

Case Report

SURGICAL TREATMENT FOR UNUSUAL BIFOCAL FRACTURE OF PAGETIC FEMUR: A CASE REPORT AND CONCISE REVIEW OF LITERATURE

Simone Cerbasi ¹, Giuliano Lattanzi ², Liverani Luca ², Donato Carola ², Raffaele Pascarella ¹

1. Unit of Orthopaedics and Trauma Surgery, Ospedali Riuniti, Ancona, Italy
2. Clinical Orthopaedics, School of Medicine, Università Politecnica delle Marche, Ancona, Italy

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ABSTRACT

Paget's disease is a chronic metabolic disorder of the bone, characterized by exuberant bone resorption, bone growth, and accelerate remodeling, which can make surgical reconstruction demanding in case of fracture. The intramedullary nail is considered the gold standard in these fractures but due to the prevalence of angular deformity in the Pagetic bone, other devices may be indicated. We present here a case of atypical post-traumatic bifocal fracture in a Pagetic femur, in a 91-year-old man, surgically treated with femur short nail, plate, screws and bone graft. The functional and radiological recovery after 1-year follow-up was excellent despite advanced age.

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1. Introduction

Paget's disease is a progressive and chronic skeletal disorder characterized by focal areas of exuberant osteoclastic resorption followed by a secondary increase in the osteoblastic activity. The result is an atypical skeletal remodeling, leading to bones with excessive and deformed tissue. This bone has low mechanical resistance^{1,2}. For this reason, frequent post-traumatic and pathological fracture is observed³. Despite Pagetic fractures being treated operatively, these lesions are associated with a high incidence of varus deformity, nonunion, delayed union, and increased risk of refracture has been reported³⁻⁵. Surgery may be technically demanding.

Although Paget's disease has a benign evolution and causes bone pain, skeletal deformity and sometimes fractures, in a small percentage of cases it gives rise to malignant degeneration such as Paget's sarcoma and osteosarcoma⁵⁻⁷.

We described a patient who sustained a bifocal fracture of Pagetic femur. This lesion is quite unusual. Internal fixation through femur short nail, plate, screws and bone graft was successful, as demonstrated by good outcome at 1-year follow-up. The patient was informed that data concerning the case would be submitted for publication, and he provided consent.

2. Case report

A 91-year-old man presented to our hospital following direct trauma to his left femur.

On examination, tenderness was present at left hip region, left lower limb was externally rotated, shortened with no wound and distal neurovascular deficit. Anteroposterior and lateral radiographs revealed a bifocal fracture of the femur: subtrochanteric and diaphyseal with saber deformity of the femur (Figure 1).

The patient underwent surgery promptly within 48 hours of hospital admission.

The man had a medical history of Paget Disease since the 1970s, discovered incidentally, arising as asthenia and localized bone pain of the lower limb.

The man was studied and subjected to adequate treatments and care until the 2000s when he decided to stop every treatment for the Paget Disease; which was mainly medical, with zoledronic acid and then bisphosphonates. The man has done laborious work all his life and in recent years he has begun to suffer from a limp due to the disease.

In the operating room, the proximal, subtrochanteric fracture was reduced and fixed using the Gamma3 locking nail system (Stryker®).

Due to pre-fracture deformity and the impossibility of providing a stable osteosynthesis of both fractures using a single longer intramedullary nail, the distal fracture was fixed using Polyax™ Locked Plating System (Zimmer Biomet, Warsaw, IN, USA), and opposing cortico-cancellous bone allograft.

With the patient in supine position on left limb traction, antegrade nailing positioning was achieved following the standard surgery technique. Subtrochanteric proximal closed fracture reduction was performed under image identifier check. A 3 cm incision was made 10 cm proximal to the tip of the greater trochanter following the extension of the femoral bow.

* Corresponding author: Simone Cerbasi, simone.cerbasi@libero.it

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Deep dissection was performed using scissors and gluteus muscle was split along its fibers. Under image identifier control, the entry-point was determined, and the guide wire was inserted using a T-handle. Progressive reaming of intramedullary canal was carried out in order to reduce the forces required to achieve nail introduction. The nail was advanced by hand.

Distal fracture approach was made by distally extending cephalic screw towards the lateral femoral condyle. Deep dissection was developed by detaching the vastus lateralis from the lateral intermuscular septum. Bridge plating principles were respected. We also realized a partial corrective osteotomy for deformity in present Paget disease.

Structural bone allograft was placed alongside the medial aspect of the femur in order to provide a framework for the growth of new living bone and increase the stability (figure 2).



Figure 1. Radiographs of left femur showing subtrochanteric fracture and middle third diaphyseal fracture, with the features of Paget's bone disease: deformity, osteolytic areas, enlargement of the affected bone, cortical thickening, loss of distinction between medulla and cortex, trabecular thickening, osteosclerosis.

