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Case report of early aseptic loosening of total hip arthroplasty in monostotic paget disease, a diagnostic challenge



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ABSTRACT

Paget's disease of bone is a localised chronic osteopathy which produces bone deformities, bone hypervascularity, structural weakness and altered joint biomechanics. Although radiological diagnosis of Paget's disease of bone is usually straightforward, monostotic cases may potentially raise specific problems which require invasive and expensive procedures such as bone biopsies. The pelvis and upper femur are frequently affected, resulting in disabling hip disease that may require total hip arthroplasty. We report a case of Paget disease of bone in an 84-year-old woman, which was initially identified as avascular necrosis of the hip, reason for which she underwent total hip arthroplasty. During follow up, the patient complained about hip pain and in a few months she was not able to walk because of an early loosening with bone destruction. Radiological and laboratory exams were carried out with normal results except for alkaline phosphatase (AP). After treatment with biphosphonates hip pain relieved but hip reconstruction was not possible. In this paper we present an early aseptic loosening of hip arthroplasty due to monostotic Paget's disease of bone, a rare etiology of loosening which poses particular diagnostic difficulties prompting an excessive use of excisional biopsies.

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1. Introduction

Paget's disease of bone (PD) was first described in 1877 by Sir James Paget at St Bartholomew's Hospital in London. Sir Paget described patients with multiple deformities of the bone and noticed that some patients developed bone sarcomas.

Nowadays PD is a localised chronic osteopathy characterised by an osteoclastic dysfunction with increased bone resorption and a subsequent compensatory formation of new bone with a defective microstructure. The disease may appear in its monostotic (only one third of patients) or polyostotic variants and the most common symptom is bone pain, which is typically constant, present at rest, and worse at night [1]. Patients may also present pathologic stress fractures, bony enlargement, gait disturbances or secondary osteoarthritis. Physiologic markers of bone turn over may be elevated. Suspicion should arise in case of bone pain with characteristic radiological findings.

We present a case of early failure of total hip arthroplasty, following hip osteonecrosis diagnosis and constant hip pain, more severe at rest despite the arthroplasty; subsequently, alkaline phosphatase was measured and elevated serum levels were found which

was reflective of a rapid new bone turn over. Therefore, a new diagnosis was reached: early implant loosening because of PD.

2. Case report

An 84-year-old woman was referred to our department because of pain in her left hip. The pain was of a constant nature and, although she was able to walk, it had risen gradually causing an antalgic gait. The pain got worse at rest and there was no history of trauma reported. Clinical examination showed a decreased active and passive range of motion, associated with relative left thigh atrophy, most likely because of a lack of use of the quadriceps due to the pain.

Anteroposterior and axial radiographs (Fig. 1) showed incipient hip osteoarthritis. Consequently, medical treatment was started with non-steroidal anti-inflammatory drugs (NSAID). The X-ray showed mixed radiolucent and radiopaque areas. Three months later the patient complained about hip stiffness; a new radiological exam and magnetic resonance imaging were carried out revealing cephalic bone necrosis. This X-ray shows lytic areas (Fig. 2). Before surgical treatment a bone biopsy was practiced to confirm the initial diagnosis: avascular necrosis of the hip. During surgery, a biopsy specimen was taken from the femoral head of the right hip due to excessive bleeding of the acetabular bone. The histological findings of the specimens and bones showed no evidence of malignant cells.

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Fig. 1. Radiographs of the left hip in anteroposterior view showing hip osteoarthritis and mixed radiolucent and radiopaque areas (white arrow).

Following the total hip arthroplasty (THA), she had pain-free and improved clinical functions. As demonstrated in Fig. 3, the immediate postoperative radiograph shows a correct implant positioning. Two months later, she developed a creeping right groin pain and a limp in spite of NSAID. The radiograph demonstrated an acetabular loosening with migration of acetabular component. She was advised non-weight bearing and crutches or wheel chair use. Regardless, three months later hip pain increased. Since we suspected another etiology other than hip osteoarthritis, we decided to broaden the imaging studies to discard any lesions in other bones. The radiograph and the computerized tomography (CT) showed acetabular and iliac bone destruction. The CT scan shows esclerotic and lytic areas on the right iliac bone (Fig. 4). In order to make certain it was not a malignant tumor or a bone infection a new biopsy was carried out and we obtained a large blood volume showing a profuse local vascularity. The histological section can be seen in Fig. 5.

