Bilateral vanishing hips, coincidence or systemic disease? A case report and overview of current literature

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ABSTRACT

Rapidly progressive hip disease (RDHD) is a rare condition of the hip joint, causing destruction of the femoral head. The pathogenesis is unknown. The disease is self-limiting, there is no treatment to stop the disease. Hip arthroplasty is a successful way to relieve pain and restore function. We present a case where both hips were involved and analysed. A favourable result was obtained by bilateral total hip arthroplasty.

1. Introduction

Rapidly progressive hip disease (RDHD) is a rare condition of the hip joint, resulting in rapid destruction of the femoral head, with inconsistent involvement of the acetabulum. Over a period of months a femoral head can completely disappear, as first reported by Forestier in 1957.¹ The pathogenesis remains unclear. Possible hypothesis are subchondral bone ischemia and cell necrosis, subchondral insufficiency fracture in patients with or without osteopenia resulting in bone collapse, enzyme disturbance in synovial fluid, and an aberrant bone response to osteoarthritis.² There is no treatment available for this syndrome. In this report we will show a case in which the condition appeared bilateral and in one shoulder, to contribute to the limited available literature.

2. Case report

A 70 year old male patient presented at our institution with pain in the left hip since three months. Patient is mentally challenged and had a history of alcohol abuse, drinking six to twelve alcohol units a day. Otherwise the patient reports that he is in good health. Physical examination showed a lack of full flexion and zero internal rotation of the hip. Radiographs showed a coxarthrosis of the left hip with narrowing of the joint space and subchondral cyst formation (Fig. 1). Patient was placed on the waiting list for a total hip arthroplasty of the left hip. While waiting on the hip replacement, the groin pain increased exponentially in a short period of time. Furthermore, he experienced mild pain in his right shoulder. Therefore additional X-rays were obtained 15

weeks after initial presentation. Complete bilateral destruction of both hip joints was seen (Fig. 2) and a flattening of the humeral head with loss of joint space (Fig. 3). A diagnostic biopsy was performed of both hips. The obtained specimen was submitted to the microbiology laboratory. The culture showed no growth. Pathological assessment excluded a malignant cause of the bilateral vanishing hips. Histological examination of the core biopsies showed necrotic and fragmented bone trabecula partly covered by fracture callus. The marrow consisted mostly of fat and callus. No granulomas or evidence of infections were seen. These findings can be caused by avascular necrosis. Patient was also referred to the outpatient clinic of internal medicine who performed an extensive internal analysis. There were no signs of infection, malignancy or metabolic disease. Patient was re-scheduled for total hip arthroplasty of the left hip.

Peroperatively, the joint capsule was thickened. The femoral head and neck were disappeared and the acetabulum was filled with fibrotic tissue. The acetabulum wall was also affected, however there was no need for bone grafting. Because the patient was mentally challenged we implanted a cemented dual mobility cup. There were no postoperative complications. The patient was seen in the outpatient clinic after six weeks for the routine checkup. He was satisfied with his left hip, clinical examination showed a pain free hip with a good range of motion. During this visit, the patient was scheduled for the total hip replacement on the other side. The second joint replacement surgery had the same good progress. Fig. 4 shows the post-operative performed X-ray. The patient recovered quickly and showed good progression in his physiotherapeutic rehabilitation. Almost a year after both hip arthroplasties, a reversed shoulder arthroplasty was performed because of

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Fig. 1. X-ray showing coxarthrosis of the left hip with narrowing of the joint space and subchondral cyst formation.

Fig. 2. X-ray 15 weeks later, complete destruction of both femoral heads.

pain and rotator cuff deficiency (Fig. 5). Histological examination of the humeral head showed architectural and histological normal bone trabecula. No necrosis or callus was present.

3. Discussion

Rapidly destructive arthroses is an uncommon subset of osteoarthritis, also known as vanishing hip and rapidly progressive osteoarthritis, and is a diagnosis per exclusionem. It is a self-limiting disease.

Patients present with rapidly onset of severe hip pain. Physical exam can show an antalgic gait and limb shortening. Radiological features are a rapidly progressive chondrolysis and subchondral bone loss.³ Initial radiographic findings may have a normal appearance, but within a few months destruction of the femoral head and acetabulum arises, to complete vanishing of the femoral head. No osteophytes have been observed. Histopathological findings of RDHD are bone resorption, synovitis and osteonecrosis. An extensive fibrosis of the joint capsule can be seen. It is reported that in the histological sections of patients

Fig. 3. X-ray shows flattening of the humeral head and narrowing of the glenohumeral joint space.

Fig. 4. Postoperative radiograph showing bilateral cemented total hip procedures.

with vanishing of the femoral head an increased amount of osteoclasts were found which were morphologically altered and there was formation of resorptive giant cell granulomas.⁴ A case series study did not found histopathological traceable reason for infection or malignant disease.⁵

The pathophysiology of rapid destructive arthritis is not fully understood. Different causes are mentioned, which induce osteonecrosis or osteoporosis and subsequently collapsing of the hip. Firstly the presence of a greater amount of osteoclasts as compared to primary hip arthritis can result in bone resorption.⁴ Further, RDHD patients could

Fig. 5. Radiograph showing the cemented reversed shoulder prosthesis.

have subchondral fractures which precede the onset of the disease with secondary superimposed osteonecrosis.⁶ Another possible cause is osteoporosis followed by insufficiency fracture and collapse of the femoral head or neck.⁷ It was proposed that a risk factor for the development of osteonecrosis was drug toxicity, primarily nonsteroidal anti-inflammatory drugs could contribute, however this was later on challenged.⁵ An important risk factor is the use of alcohol, there is a higher risk of osteonecrosis of the femoral head in occasional drinkers and even more in regular drinkers (relative odds = 13.1) than in controls.¹⁰

Most important is to do a complete work-up, no underlying pathology should be missed. This applies in particular to a potential systemic (malignant) disease or the potential toxicity of medication on vital organs. The differential diagnosis of vanishing hip should include a malignant cause of bone destruction, septic arthritis, avascular necrosis and neuropathic arthropathy.² RDHD differentiate because of the absence of bone formation and osteophytes, and the rapid onset (typically 1–6 months). The differential diagnosis should also include a disease called the Gorham-Stout disease, known as a cause of mysterious bone disappearing. It has been described to affect primarily young patients and its etiology and pathogenesis is not completely understood. A minor trauma may be present before onset of the disease and in contradiction to vanishing hip syndrome it is relatively painless until pathological fractures occur.^{8,9}

There is no cure for this condition. However, symptoms can be effectively reduced with total hip arthroplasty.

Interesting about our case is that both hips were involved. First we thought that his shoulder showed a similar disease, however histological examination excluded this. Microscopic examination of the hips of the presented case revealed necrosis and fragmented bone trabecula, there were no granulomas as seen in earlier presented studies. Similar involvement of multiple articulations suggest a more systemic process, however this is not found. Possible cause of the presented case is that his alcohol consumption led to osteonecrosis of both femoral heads, with subsequently collapsing of the femoral heads. That both femoral

heads collapsed at the same time can be coincidence, or a more systemic process remains unclear.

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Conflict of interest

None.

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