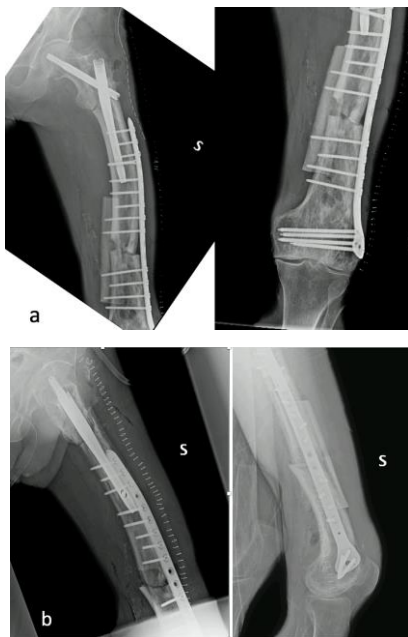


Figure 2. Immediate post-operative, anteroposterior (a) and lateral radiographs (b). Subtrochanteric fracture stabilized by intramedullary nailing and middle third diaphyseal fracture treated by plate and bone allograft with correction of pre-existing deformity.

No functional bracing was used after the surgery. Post-operative care was meant to avoid prolonged immobilization because it can exacerbate osteopenia and could provoke hypercalcemia and hypercalciuria. Hip and knee mobilization were allowed from day 1 after the surgery. Weight bearing was not allowed for 50 days after the surgery, followed by a progressive rehabilitation program. Wound healing was uncomplicated. The bone needed 7 months to be considered radiologically healed.

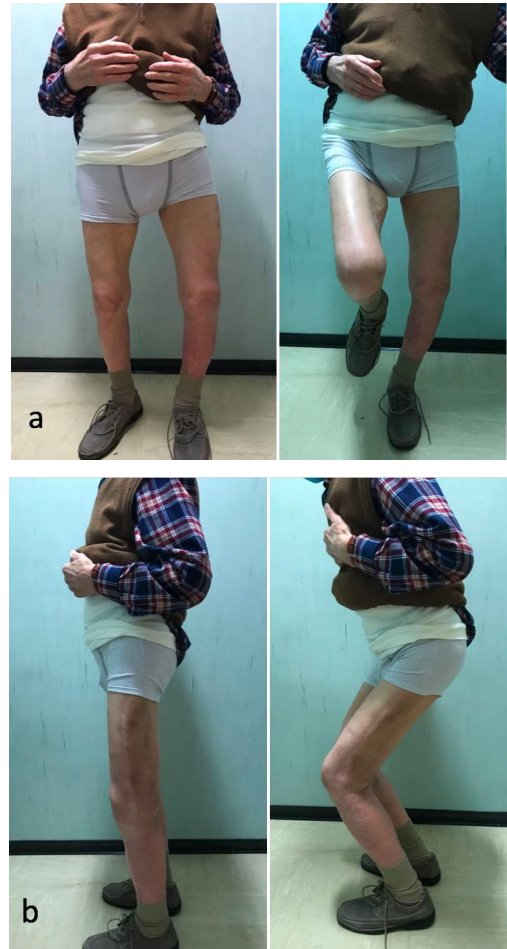


Figure 3. Satisfactory clinical outcome at 1-year follow-up (a,b).

At 1-year follow-up, the patient was evaluated clinically (figure 3) and radiologically (figure 4).

The patient refers no changes in his daily activities compared to pre-trauma. He reported no need for crutches or other aid during his activities. The functional outcome was evaluated with the Italian version of the Western Ontario and Mac Master University (WOMAC) Questionnaire⁸.

The WOMAC is a self-administered disease-specific validated outcome score that evaluates pain (5 items), stiffness (2 items), and physical function (17 items) with a total score ranging from 0-100. Lower scores are associated with better function and less pain and stiffness. Subject obtained 36 points.

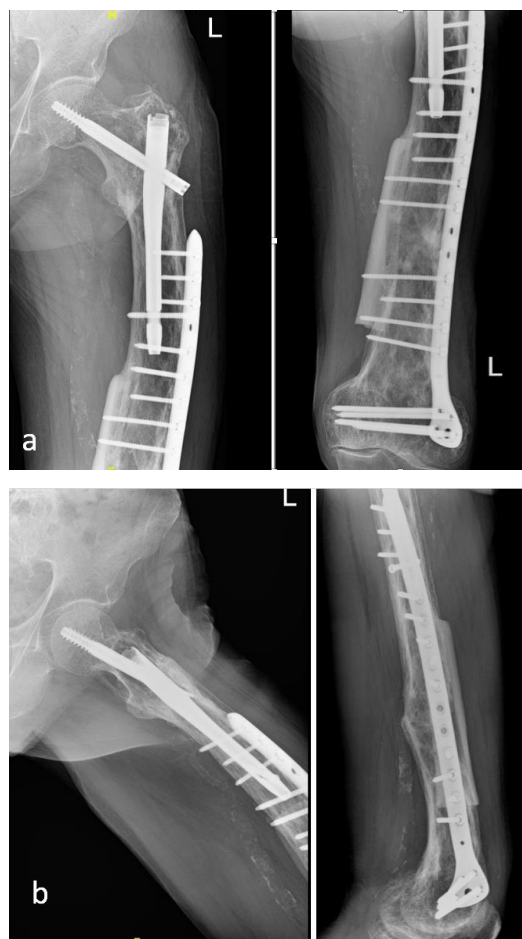


Figure 4. Radiographs (a,b) showing the union fractures at 1-year follow-up.

