

Paget's disease Of Bone Involving Tibia in an Indian Male: Deformity Correction And Fixation With An Interlocking Nail: A Case Report

Anshuman Dutta¹, Ranjit K Baruah², Russel Haque³, Saurabh Jindal⁴

¹Anshuman Dutta, Department of Orthopaedics, Silchar Medical College, Silchar, Assam, India.

²Ranjit K Baruah, Department of Orthopaedics, Assam Medical College, Dibrugarh, Assam, India

³Russel Haque, Department of Orthopaedics, Assam Medical College, Dibrugarh, Assam, India.

⁴Saurabh Jindal, Department of Orthopaedics, Assam Medical College, Dibrugarh, Assam, India.

Abstract: Paget's Disease is a chronic bone remodelling disease characterised by aggressive bone resorption and imperfect deposition resulting in structurally abnormal bone. It is a common in Caucasian population, and is rare in non-Caucasians, particularly Asians, with only stray case reports from India. We report a case of Paget's Disease of bone from North-east region of India. A 55 year old male presented with bony deformity, pain and tenderness in upper third of his right leg for 1 year. There was no history of trauma or local infection. Blood investigations revealed raised ESR, CRP and Alkaline phosphatase. X-Rays revealed bowing and two healed fracture lines at upper third of tibia. Patient was operated where internal fixation with Inter-locking nail was done after deformity correction and removal of pathological tissue. Histopathology report confirmed Paget's Disease. Calcium and vitamin D supplementation and medical treatment with Risedronate was started and it resulted in improvement in pain and reduction of serum Alkaline phosphatase levels. In conclusion, genetic factors as well as environmental triggers have been implicated in etiology of Paget's Disease. Though rare, it can occur in Indian population as well, so a high index of clinical suspicion is necessary for diagnosis.

Keywords: Paget's Disease; Intra-medullary Nail; Osteitis Deformans; Deformity Correction.

I. Introduction

Paget's disease is a chronic non-hormonal bone remodelling disease characterised by aggressive bone resorption and imperfect deposition [1,2] resulting in a patchwork of structurally abnormal bone with extensive vascularity and fibrous tissue in the marrow [3]. It is commoner in males [4] and rare before the age of 55 years, but increases in prevalence thereafter with advancing age [5,6]. Though it is usually asymptomatic and diagnosed incidentally, most clinical symptoms are skeletal [1,7]. It is a common disease in Caucasian Population, and rare in non-Caucasians, particularly Asians [6,8]. There are only stray case reports from India [9,10]. Here we report a case of Paget's Disease from North-Eastern part of India.

II. Case Presentation

A 55 year old male presented with pain, difficulty in walking and bony deformity in his right leg for past one year. The pain was continuous in nature both on rest and slightly increased on weight bearing. There was no history of trauma or local infection. Examination revealed obvious anterior bowing deformity of upper third of right leg (Fig 1) along with warmth and tenderness in the same region. Bony transmission was found to be intact with normal range of motion in the knee. Systemic examination was normal.

Blood investigations revealed raised ESR (70 mm AEFH), CRP (34 ng/ml) and Serum Alkaline phosphatase (1057 IU/L; normal range: 115-359 [11]) with normal serum protein electrophoresis and liver function test. X-Rays revealed bowing at upper one third of tibia, alongwith osteosclerosis and osteolysis and evidence of two healed fracture lines in the same region (Fig 2). A skeletal survey failed to detect any further obvious skeletal lesion. A Bone scan could not be done due to non availability of facility.

Patient wanted relief from pain and deformity correction. Patient was thus taken to operation theatre and placed in supine position with knee flexed over the end of the table. A medial longitudinal skin incision was given from the inferior border of patella to the tibial tuberosity, and extended distally for 3-4 cm to expose the deformed site. Pathological tissue was removed from deformed site and sent for HPE. Freshening of margins and realignment of the axis with osteotomy was done with further fibular corticotomy and excision of about 1 cm of fibula at two different levels. Without reaming, Internal Fixation with intra-medullary Inter-locking Nail was done followed by bone grafting and wound closure over a drain (Fig 3). Good range of knee joint movement and normal alignment of the limb achieved (Fig 4A and B)

Histopathology report showed thick, irregularly shaped bone trabeculae with variable sizes of osteoblasts with eccentric nuclei and few large multinucleated osteoclasts thus confirming Paget's disease (Fig

5A and B). Calcium and vitamin D supplementation and medical treatment with Risedronate 30 mg daily was started and it resulted in improvement in pain and reduction of serum alkaline phosphatase levels.

III. Discussion

The distribution of Paget's Disease throughout the world varies according to race and geographic deviations. They vary in populations by migration or by breach of geographic isolation [3]. It is common in Caucasian population and thus a relatively common disease in Australia, New Zealand, North America and most European countries, [12] but it has a low incidence in Scandinavia and Africa, and is extremely rare in Asian countries specially India [3,13]. Although the precise pathophysiology of PDB is still not certain, ethnic differences suggest a genetic factor in its aetiology. It is inherited as an autosomal dominant trait with high penetrance [13]. Further, PDB susceptibility loci affecting specific genes have been recently identified, such as *SQSTM1*, *VCP*, valosin-containing protein gene (which encodes p97), and sequestosome1 (*SQSTM1*) gene (which encodes p62) [11] *TNFRSF11A*, and *TNFRSF11B*, the two latter ones encoding NFkB-RANK and osteoprotegerin (OPG), respectively [14]. Viral infections could be another, because viral nuclear inclusions have been observed in osteoclasts from affected patients. Also importantly, an environmental impact has been considered, because of the geographic and ethnic variation.