The patient underwent laboratory investigations with blood and urinary tests looking for myeloma or lymphoma. Serum bone AP was high (three times greater than the normal value), which is reflective of the rapid new bone turnover. Other laboratory studies showed normal levels of calcium, phosphate, parathormone and vitamin D levels. A complete succession of bone radiographs (skull, humerus, spine, femur, pelvis, tibia) was performed, showing



Fig. 3. Immediate postoperative radiograph, cementless implants well positioned.

pathological findings exclusively in the pelvic bone: generalized mixed radiolucent and radiopaque areas with characteristic cotton-wool appearance (Fig. 4). No bone scan was done.

All these findings support monostotic PD as the diagnosis that caused acetabular loosening with bone destruction. After treatment with biphosphonates hip pain decreased but because of the severe bone destruction the patient could not walk. We offered the patient a replacement of her hip arthroplasty after the treatment with bisphosphonates due to the obvious loosening of the implant. She did not accept surgical treatment to achieve hip reconstruction. At present she needs two crutches to stand up.

3. Discussion

Paget’s disease of bone is a rare condition with an approximate incidence of 2–3% in patients older than 40 years [2–4]. The etiology is unknown, several observations suggest a viral cause (measles, respiratory syncytial virus and canine distemper virus) [5] but genetic and environmental factors have been implicated in the development of PD (mutations in the sequestosome 1 or p62 gene, SQSTM1) [6–9]. There is strong evidence that susceptibility is determined by variants within or close to genes that regulate osteoclast function, moreover the frequency of familial aggregation is high (in some studies 40% of affected patients have a family history of PD) [10–12].

It is a focal disorder of bone metabolism, characterized by an initial phase of bone resorption that begins in subchondral bone

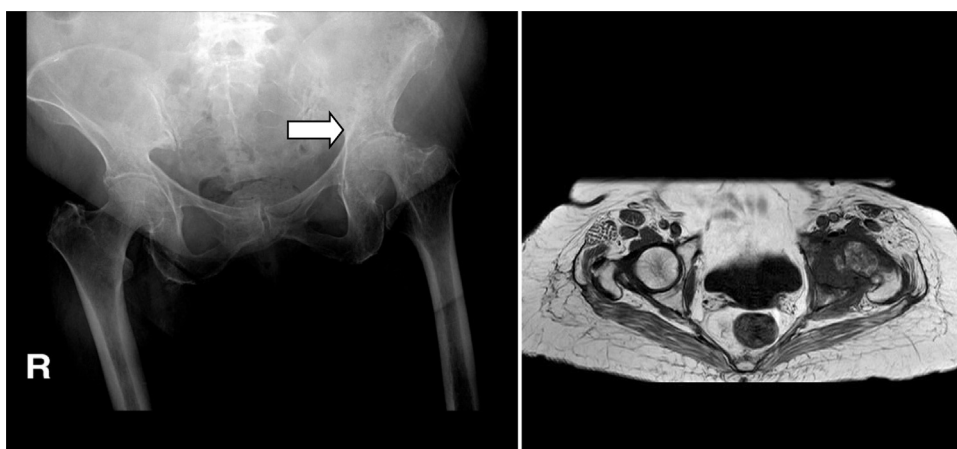


Fig. 2. Preoperative radiograph showing cephalic bone necrosis and magnetic resonance image demonstrating avascular necrosis and acetabular bone changes.

