraction. Alternatively, the area proximal and distal to the pseudoarthrosis is resected, the four ring frame is applied, a corticotomy is done proximal, and the bone is transported into the gap.

Main four methods by Ilizarov

- (1) Epiphyseolysis (epiphyseal lengthening)
- (2) Compression distraction method
- (3) Bone transport
- (4) Tibialisation of fibula (when defect is large and fibula is not involved in pseudoarthrosis)

The main drawback of Ilizarov technique is early fusion and limb length discrepancy.

Three main treatments: recently in picture

- (1) Vascularize Free Fibular Graft
- (2) Ilizarov Bone Transport
- (3) IM roding

A 5 years old male child with pseudoarthrosis tibia right side with almost right angle deformity at the junction of lower 1/3rd – middle 1/3rd tibia-fibula.

Clinical at the time of presentation.

X-ray at the time of presentation.

Dr. Dror Paley's calculation was carried out after tracing of the X-ray for placement of hinge & distractor.

1st operation compression at the site of pseudoarthrosis.

Correction as per calculation.

After correction deformity recurred.

After 1st surgery in spite of external support deformity recurred.

It is believed that after correction if you do compression at the site of pseudoarthrosis & corticotomy at upper metaphyseal region pseudoarthrosis heals. But here the deformity recurred at pseudoarthrotic region, so again Dror Paley's calculation applied.

According to Dr. Paley's calculation deformity corrected, pseudoarthrotic portion was excised & then compression done & lengthening done at upper metaphyseal corticotomy. After getting consolidation at both the sites, frame was removed & limb was supported with long leg corsette from groin to toe for 6 months.

In spite of corset deformity recurred so case was referred to Professor Catagny by e-mail & he advised to put intramedullary rod after correction with the frame & then to perform compression at the pseudoarthrotic site & recorticotomy performed at the upper metaphyseal region.

Frame was removed & rush pin kept in for consolidation. 6 months after external support intramedullary rush pin was removed. Solid consolidation achieved in tibia but fibular pseudoarthrosis in lower 1/4th persists so foot was going into mild valgus for which special shoes with orthotics were provided.

15 years follow up after surgery, pre operative photographs showing left valgus foot with external rotation deformity with angulation at the junction of lower 1/3 middle 1/3 junction with persistant pseudoarthrosis of fibula.

After 15 years follow up left tibia was formed normally but because of pseudoarthrosis lower 1/3 fibula patient had valgus foot with mild external rotation deformity in lower 1/3, middle 1/3 junction with mild valgus of the heel for which final surgery was carried out at the level of cora of left leg deformity of tibia where angulation as well as rotation was corrected and tibia was plated with long lock plate which has corrected its complete deformity.

the disease have been recognized. Even when union of the pseudoarthrosis is achieved, patients may have compromised function. Residual limb length discrepancy is common. Discrepancies of less than 5 cm may be treated with contralateral epiphysiodesis when sufficient growth remains. Larger discrepancies can be treated by lengthening of the shorter leg.

16. Conclusion

We have considered the following criteria for grading

- (1) cure of pseudoarthrosis.
- (2) movement of joints proximal and distal to the lesion
- (3) no limb length discrepancy
- (4) 100% correction of the deformity
- (5) patient moving without support

Our series shows that Ilizarov technique is an effective and practical treatment for CPT. The Ilizarov technique allow osteosynthesis, ankle stabilization and no limb length discrepancy.

Final follow up.

15. Complications

The treatment of CPT is fraught with complications, and failure is common. Complications both of treatment and of

Conflicts of interest

All authors have none to declare.

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