

Treatment of Tibial Nonunion in Paget's disease by Ilizarov Distraction Osteogenesis

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SUMMARY

Surgical treatment of fractures and deformity correction in Pagetoid bone is often complicated by nonunions and implant failure; we describe different cases of monostatic Paget's disease of the tibia treated successfully by the Ilizarov distraction osteogenesis technique. The study was conducted during my post graduate fellowship training at Maryland University Baltimore, USA under the supervision of Dr. Dror Paley and Dr. John E. Herzenburg.

Key words: Paget's Disease, tibia, fracture, nonunion, Ilizarov technique.

INTRODUCTION

The first descriptions of Paget's disease were provided before the advent of radiography and were based on the study of anatomic specimens and patients seen by the English physician, Sir James Paget. Paget described the disease as a form of chronic inflammation of bone, causing softening, enlargement and excessive production of imperfectly developed structure with increased blood supply. He stated "it begins in middle age or later, it is very slow to progress and may continue for many years without influence on the general health and may give no other trouble other than that which is due to the change in shape, size and, direction of the diseased bone. Even when the skull is hugely thickened and all its bones exceedingly altered in structure, the mind remains unaffected"²⁶.

In the early stages, there is an increased breakdown of bone by osteoclasts, followed by excessive bone formation by osteoblasts. Patients present with thickened cortices, coarse scant trabeculation, and bowing deformity. The diseased bone is structurally weak and is prone to fracture. The most commonly involved bones are the skull, spine and pelvis while the clavicle, ribs, small bones of the hand and feet, facial and jaw bones, patella, and tibia are also affected in decreasing numbers^{1,10,13,16}.

Fracture and deformities are well recognized complications of Paget's disease. Management by internal fixation (nailing and plating) is difficult because of the poor quality of bone^{15,18,19}. The presence of complex deformity^{2,23}, and excessive bleeding during surgery^{8,15,23}. There are different reports in the literature describing a high incidence of

nonunions and implant failures after internal fixation^{28,15,23,26}. We present the first report of treatment of deformity and nonunion for the Pagetoid tibia by ilizarov distraction osteogenesis techniques.

CASE 1

A 78 year old female had monostotic Paget's disease of the left tibia for 20 years. She developed an anterior bowing deformity of the left tibia and medial compartment degenerative arthritis of the knee. Twelve years ago, she underwent a Coventry osteotomy. Her knee pain was only partially improved over the years until she presented with a stress fracture manifested by pretibial pain and a minimal anterior cortical lucency at the apex of her curvilinear deformity along with other minimal cortical lucencies below that. She was treated conservatively with a long leg cast for six months followed by a hinged long leg brace. One month later, she sustained another transverse fracture through the original stress fracture site at the apex of the tibia and was once again treated with casting. Her fracture failed to unite and she was referred to our Center for treatment of the nonunion. We applied an ilizarov push constraint fixator consisting of two distraction rods posteriorly and a sliding bar anteriorly with three push half frames. A subperiosteal tibial osteotomy was performed with a Gigli saw leaving the anterior cortex intact. Post operatively, she was put on Calcitonin 0.5mg daily. Her fixator was removed after four months and she was placed in a short leg PTB cast for six weeks. Later, a removable orthotic for her left leg was used for two months. Follow-up radiographs showed solid bony union and complete correction of deformity. Mild equinus contracture at the ankle improved after physical therapy. At 3.5 year follow up, she remains asymptomatic, with no recurrence of her stress fracture or of the deformity.

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CASE 2

A 75 year old woman with monostotic Paget's disease of the right tibia sustained a fracture at the junction of middle and distal thirds of tibia. She was immobilized until her fracture healed and then placed in a hinged ankle patellar tendon bearing brace. However, while walking, she felt severe pain in her tibia. Two weeks after, she returned to her doctor and radiographs showed a new undisplaced fracture at the junction of proximal and middle thirds of tibia. She was treated with non-weight bearing and with passive and active ROM exercises to prevent immobilization induced hypercalcemia. The new fracture failed to unite, and she was referred to our Center for further treatment. At this time, her radiographs showed classical anterolateral bowing of right tibia along with a nonunion. Her bone scan demonstrated intense uptake in her right tibia. She also had a 5 cm leg length discrepancy. Our treatment consisted of bivel tibial corticotomy, fibular osteotomy and application of the Ilizarov device with an anterior push construct. On subsequent follow-up, the osteotomy site shows forward translation at the angular segment and opening up in the back as planned. After the 17th week, her x-ray showed full correction of her bowed leg and satisfactory healing at both osteotomy sites. At this time, the two intermediate rings were removed to dynamize the construct. Four weeks later, the Ilizarov device was removed and a long leg cast was applied for six weeks. She then resumed full activities without restriction. Her recent follow-up (3.75 years) examination shows satisfactory position and alignment and no complaints.

CASE 3

A 46 year old public works engineer with monostotic Paget's disease of left tibia received a six month course of calcitonin therapy and his alkaline phosphate and urinary hydroxyproline level returned back to the upper limit of normal range. However, during treatment he developed gradual, painful varus bowing of his left tibia. He underwent a tibial osteotomy and intramedullary nailing along with fibular osteotomy to align his leg mechanically. Later, the nail was dynamized by removing the distal screw in order to enhance healing. A short leg cast was applied at the same time. The cast was removed after three months and a brace was applied. He was treated before and after surgery with calcitonin injections three times weekly and received medication for hypertension and hyperuremic problems. His initial post-op course was complicated by a superficial wound breakdown which responded well to dressing changes and oral antibiotics. The

osteotomy failed to heal, and the tibial nail subsequently broke one year post-op. He was referred to our Center for further treatment of his nonunion. At that time, he had a residual anterolateral bow of his tibia along with 10 degrees extension lag movement. Our treatment consisted of broken rod removal, application of Ilizarov external fixator and fibular osteotomy. He received 100 units of calcitonin subcutaneously for a period of two months prior to surgery and after surgery he was adjusted on a maintenance dose of 50 units subcutaneously three times per week for 6 months. The nonunion was distracted and produced good regenerate bone. The fixator was removed seven months after surgery. A hinged cast was applied for three weeks. After that, he was placed in a PTB brace with gradual increased weight bearing. At follow-up, he is out of brace and has no pain. Radiographs show solid union with proximal medial tibial angle measuring 89 degrees and full ROM.