Furthermore, the Short form health survey (SF-12) questionnaire was used as a general measure of health at final follow up^{9,10,11}. Physical Component Score (PCS-12) was 39.36 and Mental Component Score (MCS-12) was 50.69. Scores were similar to normative values for subjects in the comparable age group and near the 50th percentile of > 75-year-old Italian males¹¹ (Table 1).

Age > 75 years	PCS-12	MCS-12
Mean	39.98	46.89
25th percentile	30.10	38.28
50th percentile	41.26	50.37
75th percentile	50.41	55.96
Standard Deviation	11.63	11.69
Range	11.73 - 62.77	15.63 - 69.39

Table 1. SF-12. Normative values for subjects > 75 years

3. Discussion

Fracture's treatment in Pagetic bone could be difficult and demanding, both for anatomical deformities and delayed union rate and high non-union rate³⁻⁵. The femur is a relatively usual monostotic Paget's disease of bone location^{12,13}, and the subtrochanteric fractures are the most common lesions, followed by the middle third diaphyseal fractures¹⁴.

No randomized trials were known with regard to the treatment of fractures in Paget disease of bone, but, more commonly, the outcomes of surgical treatment have been reported in several recent observational studies¹³ and case reports^{15,16}. Fracture healing normally occurs in many patients, however the clinical results in proximal femoral fractures are poor¹³.

Intramedullary fixation with a short narrow nail to control the angular deformity was successful and did not involve excessive technical difficulties. Many authors have described the successful use of intramedullary nailing^{4,14,15,17}. Shardlow et al⁴ concluded that this device is a good option for the stabilization of acute subtrochanteric or diaphyseal fractures in Pagetic femora, reliable bone stability and good healing results in eleven cases. Recently, Mahajan et al¹⁵ showed that subtrochanteric pathological femur fractures in Paget's disease are better managed with the long PFN nail, achieving good clinical outcomes and fewer complications, which advantages early weight-bearing and mobilization.

The use of long PFN nail prevents the secondary fractures because it allows spanning of the entire pathologic femur. However, attempts to secure absolute fixation by the insertion of a full-diameter long nail are inadvisable where there is any predisposition to bowing of the shaft or narrowing of the medullary cavity. In case of skeletal deformity, or if the fracture is in the lower part of the femoral shaft, other methods and devices are necessary¹.

But it is also reported that open reduction and internal fixation are not always easy if the femur is relatively undeformed. In our situation, the femur's bowing did not allow intramedullary nailing for both the fractures even if is currently considered the best method of fixation in femur pathological fractures^{2,3,5}.

Our case posed problems for the surgeon, specifically distal deformity of the femur with poor bone quality. No flexible intramedullary device was considered stable enough due to the bifocal nature of the fracture with enlargement of the affected bone. Although long intramedullary nailing was possible after osteotomy, we preferred hybrid fixation to increase stability and to give biological support to promote healing of the second fracture. Our decision ended up being to stabilize the proximal fracture with the intramedullary nail, and bridge plate for the distal one. Particular attention was reserved not to leave parts of the femur unarmed in order to prevent future fractures. Bradley et al¹³ reported near certain healing of the fracture, though, a later fracture may occur if the disease 'outgrows' the primary implant" so our purpose is to span the femur all along the length.

Although Paget's disease has a normal biological capacity for fracture healing, which can occur at a regular rate, the time to union is slightly longer than the average of non-pathological fractures¹⁸. Parvizi et al¹⁸ suggested that mechanical instability of the pagetic bone could result in a prolonged course. Bone allograft has been used to provide both mechanical and biological support¹⁶ to promote healing and avoid non-union or malunion with progressive deformity.

In this paper, we presented a technically demanding osteosynthesis. To the best of our knowledge, this is the first report analyzing the treatment strategy of Pagetic bifocal fracture of the femur, and this surgical technique shows reliable skeletal stability and satisfactory healing result. Our pre-operative research focus on understanding the bifocal femur fracture.

In addition, our patient's age could have resulted in an even higher risk of non-union.

Despite the lack of information about bifocal Paget femur fracture, the aim of this case report is to underline how following the basic principles of osteosynthesis could lead to a good result even in a pathological bone in an old patient. Anatomical deformities could require the use of multiple internal fixation devices. Bone allograft could be a useful tool to enhance fracture healing.

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