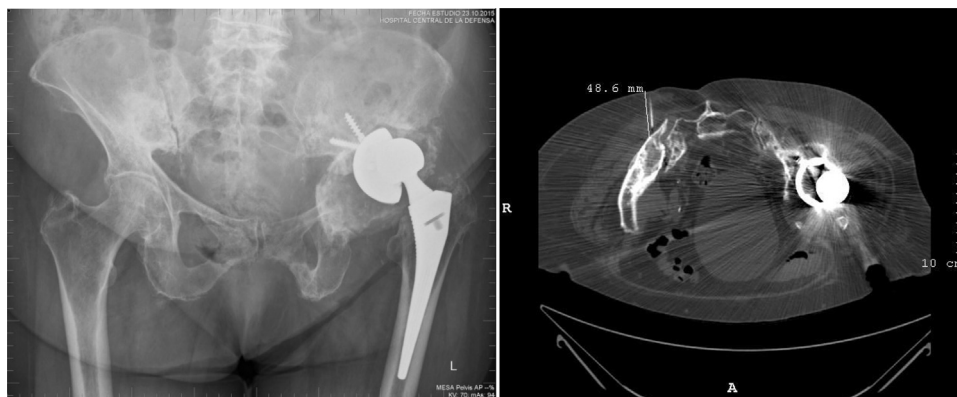


Fig. 4. Radiograph and computerized tomography (CT) demonstrating left acetabular and iliac bone destruction. The CT also shows esclerotic and lytic lesions on right iliac bone.

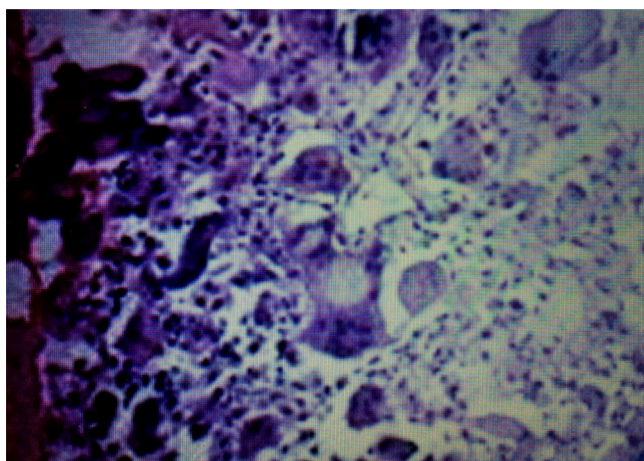


Fig. 5. Histological section: no malignant cells can be observed, but there is an increase in size and number of osteoclasts, with multiple nuclei per cell.

and moves through the affected bone. Bone growth occurs thanks to lamellar and woven bone which settle at the same time that capillary are creeping through the bone marrow with a peritrabecular fibrosis. The presence of woven bone and marrow fibrosis is also seen in other bone turnover conditions; therefore the histologic changes are not pathognomonic for PD [13].

Three tests (an X-ray, a blood test, and a bone scan) are commonly used to discover Paget's disease. X-rays are used to make the final diagnosis. In X-ray images, bones affected by Paget's disease have a particular appearance. Radiographs include both lytic (early) and sclerotic findings, osteoporosis circumscripta in skull, bowed limbs, and flame-shaped lesions in long bones. Many patients are diagnosed incidentally in the asymptomatic phase through plain radiographs that show localized enlargement of bone. These radiographs often have a high specificity because of their classic nature, but a low sensitivity. In a blood test, a higher level of serum AP is a sign that the disease may be present. An AP level greater than twice the usual level strongly suggests Paget's disease, especially if the patient's serum calcium level, phosphorus level, and kidney function are normal. A bone scan helps identify which bones have been affected by Paget's disease. Furthermore, bone scans can be used to increase diagnostic sensitivity in patients suspected of having Paget's disease, although this method is less specific and should be interpreted cautiously.

This structural change produces bone that is expanded in size, is more vascular, and may lead to osseous deformities, skeletal fragility and altered joint biomechanics. It is characterized by rapid bone resorption and reposition, resulting in formation of numerous reversal lines, which give rise to a mosaic pattern in the lamellar

bone with profuse local vascularity and fibrous tissue in the marrow. In the initial phase of PD, there is excessive bone resorption followed by increased deposition. However, both may occur simultaneously, resulting in osteoporosis and sclerotic bone. Within the sclerotic bone there is reduced vascularity (localized vascularity) and the ability for normal healing is compromised, hence it can lead to osteonecrosis [14]. This excessive growing explains some of Paget's disease complications such as deformity, bone pain, fracture, nerve compression syndromes, and orthopedic complications like implant loosening [15].