DISCUSSION

Paget's disease (osteitis deformans) is commonly encountered in middle aged and elderly individuals and has slight male predominance^{14,15}. Its incidence increases with advancing age^{6,29}. The positive family history is present in about 14% of cases²⁸. The exact etiology of Paget's disease is still unknown although various hypotheses including inflammatory, neoplastic, vascular, genetic, traumatic, endocrinological, immunological, and more recently, viral etiologies have been proposed^{26,27}.

Paget's disease is divided into four stages: an early destructive (or lytic) phase, a combined lytic and sclerotic phase, a sclerotic phase, and malignant degeneration phase. The first stage is seldom recognized since Paget's disease is often subclinical and limited to local area of the bone²⁵. Anderson et al have suggested five mechanisms by which Paget's disease appears osteolytic. These include early lesion, immobilization after fracture, advanced disease with secondary degeneration, seeding of an independent osteolytic lesion and sarcomatous transformation. The sclerotic phase is usually represented by subsequent reactive bone formation (osteoblastic activity) while the combination of lytic and sclerotic components seldom present diagnostic problems. The malignant degeneration phase is often associated with large soft tissue masses and cortical destruction. In case of lytic lesion, the possibility of angioma, brown tumour, fibrous dysplasia, giant cell tumor, malignant lymphoma, necrosis, radiolucency following fracture and sarcomatous changes must be ruled out. In sclerotic lesions, the possibility of metastatic lesion from prostate, breast and Hodgkins

lymphoma should be considered in differential diagnosis of Paget's disease^{3,9,13}.

The radiograph of patient with Paget's disease shows characteristic area of bone resorption and formation in disordered trabecular pattern with overall increase in size and shape of bone along with thick cortices. They may show candle frame appearance in long bones, picture frame appearance in spine, thickening of iliopectineal line and coarsening at sacroiliac joint or proteusioacetabular deformity^{13,20,23,27,28}.

Most patients with Paget's disease are asymptomatic (10-20%) and are diagnosed incidentally. Typically, a history of minor trauma and pain provokes an investigation which shows characteristic radiological appearance along with increased uptake on bone scan and elevated serum alkaline phosphatase and 24 hour urinary hydroxyproline levels²⁷. In patients with symptomatic Paget's disease, the most common complaints are pain, skeletal deformity and change in skin temperature due to increased vascularity and blood flow to the bone. Rheumatic manifestation such as rheumatoid arthritis, its variant ankylosing spondylitis, gout, calcific peri-arthritis, and metabolic complications such as hypercalcemia and hyperurecemia after fracture are also associated with Paget's disease and may contribute to the formation of renal stones. Cardiovascular complications including high output congestive heart failure may be present in patients with greater than 30% of skeletal involvement along with apathy, lethargy and easy fatigability. Neurological complications due to cranial involvement include mechanical compression on neural foramina to the exiting cranial nerves, particularly the I, II, V, VII and VIII nerves. Infiltration of the inner ear causes neurosensory deafness, softening and basilar invagination, which may produce vascular compression and blockage or cerebrospinal flow with resultant hydrocephalus^{12,16,18,19,26,27}.

In elderly patients, 20-30% of bone sarcomas are associated with Paget's disease²¹. Painful symptoms may be due to combination of factors including: metabolic activity of disease, neurological impingement, impending fractures and secondary arthritis.

Orthopaedic treatment in Paget's disease is indicated for: severe disabling arthritis, particularly hip, knee and shoulder, severe bowing deformity of femur or tibia, sacromatous degeneration, and pathological fracture.

Pathologic fractures occur with an incidence of 10-30% and present very challenging problems to the orthopaedic surgeon²⁶. They may occur most frequently in the long weight bearing bones of the

lower extremities. The femoral neck, subtrochanteric and tibial regions are most common sites although they may occur in the humerus, vertebral column, and the pelvis^{26,13,15,16}. They are usually transverse or short oblique. The surgical treatment by intramedullary nailing or plating is associated with high incidence of implant failure, nonunion, residual deformity, revision surgery and failure to return to walking status.

Bradley and Nade⁴ reported seven implant failures out of 18 transcervical fractures, 17 out of 36 subtrochanteric fractures and nonunion at both sides in all five cases of segmental fracture of the femur. The reported overall rate of nonunion was 35.5% with 69% failure of initial and revised surgery in their series. Nicholas and Killoran²³ from New York and Barry² from Australia have reported 15% rate of nonunion in their series of 23 and 70 patients respectively whereas Grundy¹⁵ from England reported 25% rate of nonunion in his series of 63 patients. Similarly, Dove[^] reported 40% rate of nonunion in his series of 150 patients. We have treated successfully three cases of established nonunion and deformity of Pagetoid tibias by distraction osteogenesis techniques with the Ilizarov external fixator. The procedure is less invasive with minimum blood loss and a high percentage of success. Therefore, we suggest that nonunion and deformities of the tibia in Paget's disease of the bone can be successfully treated with the Ilizarov distraction osteogenesis technique. Our series in small and follow-up is limited. Clearly however, the method of distraction osteogenesis shows promise in the treatment of nonunions and deformities in Paget's disease.

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