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CASE REPORT

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Rapidly destructive bilateral hip disease: a case report and review of the literature

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Abstract We describe the case of an elderly woman with rapid destruction of the right hip followed by the same involvement of her left hip 10 months later. The clinical history, the physical examination and radiographic images suggested the diagnosis of rapidly destructive hip disease. This disease is a distinct entity, unilateral in 80%-90% of cases, which requires extensive investigation and special efforts for its identification. Essential elements for the differential diagnosis are discussed.

Key words Hip • Total hip arthroplasty • Joint destruction

Introduction

Rapidly destructive hip disease is an unusual hip arthropathy which leads to partial or complete destruction of the involved joint, until it stops spontaneously. The entity is typically encountered in elderly women and it is mainly unilateral. It was first described by Forestier in 1957 [1] and its etiology and pathogenesis are still debated.

Subsequent studies gave it a large variety of titles, such as rapid destructive osteoarthritis, atrophic osteoarthritis, Postel's osteoarthritis and rapidly progressive osteoarthritis [2, 3]. This entity is rarely encountered in the classic orthopedic texts and seldom reported in the literature, so that it is difficult to suspect and the diagnosis can be particularly troublesome and time consuming.

The result of this process is the rapid destruction of the hip joint that often occurs within months of the onset of symptoms, associated with joint instability and impaired function. Bilateral involvement is uncommon. We describe a case with rapid onset and bilateral involvement of both hips.

Case report

A 69-year-old woman was admitted to her local hospital with an eight-month history of right hip pain, without history of preceding trauma or pelvic surgery. A radiograph of the right hip taken three months before showed osteolytic cystic-like lesions of the upper part of the femoral head and of the outer third of the acetabulum (Fig. 1a, b).

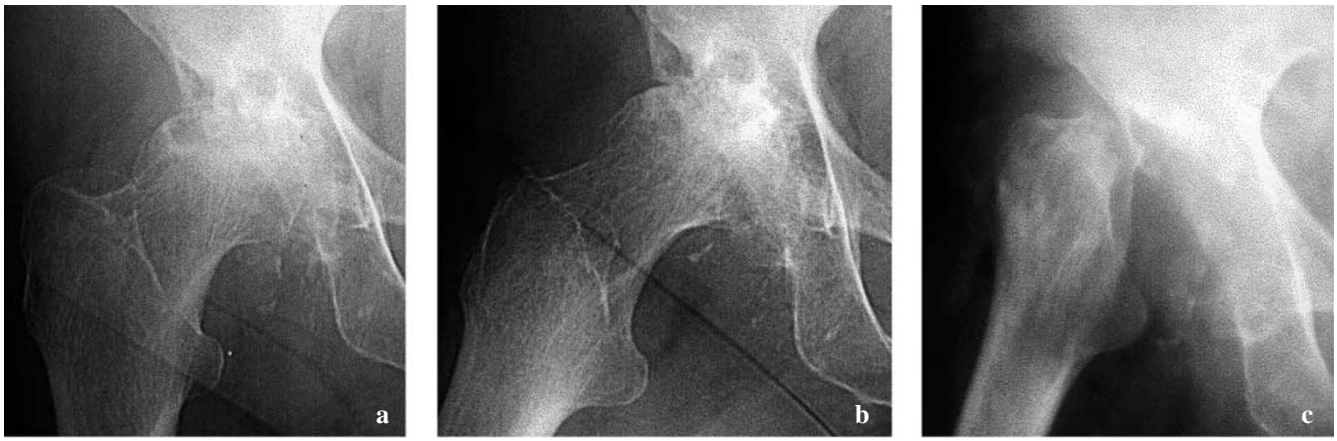

Fig. 1 a, b Anteroposterior radiographs of the right hip in neutral position (**a**) and in abduction and external rotation (**b**) showed the complete disappearance of the joint space and osteolytic cystic-like lesions of the upper part of the femoral head and the outer third of the acetabulum. **c** Three months later, a complete destruction of the hip joint with proximal migration of the femoral diaphysis was observed