Prevalence of PD has been shown to increase with age [16] and the most commonly involved regions include the pelvis, femur, spine, skull and tibia. The pelvis and upper femur are involved in 20–80% of patients with PD resulting in frequent disabling hip disease [17].

When PD is symptomatic, the most common presentation is poorly localised bone pain. Studies have demonstrated that the level of bone pain is proportional to the level of disease activity [18]. Deformities such as coxa vara, femoral bowing, acetabular protrusion and bony enlargement can be observed in advanced stage and may lead to Paget's arthropathy. However, in initial stage this disease can be silent or asymptomatic so an early diagnosis is impossible sometimes. Our patient complained about hip pain but no deformity was present, and the X-ray exam did not show any pathological findings in other bones other than the left hemipelvis.

Increased bone turnover and remodeling is associated with elevated levels of serum AP. Such a marker allows an earlier diagnostic and assessment of the activity of Paget's disease and monitoring of the effectiveness of the medical treatment by bisphosphonates [19,20].

Antiresorptive treatment suppresses bone turnover in Paget's disease [13] and improves the appearance of pagetic lesions on the isotope bone scan, but evidence-based medicine only supports the use of antiresorptive drugs for the treatment of bone pain. Bone pain in Paget's disease can be managed either with simple analgesics and NSAID or with antiresorptive drugs such as calcitonin and bisphosphonates. In our case we began treatment with NSAID but antiresorptive drugs would have prevented bone destruction and implant loosening and would have been more effective for pain. Use of bisphosphonates has been shown to stop the cycle of macrophages that absorb small particles of wear debris activating osteoclasts to start bone resorption, although this clinical outcome has not been clearly defined [14].

It is well known that in such cases total hip arthroplasty relieves pain, but long-term outcome in these patients has raised concerns about the potential for implant loosening. Cemented implants are preferred but recently, the use of cementless implants in patients with PD and preoperative antipagetic medication has been reported

with encouraging results [15,16]. We decided to implant a cementless hip arthroplasty but the patient suffered an early loosening. To our knowledge, total hip arthroplasty early loosening with bone destruction because of monostotic PD has never been reported.

In this case we should have thought about PD during the first assessment of hip pain because of the patient's age and suspicious bone lesions in radiographs. Avascular necrosis development was so rapid that we decided to take a bone biopsy, from which we didn't obtain any malignant findings, but it did not occur to us to look for alkaline phosphate serum levels, which is why we erred in the diagnosis.

When patients with this condition require surgical treatment, the orthopedic surgeon should be aware that this is a hypervascular disorder with increased perioperative blood loss. Elective surgical prophylaxis such as biphosphonates is required to decrease perioperative bleeding. Biphosphonates include oral etidronate, alendronate, risedronate and intravenous pamidronate. Optimal dosage remains controversial [17]. When implanting hip or knee arthroplasty in patients suffering PD it can be useful to treat pain with biphosphonates, on the one hand to control pain, and on the other to prevent early loosening [18] and avoid catastrophic cases as the one we present here.

The correct diagnosis was belated and it was astonishing how quickly the pelvic bone was destroyed. For future cases it would be advisable in older patients with suspicious bone lesions to determine serum AP levels before performing invasive procedures. In order to avoid bone destruction it is useful to use oral biphosphonates before orthopedic surgery. Medical therapy achieves osteoclast inhibition and reduces excessive bleeding during arthroplasty.

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Consent

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Author contribution

Diana Crego-Vita wrote the paper.

Daniel Aedo-Martín did the bibliography search.

Coral Sánchez-Pérez described the radiological findings.

Guarantor

DR. DIANA CREGO-VITA.

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