Bone that is remodelled due to Paget's Disease becomes enlarged, disorganized in structure, and weakened. Patient usually present with complains of pain(80%), skeletal deformity(15%), and pathological fracture (9%) due to structurally inferior bone and neurological compression syndromes due to bony expansion [1,4]. The bowing deformity is usually due to stress fractures that heal over time. Pagetic bones have increased diameter and have areas of sclerosis and lysis. If vertebra is involved it causes sclerosis at the periphery and lysis at the centre, the so-called 'picture frame' appearance [13].

Complications include pathological fractures and malignancy (incidence <1 %; osteosarcoma mainly). The treatment of such fractures is challenging due to increased rate of delayed union, non-union, and malunion in pagetic bone fracture [12]. Open reduction and internal fixation of fractures has been recommended to prevent such complications. The goal of operation of such fractures is to restore a normal axis in the limb and to allow early mobilisation preventing further weakening and mineral loss. Good fracture healing along with correction of mild deformities with the use of intramedullary nailing has been reported. It does not require the extensive exposure of plate and screw and is biomechanically more stable [15,12]. Intramedullary nailing may be technically difficult due to abnormal mechanical axis and obliterated medullary canal in deformed pagetic bones. However in mild deformities as was our case, it can be performed with osteotomy to realign the axis.

To conclude, Paget's disease is rare in Indian population, therefore a high index of suspicion is necessary to make an accurate diagnosis. Deformity and pathological fractures in such cases can be successfully treated with intra-medullary nailing. Concurrent therapy with bisphosphonates gives good outcome and regular monitoring of serum alkaline phosphatase is necessary.

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Figures



Figure 1- Anterior bowing deformity involving proximal third of the leg.



Figure 2 - Radiological confirmation of bowing with healed fracture lines.



Figure 3 - Xray showing realigned bone after short fibular excision, tibial osteotomy and fixation with intramedullary nail



Figure 5- Normal alignment of limb with healed scar and good range of joint movement.

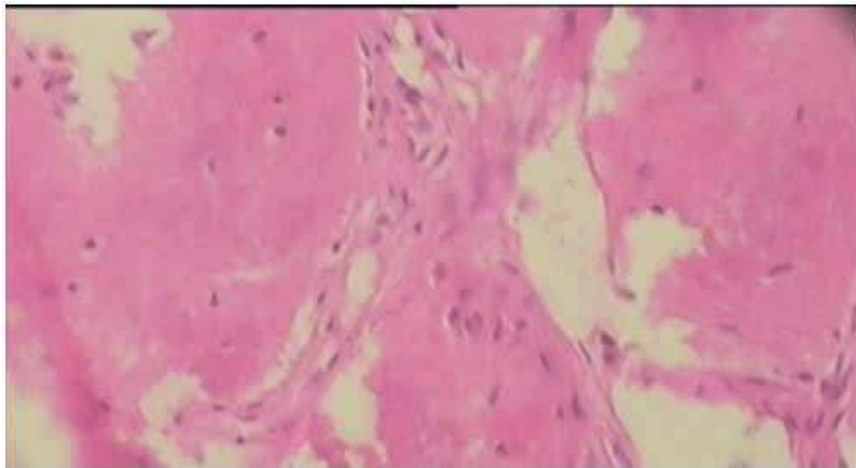


Fig 5A

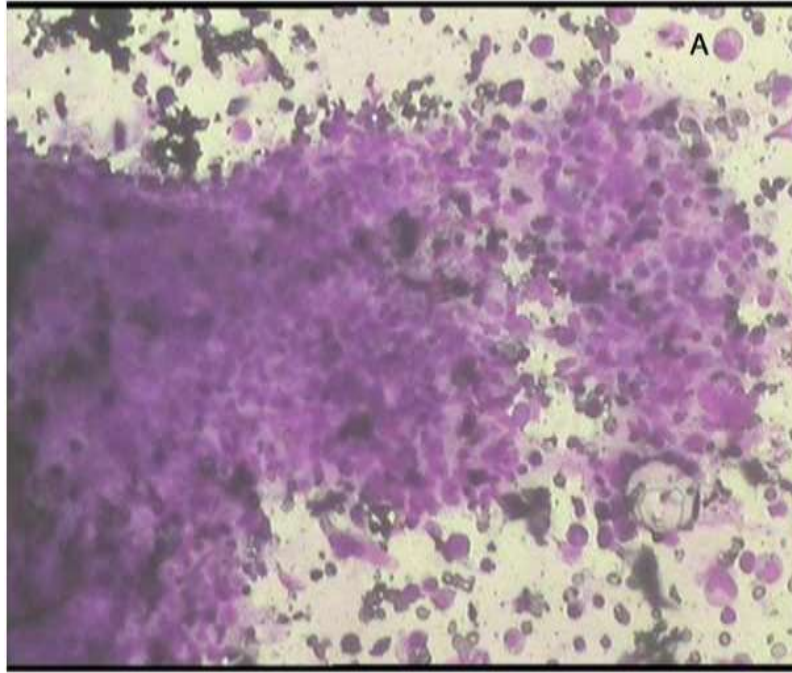


Fig 5B

Fig 5A and 5B: Histopathology report showed thick, irregularly shaped bone trabeculae with variable sizes of osteoblasts with eccentric nuclei and few large multinucleated osteoclasts thus confirming Paget's Disease.