

REALIGNMENT OF TIBIAL DEFORMITY IN PAGET'S DISEASE A CASE REPORT

J. A. BECERRA, M. LLUSÀ, E. GUERRA, B. R. DENIZ, V. MOLERO, J. NARDI

Paget's disease of bone is a relatively common condition which usually is asymptomatic. Furthermore, there is limited experience in surgical procedures, especially in realignment of severe skeletal deformities. Likewise, there are often relapses in tibial deformities treated with plates. We report a case of a severe tibial deformity treated with multiple osteotomies and stabilized with an unreamed medullary nail.

Keywords : Paget ; tibial deformity ; osteotomy ; realignment.

Mots-clés : Paget ; déformation tibiale ; ostéotomie ; réalignement.

INTRODUCTION

The incidence of Paget's disease ranges between 2 and 4% in the population older than 60 years. Only 5 to 10% of these patients are symptomatic (4).

The main complications of this bone disease are neurologic disorders, pathological bone fractures and joint degeneration. Particular features of pagetic bone (5) and the reduced strength of bone structure (7) are factors that influence the surgical treatment. Furthermore, there is limited experience in surgical procedures, especially in realignment of severe skeletal deformities. On the other hand, conservative treatment of osteoarticular disorders, which involves prolonged immobilization, is also associated with complications(7).

We report a case of a severe tibial deformity realignment in a patient with polyostotic Paget's

disease treated with multiple osteotomies and stabilized with an unreamed medullary nail.

CLINICAL CASE

The patient, a 65-year-old male, had left knee pain. He was diagnosed with Paget's disease 40 years previously, presenting a left tibial deformity. When he was 50 years old, he suffered a proximal metaphyseal pathologic fracture, which was conservatively treated. Additionally he received calcitonin.

The reason for the consultation was left knee pain localized on the medial side, with severe varus tibial deformity (fig. 1). Pain was constant and did not decrease with rest and analgesics. The patient walked with a cane. Walking perimeter was about 300 meters. Knee range of motion was 0/90°. There was no instability or articular effusion. Roentgenographic tests revealed a tricompartmental arthritis of the knee predominantly in the medial femorotibial joint, and a 34° varus deformity at the expense of the proximal tibial metaphysis (fig. 2). Scintigraphy demonstrated increased uptake in the left tibia and sternum. Electromyography was normal, and blood tests revealed high levels of total alkaline

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Fig. 1. — Clinical appearance of the tibial deformity with obvious varus.

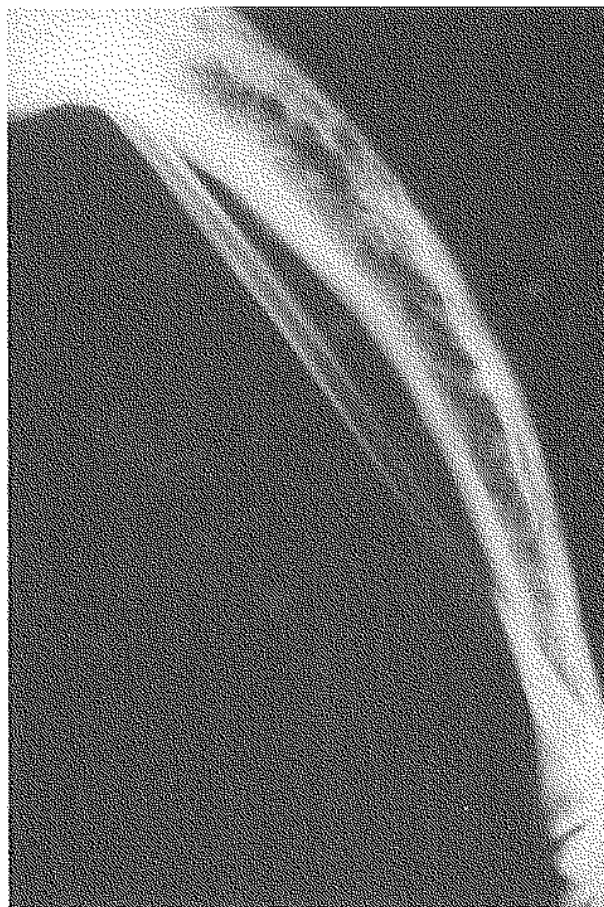


Fig. 2. — Radiograph shows twisting deformity.

