

Total hip arthroplasty and Paget's disease: case report and literature review

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Abstract. Paget's disease (PDB) is a chronic osteopathy more common in male, Caucasian, European population, after the age of 50 years, that can lead to bone deformities. A challenging surgical solution for affected hip with severe hip osteoarthritis is total hip replacement (THA). We describe a case of THA in a 71-year-old patient with PDB and we present a literature review. In particular we find out that more studies comparing cementless THA with cemented one are necessary, in order to understand if one implant is better than the other. (www.actabiomedica.it)

Key words: Paget, osteopathy, total hip arthroplasty, hip replacement, hip arthritis

Introduction

Paget's disease of bone (PDB) is a chronic, non-metabolic, benign bone disease characterized by an unbalance between osteoclastic and osteoblastic activities (1).

This pathology is more common in male, Caucasian, European population, after the age of 50 years (2).

Focusing on total hip arthroplasty (THA), in Paget patients there is a higher risk of heterotopic ossification and Paget bone's deformities may make surgery more challenging. In fact, concerning the acetabulum, sclerotic bone could make acetabular reaming and cement interdigitation so difficult that cementless implant are recommended with the use of adjuvant acetabular screws. Moreover, deformities may lead to acetabular protrusion, so it could be awkward to put acetabular component in the correct position, therefore antiprotusio cage could be indicated. Considering the femoral component, coxa vara is the most frequent deformity found, so realignment osteotomies can be necessary to restore the correct limb axis (3).

We describe a case of THA in a 71-year-old patient with PDB and we present a literature review.

Case report

A 71-year-old man, with a diagnosis of Paget bone disease from 2019, was operated in March 2021 with cemented total hip arthroplasty, because of severe coxarthrosis (Figures 1, 2, 3). He had started a therapy with an oral bisphosphonate (Difosfonal 100 mg 1 vial every week) in July 2019 and from October 2020 he underwent a treatment with zoledronic acid 4 mg, prescribed by the referral center for Paget's disease.

At preoperative clinical examination, he suffered from inguinal pain, with compromised range of mobility (Flexion 100°, Intrarotation 0°, Extrarotation 30°), quadricipital and gluteal muscular hypotrophy and severe limitations of his quality of life.

Pre-surgery, because of the high risk of bleeding due to the Paget's disease, the patient performed recommended prophylaxis with bisphosphonates and in addition to this, to minimize bleeding, we added the usual prophylaxis with tranexamic acid: 1 gram pre operative and 1 gram 6 hours post operative. Pre-operative optimization of Haemoglobin level could compensate intra-operatively blood loss;

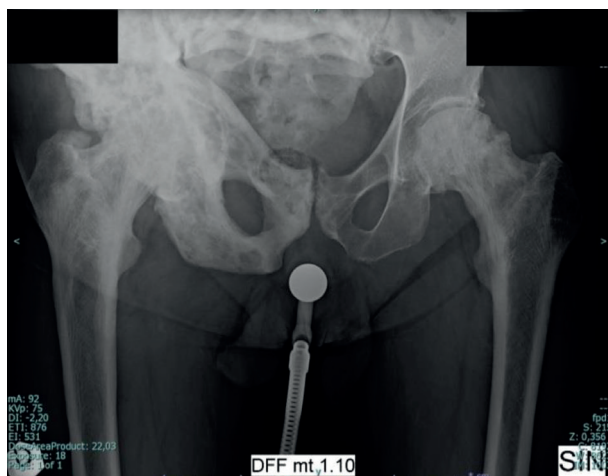


Figure 1. Paget's bone osteopathy. Right hip: pre operative x-ray.

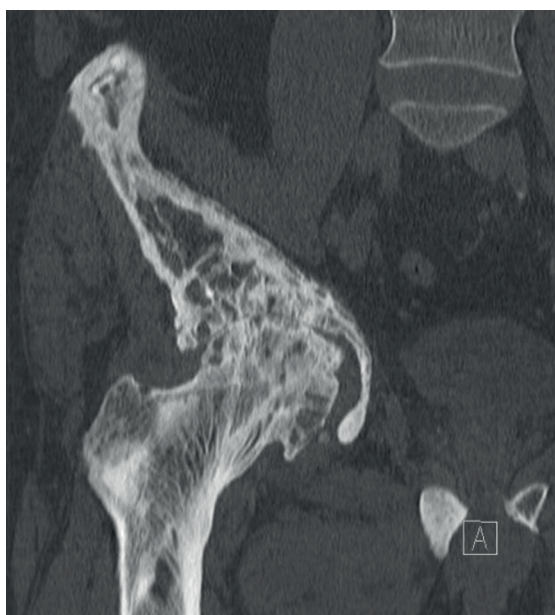


Figure 2. Paget's bone osteopathy. Right hip: pre operative CT scan.

in our case the patients had high level of pre operative haemoglobin (15,2 gr/L), so no therapy was necessary.

