Hip Arthroplasty in Paget’s Disease: Case Report and Literature Revision

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Abstract

Introduction: Paget’s disease of bone represents a relatively frequent pathology affecting the elderly. In order to appropriately treat fractures of the femoral neck in these patients, it is mandatory to correctly diagnose the disease and to thoroughly study the anatomy to choose the correct prosthetic implant.

After thorough pre-operative planning, we performed hip arthroplasty in an 88 years old PDB patient. We chose Smith and Nephew REDAPT Modular Shell and a Smith and Nephew POLARSTEM stem, even though a REDAPT stem was available. We then planned a six weeks, three months and six months follow up and we collected data about radiological findings and clinical outcome each time.

X-rays findings were always normal with no sign of aseptic loosening, infection or heterotopic ossification. The HHS improved from 60.2 points at six weeks to 82 points at three months.

The REDAPT system allows to obtain excellent stability of the implant even in Pagetic bone and it may induce excellent secondary bone ingrowth. In our opinion, multicentric studies are needed to improve our ability to diagnose PDB and properly perform joint arthroplasties in patients affected by PDB.

Keywords: Paget’s disease; Hip replacement; Pelvis deformity; Bone disease; Femoral fracture

Introduction

Paget’s Disease of Bone (PDB), firstly described in 1877 by Sir James Paget, is characterized by altered bone metabolism with increased osteoblastic and osteoclastic activity that leads to an increased bone production resulting in more vascular, less compact and mechanically weak bone, thus being more susceptible to fractures [1]. The osteoblastic and osteoclastic activity is not constant, and it varies over time depending on disease activity. The disease affects the skeleton asymmetrically, involving more frequently the pelvis (60.3%), spine (35.1%), femur (32.3%), skull (22.2%) and tibia (15.5%) [2]. The deformities that we find in advanced stages of PDB include coxa vara, acetabular protrusion and anterolateral femoral bowing [3].

PDB represents the second most frequent metabolic disease of bone after osteoporosis [4]. The prevalence in Italy is about 1% [2], with higher prevalence between males compared to females, in accordance with prevalence in the USA [5], but lower than that of UK which is about 5% [6]. Recent findings suggest a reduction trend in prevalence of the disease in the last years [7].

Orthopedic issues related to PDB are pathologic fractures, delayed fracture union, skeletal deformity, and chronic bone pain [8]. At last, osteosarcoma is the most feared issue related to PDB. Even if it affects mostly patients in the second and third decade of life, it may affect 1% of long-standing PDB patients with polyostotic disease [9].

Joint replacement might be difficult in patients affected by PDB due to bone deformities, and the difficult may increase if the procedure has to be performed on a fractured bone, because there is little time to appropriately study and treat the patients. In our experience, we had the opportunity to perform a hip replacement on a patient affected by femoral neck fracture who wasn’t aware of being
affected by PDB too.

**Case Presentation**

The patient, 82 years old female, came to our attention because of sudden pain and loss of function of the left hip while standing up from the sofa. When interviewed, the patient reported the onset of mild pain of the left hip a couple of days before. The left lower limb appeared shortened and extra-rotated. Given the radiographic image of great deformity of the pelvis (Figure 1), a CT scan was performed in the emergency department (Figure 2a, 2b).

Blood tests performed during the first day of hospitalization showed serum levels of ALP of 188 U/L (normal value 33 U/L to 98 U/L), and normal serum levels of AST and ALT, clinical finding that was highly suspicious for PDB. The patient wasn’t fully aware of her condition, in fact she reported three childbirths normally completed. She reported the dysmetria of her lower limbs to her general practitioner who suggested the use of a wedge in her left shoe.

The bone deformity affected the left side of the pelvis with resulting asymmetry of the pelvis, reduction of the transverse diameter and one oblique diameter with consequent reduction of inner pelvic volumes, while the anteroposterior diameter was preserved.

In order to correctly diagnose PDB, the patient underwent body scintigraphy with HDP Technescan (Technetium-99m oxidronate) that showed hypercapitation restricted in the left side of the pelvis, meaning that the patient was affected by monostotic PDB. We then decided to proceed with the hip arthroplasty procedure.