At the time of admission, no risk factors for avascular necrosis and no steroid intake were recorded. No evidence of other concurrent disease was present. The pain was worsened by walking and by digital pressure on the greater trochanter. Physical examination showed a reduction of the passive and active ranges of motion of the right hip. Pelvic radiography (Fig. 1c) and computed tomography (CT) (Fig. 2a) of the right hip showed an almost complete resorption of the femoral head. Tc-MDP scintigraphy showed a wide increase in uptake in the region of the right hip corresponding to the radiographic abnormalities (not shown).

At that time, an open biopsy of the right hip was performed, revealing non-specific signs of aseptic necrosis of the articulation. Biochemical investigations showed a normal white blood cell count; liver and kidney functions were normal, as were serum calcium and phosphate levels. The sedimentation rate and reactive C-protein and acid alpha-1 glycoprotein levels were over the reference values; RA-test and ASO were normal. Blood pressure was 140/80 mmHg under pharmacological control and body temperature was 37° C.

In order to complete the diagnosis, the patient was transferred to our department where radiographs confirmed a progressive massive resorption of the femoral head and acetabulum. Magnetic resonance imaging (MRI) performed one week later showed the complete resorption of the right femoral head, the cranial migration of the femoral diaphysis with an associated joint effusion and no other pathological features (Fig. 2b). All laboratory tests, including those for tuberculosis, Widal-Wright reaction and tumor markers (GICA, AFP, CEA test) were negative and the neurologic examination did not show any abnormal findings. A second open biopsy led to the diagnosis of non-specific chronic synovitis, intense osteoporosis and marrow substitution with adipose tissue. Routine bacteriologic culture of specimens

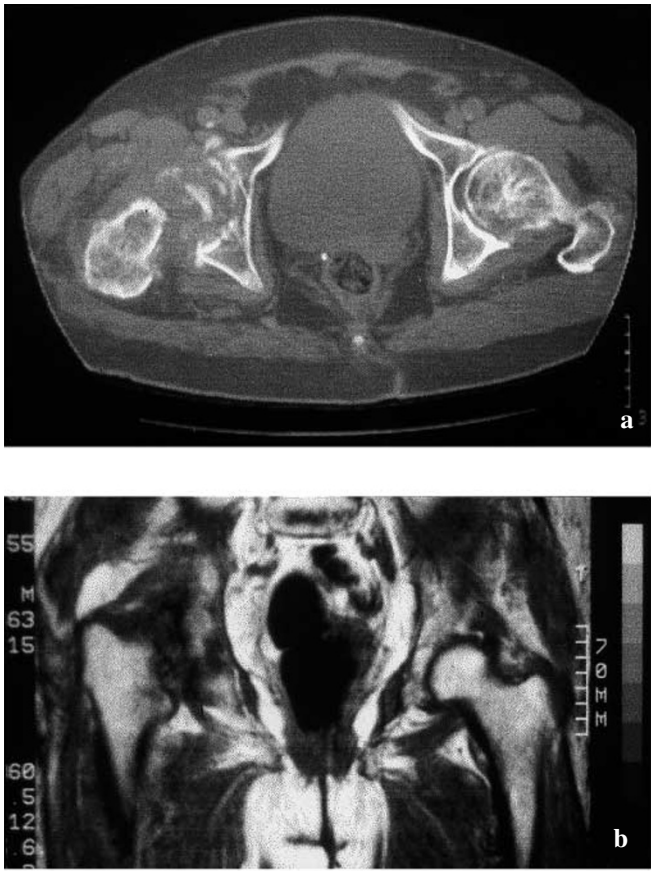

Fig. 2 a CT scan of the pelvis four months after the first radiograph showed the disappearance of the right femoral head and the partial disruption of the acetabular structure. **b** Coronal T1-weighted MR image of the pelvis four months after the first radiograph showed the resorption of the right femoral head and neck and disruption of the acetabular cavity with concurrent joint effusion