During surgery, the patient was placed in lateral decubitus to allow Moore's posterolateral approach to hip and we had implanted press-fit Delta Multihole (Lima) acetabular cup, fixed by 9 bone screw, a polietilene liner, a cemented lateralized



Figure 3. Paget's disease right hip CT 3d reconstruction.

femoral stem C-MAX with ceramic femoral head. (Figure 4).

X-rays were done after surgery. (Figures 5, 6)

In the post operative we monitored ALP levels because this is an indicator of active disease and if this was high biphosfonate treatment should continue. In our case ALP was stable.

Furthermore we started prophylaxis against Heterotopic ossification using indomethacin (25mg three times a day for 21 days).

Post operative hemoglobin levels didn't require blood transfusions during hospitalization.

The day after the operation, he started physical therapies in our orthopaedic service, followed by 3 weeks of rehabilitation protocol in a rehabilitative hospital. X-rays and clinical evaluations were done after 1, 3 and 6 months. At 1 month the patient showed no pain and he was able to walk without crutches. At 3 months follow up the patient showed complete range of motion and x-ray demonstrated excellent osteointegration of the THA.

At the last evaluation (12 month follow up) we observed full rom recovery with excellent Harris HIP score (post operative 91,70 : slight pain, no use of support devices, no limp, put on shoes and socks with moderate difficulty, flex 100°, abduction 15°, externa rotation 10°. Adduction 10°). The pre operative HHS was 35.70. (Figure 7).

12 months x-ray showed no differences with previous control. (Figures 8, 9).

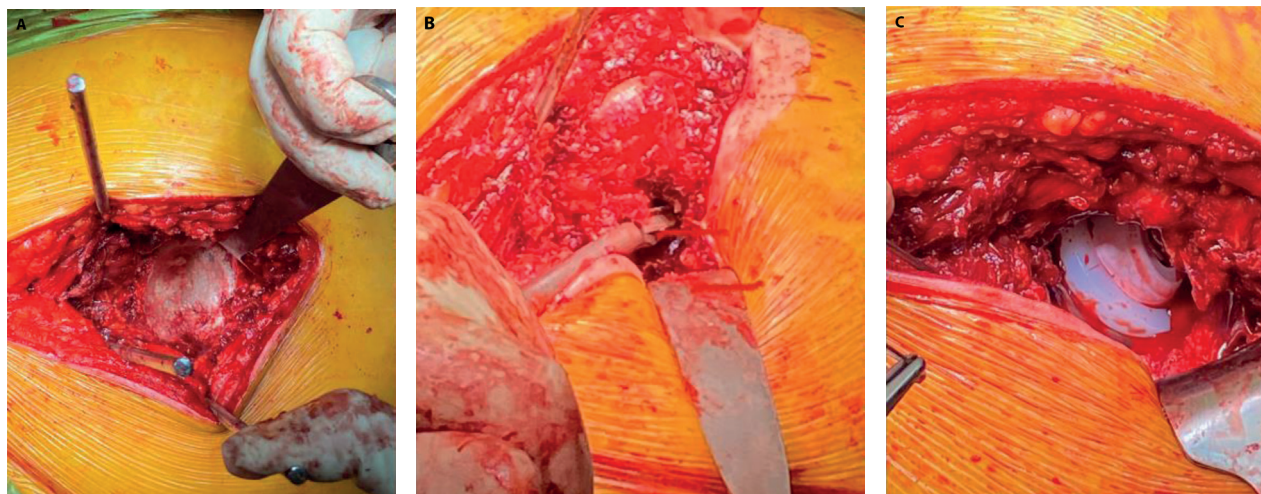


Figure 4. Intraoperative pictures show sclerotic bone. It was necessary to use a hand reamer to prepare the femoral canal.