We performed a direct lateral approach with detachment of gluteal musculature according to Hardinge technique. The good quality of tendons allowed us to perform a clean procedure. We identified the tendon of the gluteus minimus that appeared retracted and partially calcified. The acetabular reaming has been performed with care using increasing diameter motor powered acetabular reamers. In order to fill the bone gap we decided to proceed with an autologous bone graft. The acetabular cup choice was a REDAPT MODULAR shell (Smith &Nephew Memphis – TN - USA). We obtained a good peripheral rim fit with a 54 mm size cup, and we improved the stability of the implant using 1 conventional screw and 5 locking screws as allowed by the REDAPT system. The normal aspect of the femur and the correct positioning of the acetabular cup allowed the implant of a
PDB, it is mandatory to identify the correct implant model. Literature 
investigation of PDB complications such as osteosarcoma or spinal 
of little use to diagnose the disease [12], while they are useful for the 
study its extent [11]. It is more sensitive compared than plain X-rays, 
underwent radio marked bone scintigraphy with Technetium-99m 
this finding is not enough to diagnose PDB, which is why our patient 
reliable screening marker, largely available and low-cost. As observed 
by Eekhoff et al. [10], while PDB diagnosis is made on 1 patient over 
43 with normal serum ALP levels, 1 over 5 patients with high serum 
ALP levels had PDB. The relative risk of developing PDB is almost 
11 times higher in patients with high level of serum ALP. However, 
this finding is not enough to diagnose PDB, which is why our patient 
underwent radio marked bone scintigraphy with Technetium-99m 
oxidotrat, which is the most valuable exam to diagnose PDB and 
study its extent [11]. It is more sensitive compared than plain X-rays, 
but it may be negative in sclerotic lesions or whether the disease is not 
active. We didn’t perform CT scans or MRI scans because they are 
of little use to diagnose the disease [12], while they are useful for the 
investigation of PDB complications such as osteosarcoma or spinal 

Once diagnosed the condition or if there is high suspicion of 
PDB, it is mandatory to identify the correct implant model. Literature 
is still debated about the advantages and disadvantages of cemented 
and cementless implants in PDB patients. At the beginning only 
cemented implants were used, but several studies reported higher 
incidence of symptomatic and asymptomatic radioluencies around 
the bone-cement interface that might lead to higher rates of aseptic 
loosening [13]. Some authors believe that hypervascularity may 
impair the ability to achieve a dry bed on cancellous bone for cement 
interdigitation [3]. Cementless implants showed great outcomes 
both on the acetabular side and on the femoral side at a medium-
term follow up with HHS between 78/100 and 100/100 [14] and at 
a long-term follow-up with HHS between 72/100 and 90/100 [15]. 
However some authors are skeptical about the use of cementless 
implants on Pagetic bone because it is unclear if the altered bone 
quality may impair bone ingrowth [13]. Therefore, in addition to a 
good peripheral rim fit it is appropriate to increase the stability of the 
acetabular cup by inserting locking or compression screws depending 
on the model [16]. In any case, revision rates with aseptic loosening 
appear to be higher in the cemented implant cohort [1].

PDB patients present higher risk of complication during the 
surgical procedure and in the postoperative period, especially blood 
loss due to the high hypervascularity of Pagetic bone. Heterotopic 
ossification and aseptic loosening are more likely to develop in this 
kind of patient too [17]. Moreover, late disease activity might lead 
to rapid periprosthetic osteolysis and to premature implant failure 
[18]. Some authors, in order to reduce the incidence of intraoperative 
bleeding and aseptic loosening, advocate pre-treatment of the 
patient with bisphosphonate [16,19]. Patient should start treatment 
six weeks before surgery and disease activity should be monitored 
with measurement of serum ALP [1,20]. This strategy unfortunately 
doesn’t apply on trauma patients who need surgery in short time. In 
order to prevent heterotopic ossifications we treated the patient with 
indomethacin, whose effectiveness is well documented [21,22] at the 
dosage of 50 mg twice a day for 15 days post-operatively. The patient 
did not develop HO.

Joint replacement surgery is also linked to pulmonary 
complications such as pulmonary embolism and pneumonia [23]. 
Hernandez et al. [24] found that PDB patients had higher risk of 
pulmonary complications compared to control groups. This leads to 
longer lengths of stay with higher costs for NHS and higher risks of 
hospital acquired infections for the patient.

When we deal with joint replacement surgery, in our opinion it’s 
mandatory to carry out a thorough preoperative planning, especially 
for most complex cases. We rely on Traumacad software (Brainlab 
AG – Munich – Germany), that allows to estimate implant sizes and 
surgical gestures to restore correct joint biomechanical parameters.

Discussion

Usually, patients come to our attention aware of being affected by 
PDB. If not, as in our case, it’s mandatory to diagnose the condition 
in order to study its extension and severity. Serum ALP is the most 
reliable screening marker, largely available and low-cost. As observed 
by Eekhoff et al. [10], while PDB diagnosis is made on 1 patient over 
43 with normal serum ALP levels, 1 over 5 patients with high serum 
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11 times higher in patients with high level of serum ALP. However, 
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Once diagnosed the condition or if there is high suspicion of 
PDB, it is mandatory to identify the correct implant model. Literature
excellent secondary bone ingrowth.

At last, when we perform hip arthroplasty in a PDB patient, it is mandatory to preserve as much bone stock as possible in order to allow potential future revision procedures due to aseptic loosening that in this kind of patients are more likely to happen.

References