phosphatase (2055 with a normal value of 94-280). To evaluate the deformity precisely a CT- scan of the entire length of the tibia was performed, showing a curve of the shaft with an anterolateral convexity and internal rotation of the leg. Incisional biopsy excluded sarcomatous degeneration. Because of the severe tibial deformity surgical correction was performed, leaving for a second surgery a total knee arthroplasty. Through an anterolateral approach over two-thirds of the leg, a triple oblique-angled osteotomy was made, obtaining a good tibial alignment. An unreamed medullary nail locked distally and proximally was used to stabilize the osteotomies. A midshaft peroneal osteotomy had been done previously (figs. 3a, b).

Calcitonin treatment was started. An external leg orthosis was used until initial signs of bone healing, which were observed five months after surgery. The correction of the deformity produced good pain relief and functional improvement. Joint motion was 0/100°, with a 10° residual varus (fig. 4). After a follow-up of 4 years the patient remains asymptomatic, and no recurrence of the deformity has been observed.

DISCUSSION

Lower limb Pagetic deformities that produce a severe functional limitation are relatively uncommon, and the surgical experience with correction techniques is limited.

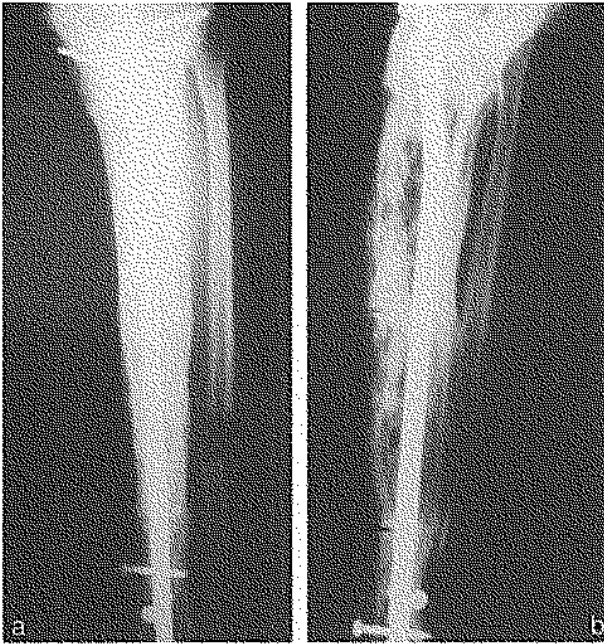


Fig. 3a. — AP radiograph after surgery.
Fig. 3b. — Lateral radiograph after surgery.

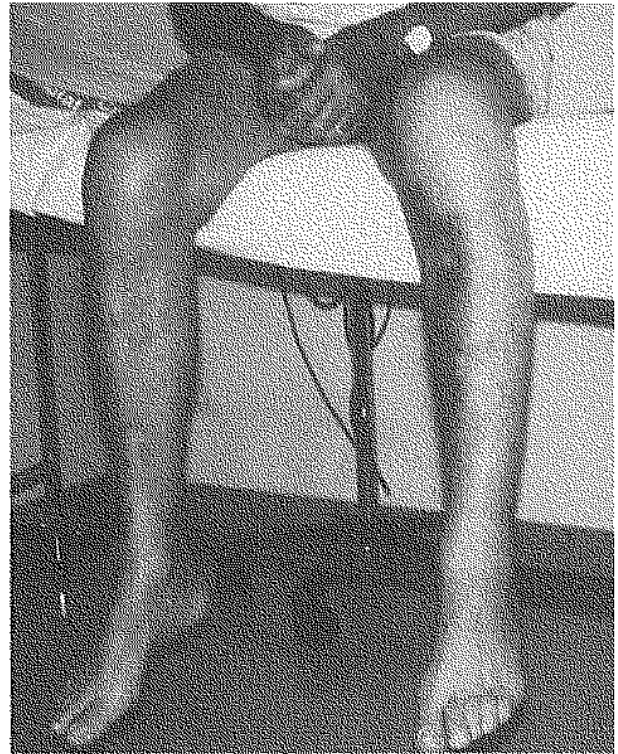


Fig. 4. — Postoperative clinical aspect.