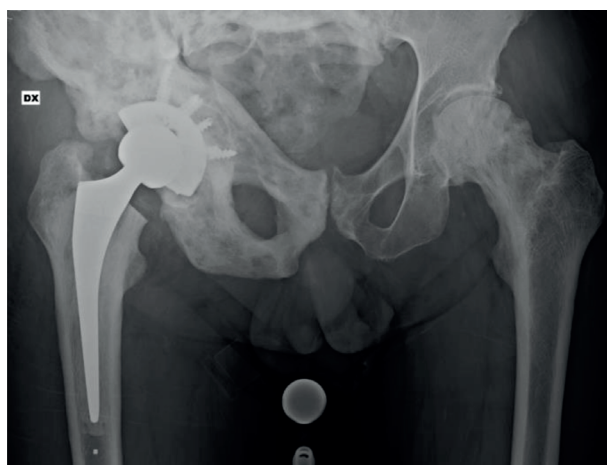


Figure 5. Post operative x-ray antero-posterior view.

Discussion

In Paget's disease we can detect 3 different phases: At the beginning there is a predominant osteoclastic activity, followed by a mixed phase in which osteoclastic and osteoblastic activities are increased and equivalent and a third phase, when osteoblastic activity is predominant.

As a result, we can find osteolysis lesion, such as cavities, marrow fibrosis, hypervascular bone and a disorganized formation of sclerotic woven bone, which is typical lamellar, weaker and more susceptible to deformities and fractures.

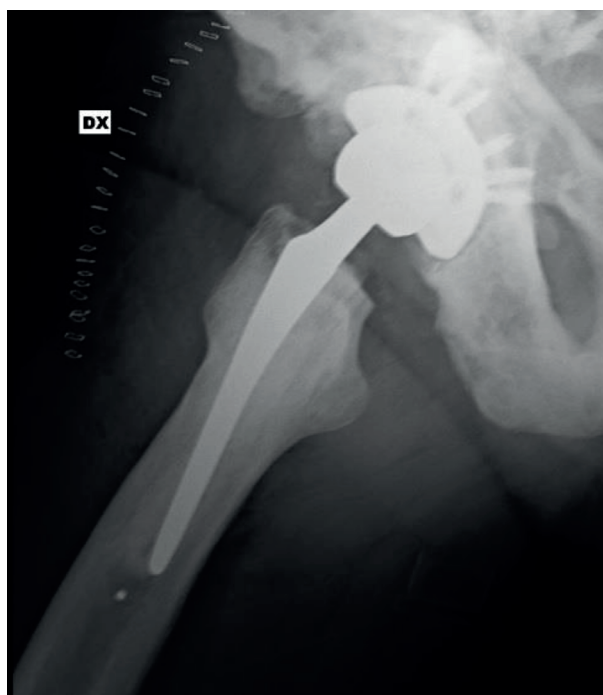


Figure 6. Post operative lateral view.

The clinical course of pathology is difficult to clarify, because its evolution is slow and the most frequent symptom is chronic, moderate bone pain which is, nevertheless, very aspecific.

Complications can occur as first manifestation of PDB, such as fractures, early onset osteoarthritis,



Figure 7. 12 month follow up: full active flexion.

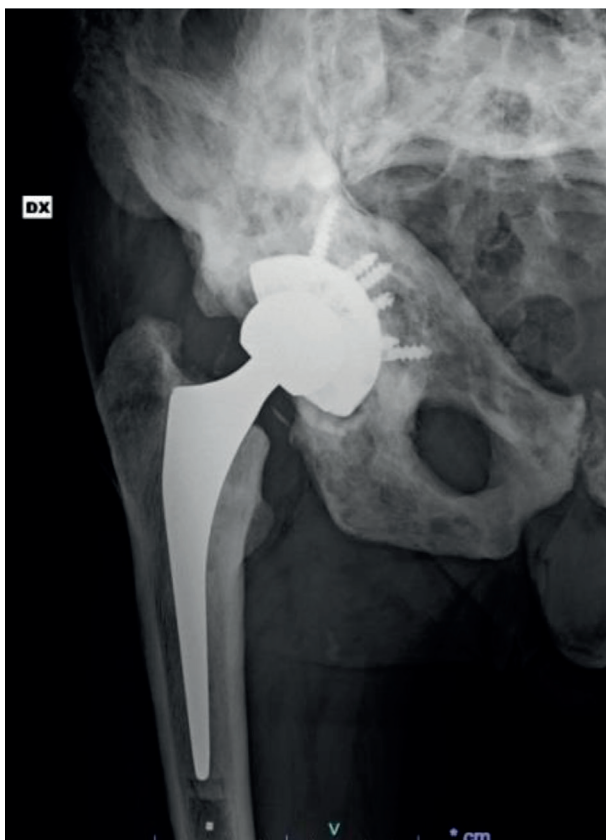


Figure 8. 12 month follow up x ray: antero-posterior view.