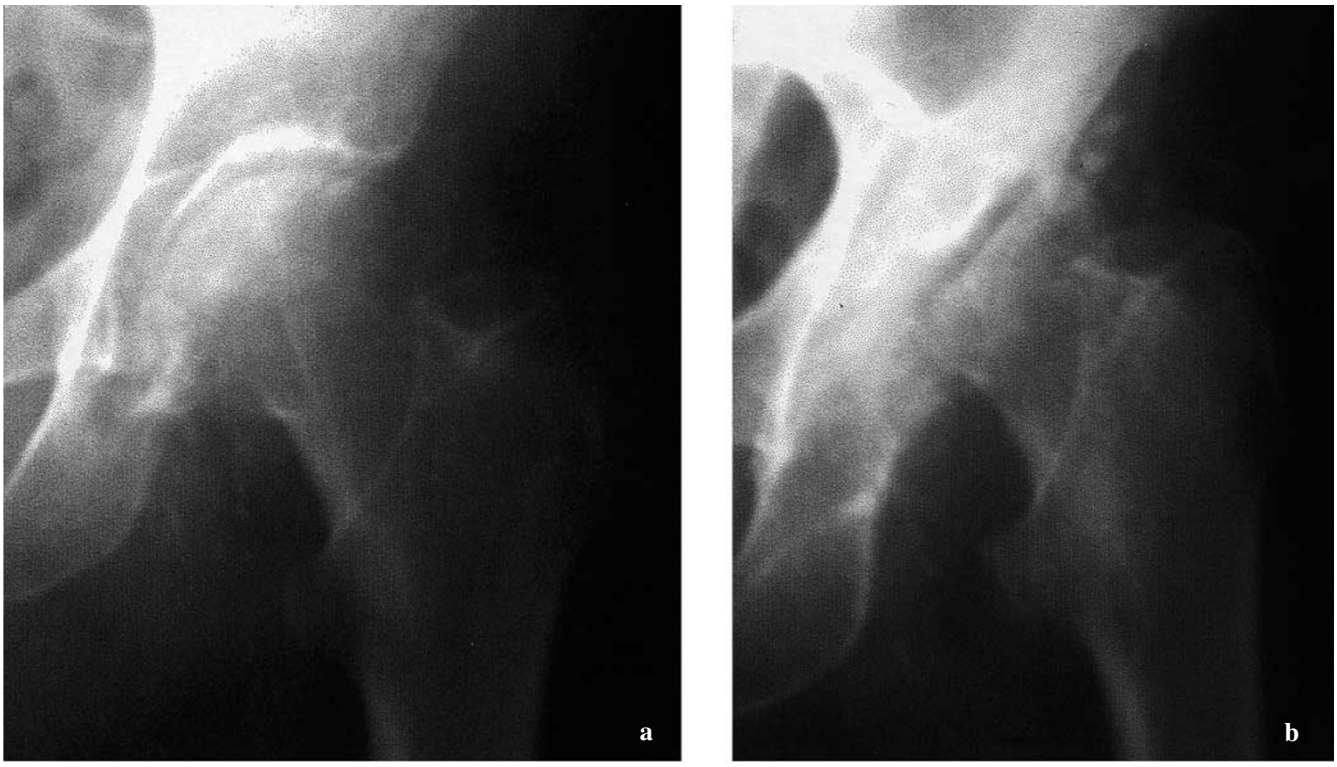

Fig. 3 a Anteroposterior radiograph of the left hip in neutral position nine months after the surgical treatment of the right hip showed no abnormalities. **b** One month later, partial destruction of the left hip joint

was negative. Two weeks later a total hip arthroplasty was performed, and the patient was discharged from the hospital.