Pagetic bone deformities mainly involve femur and tibia, with equal frequency. The type of deformity is variable. Thus, the tibia is characterized by an anterolateral convexity with internal rotation, without fibula involvement. On the other hand, the femur is affected with an anterolateral convexity associated with external rotation. When Paget's disease involves both ipsilateral segments rotational deformities are neutralized, giving a parenthesis aspect to the extremity. Pain could result either from stress fractures at the apex or from degeneration of neighboring joints produced by mechanical overloading as a result of alteration of the limb axis.

The elective treatment of Pagetic deformities is corrective osteotomy. In 1965 Merle d'Aubigné and Witvoet (6) described plane oblique osteotomies, abandoning the wedge osteotomies used until then. Plane oblique osteotomy, although more difficult and technically demanding, avoids limb shortening and provides a wider contact between bone surfaces (2, 6). Internal fixation of the

osteotomy improves anatomic and functional outcomes, decreasing at the same time cardiopulmonary and thromboembolic complications by early mobilization (1). Moreover owing to early mobilization, phosphocalcic metabolism alterations are reduced (9). In the femur, fixation with a reamed endomedullary nail is the most commonly used technique (2, 3, 6, 9). In the tibia the osteotomies used to be fixed with plates (1, 2, 3, 6, 9), although on a few occasions elastic endomedullary nails were preferred (3, 7). In addition, therapy with calcitonin pre- and postoperatively is recommended in order to decrease Pagetic bone bleeding.

Pain abolition is common after osteotomy healing. The healing period of pagetic bone seems to be the same as for normal bone (6, 9).

One of the most important aspects in the surgical treatment of the skeletal deformity is the moment of the correction: neither too early to avoid recurrence, nor too late to avoid degeneration arthropathy of neighboring joints. It is recommended to

treat the patient medically prior to surgery in order to reduce the activity of the disease (2, 3, 6, 8).

Recurrence of the deformity is related to the degree of correction and the stability of the osteosynthesis method. Recurrences are frequently reported in the pagetic tibia. This finding may be related to the more accurate and stable correction which is obtained in the femur, owing to the use of endomedullary nail fixation. On the contrary, plate osteosynthesis often used in the tibia is associated more commonly with material failure in relation to load.

In the present case medullary nailing was preferred to stabilize the three-level plane oblique osteotomies. An unreamed nail was used in order to limit damage to endosteal vessels and pagetic bone bleeding. An important advantage of using an interlocking nail is that rotation of osteotomized segments can be prevented, making bone fragment alignment easier. As a result, this osteosynthesis gives higher stability than plates, decreasing recurrences and material failures.

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SAMENVATTING

J. A. BECERRA, M. LLUSÀ, E. GUERRA, B. R. DENIZ, V. MOLERO, J. NARDI. Realignatie van een tibiale misvorming bij ziekte van Paget.

De ziekte van Paget van het skelet is een relatieve frequente aandoening, meestal asymptomatisch. Bovendien is de ervaring met chirurgische technieken beperkt, vooral wat het realigneren van ernstige skeletmisvormingen betreft.

Er bestaan ook frequente recidieven van de tibia afwijking na plaatfixatie. De auteurs beschrijven het geval van een ernstige tibiamisvorming behandeld met multiple osteotomieën en gestabiliseerd door middel van een centro-medulaire pen, zonder voorboring, met proximale en distale vergrendeling.

RÉSUMÉ

J. A. BECERRA, M. LLUSÀ, E. GUERRA, B. R. DENIZ, V. MOLERO, J. NARDI. Réalignement d'une déformation pagétique du tibia : présentation d'un cas.

La maladie osseuse de Paget, relativement fréquente, est habituellement asymptomatique. La correction chirurgicale de déformations osseuses majeures n'a donné lieu qu'à peu de publications. Les déformations tibiales réalignées et fixées par des plaques ont souvent récidivé. Les auteurs présentent un cas de déformation majeure du tibia qui a été corrigée par des ostéotomies étagées, fixées par un clou intra-médullaire non alésé et verrouillé.