hearing loss, symptoms linked to hypercalcemia and hypercalciuria. The patient can suffer also from neurological symptoms due to radicular or medullar compression, caused by spine deformities. Skull asymmetric

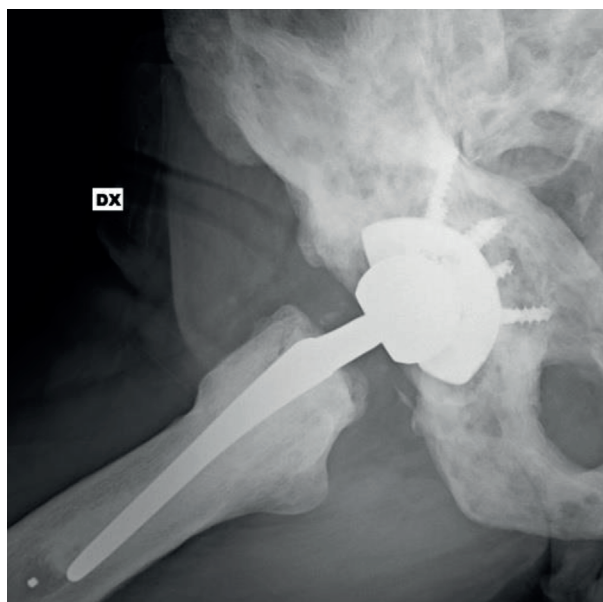


Figure 9. 12 month follow up: x ray an lateral view.

abnormalities need years to be appreciated, so rarely consists of the first sign of the disease. Finally, osteosarcoma and giant cell tumor as PDB's complications are rare.

The diagnosis is guided by X-rays, which show skull deformities, osteolytic or sclerotic processes, cortical thickening, osteoarthritis; CT-scan and MRI are second level exams, used to detect complications.

In combination with normal liver tests, the diagnosis can also be supported by increased levels of serum total alkaline phosphatase (ALP), which reflects the grade of activity of PDB, because the increasing of bone turnover determines an increasing of production of that enzyme. If ALP is normal, but a strong clinical suspicion exists, it could be useful to measure serum levels of bone alkaline phosphatase, procollagen type I N-terminal propeptide and urinary cross-linked procollagen type I N-terminal telopeptide, which are all markers of bone turnover.

Bisphosphonates are the gold standard for PDB's symptomatic patients treatment and zoledronic acid is the most effective one. Concerning asymptomatic patients, some authors suggest to treat those who have cranial and spine deformities, in order to prevent neurological deficit. Complications may require surgery and bisphosphonates are indicated also in the

Table 1. Literature review.

Author (year) reference in the text	Patients (n)	Hips (n)	Mean Age (years (range))	Gender (% male)	Mean Follow-up (years (range))	Revision Rate (%)	Medical complications up to 1 year postoperatively (n)	Heterotopic Ossification (%)	Surgical complications (n)
Sochart (2000)8	76	98	67.4 (51e79)	37.8%	10.4 (5.3e20)	8.2	DVT(3), PE(2), DVT & PE (1), peroneal nerve palsy (1)	29	Dislocation (1), aseptic loosening (2), deep sepsis (1), periprosthetic fracture (1), trochanteric non-union (1)
Kirsh (2001)9	20	20	72 (62-82)	60%	5.7 (4-8)	0	0 (0)	50	Recurrent dislocation (1)
Parvizi (2002)10	18	19	71.3 (55-89)	77.8%	7 (2-15)	0	0 (0)	32	Instability (1)
Wegrzyn (2010)11	32	39	74.2 (55-89)	50%	6.6 (2-12)	5.1	DVT (2) Total 6.25%	56	Periprosthetic fracture (2), intraoperative fracture (1), haematoma (1)
Imbuldeniya (2014)12	27	33	75 (63-85)	56%	12.3 (10.3-17)	18.2	0 (0)	45%	Aseptic loosening (4), periprosthetic fracture (2)
Makaram (2019)5	144	144	76.6 (52-97)	52.8%	10.1	2.8	Anaemia/transfusion (3), cardiovascular event (8), heart failure (1), CVA (1), non-surgical site infections (7), pleural effusion (1)	No data	Haematoma (2), dislocation (4), revision (4)
Tibbo (2020)13	25	25	78 (64-92)	52%	7 (2.4-14.7)	0	Anaemia/transfusion (7)	56%	None
Di Martino (2021)7	66	66		41%	8.4 (0-16.6)	12%	Anaemia/transfusion (22)	52%	Haematoma (2) Aseptic loosening of the stem (4) Aseptic loosening of the cup (1) Recurrent prosthesis dislocation (1) Breakage of head (1) Unknown (1)