At a scheduled follow-up after nine months, the prosthesis did not show any radiographic or clinical sign of loosening and the contralateral hip was normal (Fig. 3a). One month later, the patient went back to the hospital because of the sudden onset of pain and impaired function at the left hip. A plain radiograph of the pelvis showed the almost complete destruction of the left hip (Fig. 3b). The woman was soon hospitalized. Clinical, radiological and laboratory investigations were undertaken with the same results of the first experience and a total hip arthroplasty was performed three weeks later.

Discussion

Rapidly destructive hip disease is an uncommon arthropathy which is still variously classified as a distinctive entity, a subset of osteoarthrosis or a particular form of osteonecrosis of the femoral head. The interaction of mechanical stress, cartilage degeneration and bone response in the genesis of the disease has been suggested by Salomon et al. [4]. In their opinion, rapidly destructive hip disease is a subset of osteoarthrosis where destructive phenomena prevail due to the rapid degeneration of cartilage and the poor bone response. More recently, Inoue et al. [5] reported that the immunoresponsiveness to the articular cartilage might mark the initiation of joint destruction, and Komiya et al. [6] found elevated levels of prostaglandins, interleukin-1β and proteolytic enzymes in the synovial fluid of patients with rapidly destructive hip disease but not in patients with coxarthrosis. These observations suggest that rapidly destructive hip disease is a separate entity but its cause is still unknown.

The disease is typically encountered in elderly women in the seventh decade of life and it is unilateral in 80%–90% of cases [7, 8]. The average time from clinical presentation to the appearance of severe hip destruction ranges from 2 to 16 months.

Radiographic findings of the involved hip show an asymmetric rapid narrowing of the joint space in its outer bearing portion with bone sclerosis, subchondral cysts and minimal or no osteophytes at the upper bearing portion of the femoral head and in the corresponding outer third of the acetabulum. In a few months, an extensive subchondral bone loss takes place in the acetabulum and femoral head with final flattening of the cotyloid cavity and complete disappearance of the entire proximal epiphysis of the femur. A

similar involvement of other joints, particularly the shoulder, has been reported [3, 7, 8]. In most cases, histologic examination demonstrates extensive fibrosis in the joint capsule, disappearance of the cartilage, mild signs of acute or chronic inflammation of the synovium and bone with non-specific inflammatory cells, bone resorption by osteoclasts and focal loci of osteonecrosis [7, 8].

When the joint destruction progresses so rapidly as in the case we reported, other possible causes must be promptly ruled out. Radiographic findings may simulate primary osteonecrosis, rheumatologic disorders, septic arthritis or neuropathic arthropathies. Primary osteonecrosis can be excluded by the rapidity of bone destruction, the acetabular involvement and the early joint space disappearance. Rheumatologic arthritis may be excluded by the history and the clinical presentation, the lack of serologic abnormalities, the monoarticular involvement and the absence of marginal erosions on radiographs. Septic arthritis usually is associated with leukocytosis and local and systemic signs of infections. In neuropathic arthropathies, abnormalities are present at the neurologic examination and sometimes the patient is unusually pain free.

A rapid and severe destruction of large joints may follow other disorders. Anti-inflammatory drugs, especially indomethacin, may cause a destructive hip arthropathy by a direct drug toxicity or by induction of an analgesic state (iatrogenic neuropathic joint) [9]. Chondrocalcinosis and apatite-associated arthropathy may be implicated as a cause of joint destruction and may be diagnosed by looking for pyrophosphate or hydroxyapatite crystals in the synovial fluid [10, 11].

In conclusion, rapidly destructive hip disease is a distinct entity, sometimes dramatic in its onset and progression, which requires extensive investigation and special efforts for its identification. Most of the patients require a total hip replacement with good final results as reported in the literature [2, 3, 8]. Familiarity with this entity may reduce the need for extensive diagnostic tests.

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