



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

Giaretta S^{1*}, Spoletini P², Marcanzin M³, Colombini AG², Giarola D², Siliquini E², di Benedetto P^{3,4} and Momoli A¹

¹Department of Orthopedic and Traumatology, San Bortolo Hospital, Italy

²Department of Orthopedics and Trauma Surgery, University of Verona, Italy

³Department of Orthopedics, University Friuli Centrale, Italy

⁴Department of Medicine, University of Udine, Italy

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.

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*Correspondence:

Stefano Giaretta, Department of Orthopedic and Traumatology, San Bortolo Hospital, Viale Rodolfi 37, 36100, Vicenza, Italy,
E-mail: giarettavicenza@gmail.com

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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 e 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 hypercaptation 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



Figure 1: X-rays showing left femoral neck fracture and great deformity of the pelvis.

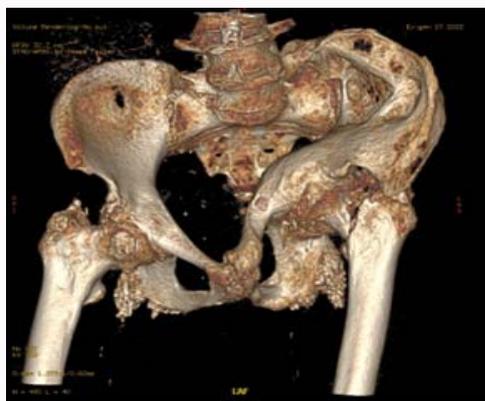


Figure 2a: 3D CT scan of the pelvis performed at the emergency department.

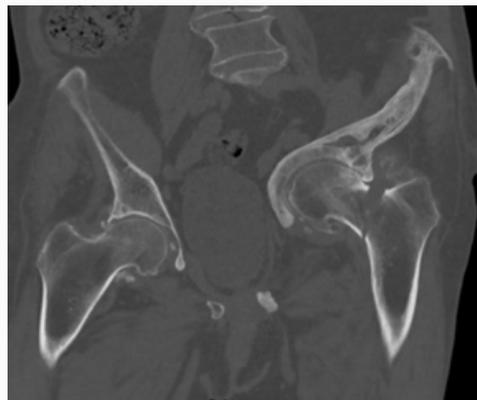


Figure 2b: CT scan of the pelvis performed at the emergency department.



Figure 3a: Post-operative X-rays AP view.



Figure 3b: Post-operative pelvis X-rays AP view.

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



Figure 4: Three months follow up Pelvis X-ray.

POLARSTEM lateralizing size 5 stem (Smith & Nephew Memphis – TN - USA). In case of difficulty in obtaining a correct orientation of the prosthetic neck, a Wagner type stem (REDAPT Smith & Nephew Memphis – TN - USA) was available (Figure 3a, 3b).

Given the risk of osteosarcoma, we requested histological study of the femoral head and of the femoral cancellous bone that didn't show any sign of malignant transformation.

After an appropriate stay in hospital during which the patient underwent physiotherapy to recover the Range of Motion (ROM) of the hip and to maintain the ROM of knee and ankle, the patient was discharged from hospital. We suggested the beginning of physiotherapy with gait training and a clinical and X-ray examination at six weeks since the surgical procedure.

At the six weeks follow up, X-rays didn't show any sign of radiolucency, the scar was normal, and the patient didn't show or report any sign of infection. The ROM was limited with a Harris Hip Score (HHS) of 60.2.

At the three months follow up, X-rays were still normal (Figure 4) and the patient was able to walk using a cane. The ROM was complete and free of pain with a HHS of 82. We suggested then to gradually cast off the cane and a new follow-up at 6 months since the surgical procedure.

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 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 oxidronat, 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 stenosis [11].

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 radiolucencies 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.

Conclusion

PDB is a rare condition that requires thorough study and preoperative planning to achieve satisfactory outcomes. Unfortunately, the experience of one case is not enough and multicentric studies should be carried out to optimize therapeutical strategies.

The short follow up of just 3 months allows us to evaluate clinical and radiographical results at a short-term follow up and doesn't allow the treatment of long-term complications such as aseptic loosening due to disease activity.

Our experience with the Smith and Nephew REDAPT system makes us trust that the great stability of this implant may induce

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.

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