preparation of orthopedic operation, to decrease the amount of blood loss, which is higher in these patients, since the bone is more vascularized as a result of elevated bone turnover (1,2).

Paget's disease of bone is a chronic pathology, which can cause as complication hip osteoarthritis, requiring total hip arthroplasty. In our case, we have planned to put a cementless THA, because bone sclerosis makes cement interdigitation very difficult, with higher risk of aseptic loosening; nevertheless, while acetabular component has been fixed without cement and using multiple acetabular bone screws, we have had to change our preoperative plan for femoral component, since, during intraoperative evaluation, we have estimated femoral bone too fragile, so that we have used a cemented stem.

THA is more challenging in this kind of patients compared to general population, because they have higher risk of complication, such as acute renal failure, pneumonia, pulmonary emboli, UTI, respiratory failure, ileus. Moreover, they have more blood loss during intervention so blood transfusions are more frequent. Furthermore, the incidence of aseptic loosening of the implant and the rate of revision are higher in these patients. Thereby, the length of hospitalization is longer and costs higher (4).

Makaram et al. (5) too found a higher risk of medical complication after THA in patients with PDB compared to general population, whereas there is no difference concerning surgical complications. According to these authors, implants survivorship after 10 years follow up is over 95 %, which results comparable with that of THA in general population.

In their studies, Hanna et al.(6) and Di Martino et al. (7) find excellent functional outcomes after THA in PDB patients, with values similar to those of general population; however, they find an increasing rate of aseptic loosening and revision's rate in PDB group, with no statistically significant differences, between cement THA compared to cementless implants.

Sochart et al.(8) analyze a group of 98 cemented THA and they find at 10 years follow up excellent improvement in clinical and functional outcomes. Moreover, rates of survivorship, heterotopic ossification and aseptic loosening are similar to those of general population undergone THA. They find, instead,

an increased risk of trochanteric osteotomy nonunion or delayed union.

Kirsh et al. (9) describe a cohort of 20 THA with hydroxyapatite-coated stems, combined with cementless or cemented or hydroxyapatite-coated cups. They find at a medium term follow up improvement of clinical and functional outcomes.

Parvizi et al. (10) presents a cohort of 19 cementless THA in 18 patients with PDB. After an average follow up of 7 years, clinical and functional outcomes improve in all patients; moreover, radiographies show optimal implants ingrown.

Wegrzyn et al. (11) analyze a cohort of 39 uncemented hydroxyapatite fully-coated THA in PDB patients, finding excellent improvement of clinical and functional outcomes after surgery. They also find no correlation between heterotopic ossification and Harris Hip Score or serum levels of alkaline phosphatase. In patients treated by bisphosphonates before surgery, they observe less intraoperative blood loss and lower levels of serum alkaline phosphatase.

Imbuldeniya et al. (12) describe a cohort of 14 uncemented THA in PDB, finding good implants survivorship and excellent improvement of functional and clinical outcomes.

Tibbo et al. (13) present a cohort of 25 THA in PDB patients, with uncemented acetabular component associated to uncemented or cemented femoral stem. They find an improvement of clinical and functional outcomes, with a survivorship of implants over 90%. The main complications were excess blood loss and heterotopic ossification, that result much higher than in general population.

Conclusion

THA is an optimal choice of treatment in PDB patients with severe hip osteoarthritis; clinical and functional outcomes improve after surgery and implants survivorship is excellent. However, it is more challenging because of bone alteration induced by the pathology. More studies with long-term follow up are needed to detect rates of survivorship and revision, comparing them to those found in general population; moreover, more studies comparing cementless THA

with cemented one are necessary, in order to understand if one implant is better than the other.

The patient has given his informed consent for this case report.

Conflict of Interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article.

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