Neglected Congenital Pseudarthrosis of the Tibia in an Adult Treated by Deformity Correction and limb Lengthening using Ilizarov Ring Fixator

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Abstract: Congenital Pseudarthrosis of the tibia (CPT) is one of the challenging problems in paediatric orthopedic surgery. It is usually associated either with neurofibromatosis or fibrous dysplasia. Different techniques for the management of CPT are present. Success of procedure depends on complete resection of hamartomatous fibrous tissue and maintenance of union. Success rate of treatment methods is directly proportional to age. We are presenting a case of adult neglected Pseudarthrosis of tibia treated by ilizarov ring fixator and corticotomy.PTB brace used to maintain union.

Keywords: Pseudarthrosis of tibia, ilizarov ring fixator, corticotomy, PTB brace.

1. Introduction

Congenital Pseudarthrosis is a specific type of nonunion that at birth is either present or incipient. The incidence ranges from 1:140000 to 1:250000. Its cause is unknown, but it often associated either with neurofibromatosis or related stigmata (50%), with fibrous dysplasia (10%) or idiopathic (40%). Congenital Pseudarthrosis of the tibia (CPT) is one of the challenging problems in paediatric orthopedic surgery. Different techniques for the management of CPT are bypass fibular graft, double on lay graft taken from the opposite tibia combined with autologous iliac crest graft, intramedullary rods, ipsilateral transfer of the fibula or contra lateral free vascularized fibular transfer, ilizarov circular external fixation, combined external fixation and intramedullary devices, direct current or pulsed electromagnetic field, "fibula pro tibia" technique with fibular fixation to the Pseudarthrosis site, recently bone morphogenic protein (BMP) and bisphosphonate therapy. Refracture rates are high with all of these methods.

2. Case Report

20 yrs old male patient presented to our orthopaedic OPD with complaint of deformity in right lower limb since birth. Associated neurofibromas and café au lait spots seen all over his trunk. Past history of surgical treatment with intra medullary nailing of tibia in childhood present but deformity to progress. His mother also continued having neurofibromas. On examination anteromedial bowing of leg with gross shortening of right lower limb present (picture1). On deep palpation mild tenderness and gross abnormal mobility present at pseudosrthrosis site. We confirmed the diagnosis with radiographs (Fig. 1) which showed involvement of both tibia and fibula at middle and lower third junction with broken rush pin in tibia.



Figure 1: Xray showing Pseudarthrosis involving both tibia and fibula with broken rush pin

As patient having associated neurofibromas, we doubted the union chances at Pseudarthrosis, so initially we planned for deformity correction and union at Pseudarthrosis as first surgical procedure and corticotomy for limb lengthening later. After spinal anaesthesia under tourniquet we approached tibia through anterior longitudinal incision and opened the Pseudarthrosis to remove broken rush pin pieces. Dissection between periosteum and surrounding soft tissue carried out and hamartomatous periosteum excised circumferentially. Osteotomy of tibia done and deformity correction done. The fibular periosteal hamartoma resected along with segment of fibula. Proximal and distal fragments of tibia aligned and stabilized with ilizarov ring fixator. Pseudarthrosis site augmented with autogenous iliac crest bone grafting. Mobilization started immediately in post operative period with shoe rise. We followed the patient at 6, 12, 18 weeks with radiographs. As the signs of union visible at Pseudarthrosis site after 4 months (Fig.2),



Figure 2: Showing union at Pseudarthrosis site

we did corticotomy and lengthening of tibia by 7 cms (Fig.3). Ilizarov ring fixator was kept until consolidation of corticotomy site (Fig.4). Later at the end of 10 months ring fixator was removed and above knee pop cast applied.



Figure 3: Showing corticotomy and lengthening of tibia. Fig.4.Showing consolidation of corticotomy site

After removal of pop cast custom made PTB brace (**picture 2&3**) was given for support and mobilization. Total duration treatment was nearly 12 months.



Picture1: Showing preoperative deformity



Picture 2: After correction of deformity



Picture 3: PTB brace for support and to maintain union.

3. Discussion

Treatment of Congenital Pseudarthrosis of the tibia (CPT) and achieving union is one of the challenging problems in orthopedic surgery. Codivilla was the first to implicate the periosteum in the pathology of CPT. Mc Elvenny reported a markedly thickened, closely attached periosteum that caused constriction of the bone with subsequent atrophy and Pseudarthrosis. A recent report suggested that the thickened periosteum was caused by neural like cells that form a tight sheath around the small periosteal vessels causing narrowing or obliteration of vessels This results in disturbance of the blood circulation of the periosteum, which in turn results in impaired oxygen and nutrient supply of the subperiosteal bone with subsequent fracture and recalcitrant nonunion. Cho et al. studied osteoclastic and osteoblastic activities of the periosteum. They concluded that refracture after initial healing and resorption of bone grafting are related to osteoclastic activity of the periosteum. The main surgical options for treatment of CPT are vascularized fibular grafting, intra medullary stabilization, external fixation with ilizarov circular frame, and rarely amputation. Codivilla recommended osteo-periosteal grafting. Recently Paley combined his periosteal grafting methodology with pharmacologic treatment using BMP and bisphosphonate infusion. This combination has reduced the refracture rate and accelerated the union rate. Ilizarov ring fixator is having advantages like early mobilization in post operative period and correcting limb length discrepancy by corticotomy and lengthening. Success of procedure depends on complete resection of hamartomatous fibrous tissue, but it does not ensure healing or prevent refracture. The refracture incidence is reversely proportional to age. Success rate of treatment methods is directly proportional to age.Factors that significantly decrease refracture were increasing the cross sectional area of union and eliminating angulation especially at the Pseudarthrosis site. The natural history of CPT is recalcitrant nonunion, atrophy of the bone and the leg, progressive deformity, limb length discrepancy (LLD) and recurrent refracture even after achieving union. The primary objective of treatment for CPT is to obtain union. The secondary objective is to maintain union using braces externally until skeletal maturity of patient. Brace wear at all times including during sleep and the only time the brace is taken off is for bathing and for physical therapy.

4. Conclusion

Since Congenital Pseudarthrosis of the tibia is a rare disease and failure is devastating, it is more important to have a successful method than to be certain which component of the treatment regimen is the most important to achieving successful union. The combination treatment like hamartoma resection, stabilization of bone fragments, bone grafting play vital role in treatment of Ilizarov ring fixator helps in union with advantage of early mobilization and limb lengthening.

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