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FUNDING: Nil.

ETHICAL APPROVAL: Informed consent was obtained from the patient included in this case report.

CONFLICT OF INTEREST: The authors declare that they have no conflict of interest.

BLINDED MANUSCRIPT

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KEYWORDS: Chondrolysis; Hip Pain

ABSTRACT (Word count – 82 (MAX 250)):

We report the case of a 23 year old male diagnosed with adult-onset idiopathic chondrolysis of the hip. Chondrolysis of the hip is a disorder most frequently seen in children who have suffered with slipped capital femoral epiphyses. Idiopathic chondrolysis of the hip is extremely rare and its onset has never been documented in adults aged over 20. With reference to the available medical literature, we summarise the current clinical management of this unusual but important cause of young adult hip pain.

INTRODUCTION

We report the case of a 23 year old gentleman with adult-onset idiopathic chondrolysis of the hip. This extremely rare condition largely affects children and the onset has not previously been reported in anyone aged over twenty years. We discuss the clinical course and subsequent management of this disease with reference to the current medical literature.

CASE REPORT

A normally fit and well 23 year old male was referred to the orthopaedic clinic with suspected primary osteoarthritis of the hip. He described a one-year history of progressively worsening severe right sided hip pain, which had become increasingly debilitating to the point of the patient relying on crutches to mobilise. He had no features of underlying systemic illness such as fevers, rigors, weight loss or nausea. He denied any preceding trauma, had no relevant past medical issues and no conditions were known to run in his family. At the time of orthopaedic review he was taking several regular analgesics which were not providing adequate pain relief. He received no courses of antibiotics during this period.

On examination, the patient was mildly uncomfortable at rest and walked with an antalgic gait. His right hip was held in fixed flexion and appeared to be extremely painful on all attempted active and passive movements. The contralateral hip was clinically normal and there was no objective evidence of restriction in any of his other joints.

Initial investigations focussed on exclusion of an underlying inflammatory process. Blood tests including white cell count, erythrocyte sedimentation rate and C-reactive protein were consistently within normal limits. Markers of auto-immune disease, including Anti Nuclear Antibody, Anti-cyclic citrullinated peptide (Anti-CCP) antibody and rheumatoid factor, were also negative on separate occasions.

Plain radiographs showed uniform loss of joint space and juxta-articular osteopenia at the right hip. There was no evidence of femoro-acetabular impingement, with an alpha angle calculated as 48° (Figure 1A & 1B). Magnetic Resonance Imaging of the hip showed synovial thickening and marrow oedema in the acetabulum and femoral head (Figure 1C & 1D). These findings were more consistent with an inflammatory process rather than progressive osteoarthritis. However, an ultrasound-guided aspirate of the hip was sterile on culture.

In the absence of a confirmed underlying inflammatory or infective process, the decision was taken to proceed to a right total hip replacement for symptomatic relief of this rapidly progressive condition.

An elective cemented total hip replacement was performed using a standard posterior approach. There was no significant joint effusion, however a marked inflammatory synovitis was clearly identified throughout the hip. Following dislocation, a large ring of synovitis around the inferior aspect of the neck was observed; given these findings the femoral head and multiple tissue samples were sent for pathological assessment. The operation was completed without undue complication. On the table, this resulted

in a good range of movement and satisfactory limb length. Aside from the usual peri-operative prophylaxis, no regular antibiotic therapy was prescribed for this patient.

Gram stains and prolonged cultures (standard 21 days incubation in our institution's Microbiology department) of intra-operative samples showed no evidence of underlying bacterial infection. Microscopy of the synovial samples showed a mild degree of synoviocyte hyperplasia and hypertrophy with a mild to moderate mixed chronic inflammatory cell infiltrate composed predominantly of plasma cells with smaller numbers of lymphocytes. Histology of the femoral head showed a range of degenerative and regenerative changes within the articular cartilage and subchondral bone, the remainder of the femoral head appearing unremarkable with no evidence of avascular necrosis or other significant abnormality (Figure 3). The articular cartilage was predominantly smooth although there was an occasional focus of fibrillation. Fissures were not prominent although an occasional horizontal crack was present. The articular cartilage changes included areas of chondrocyte loss which in some areas was full thickness in nature but in other foci deep zone chondrocytes remained viable (Figure 3A). In other, often neighbouring foci, there was evidence of intrinsic cartilage repair with prominent chondrocyte cluster formation (Figure 3B & 3C). Within the articular cartilage, proteoglycan content, as shown by toluidine blue staining was mostly maintained although staining was decreased in areas of extensive chondrocyte loss or in relation to chondrocyte clusters. In many of the sections viewed there was extensive vascular penetration of the subchondral bone plate and invasion of the articular cartilage (Figure 3D). This was independent of whether the articular cartilage was viable or showed full thickness cell death. Where extensive, the vascular invasion was associated with a fibrous pannus containing a scattering of

lymphocytes and plasma cells. In some sections a similar fibrous pannus overlay the surface of the articular cartilage whereas in others the full thickness of the articular cartilage was replaced (Figure 3E). In one of the blocks the process of vascular invasion and intrinsic cartilage repair appeared to result in the formation of a new articular cartilage surface with subchondral bone plate that was separated from the partially resorbed pre-existing articular cartilage by trabecular bone and fatty marrow (Figure 3F). The histological features were not consistent with those of osteoarthritis and a diagnosis of idiopathic chondrolysis of the hip (ICH) was made.

The patient described immediate symptomatic improvement following the operation. He was mobilising independently on the day of surgery and was successfully discharged 2 days later. He experienced no immediate complications during this period.

The patient was satisfied with the significant reduction in hip pain and is now 2 years post-procedure. He is managing to mobilise independently without walking aids and is increasing his walking distance on a daily basis. The affected hip has a good range of pain free movement, similar to that of his unaffected hip. During this time, he has experienced no symptoms in his other joints. The patient has now returned to work and he will be followed up in three years, as per our local policy. It is expected that he will likely require at least one further operation for his hip over the course of his lifetime.

DISCUSSION:

Chondrolysis is a rare disorder, first described by Waldenstrom, which can affect any diarthrodial joint and is associated with a rapid loss of articular cartilage [1, 2]. Provencher et al. systematically appraised the published literature on joint chondrolysis and broadly characterised the potential aetiologic contributors as mechanical, chemical or thermal [1]. In the hip, it is a widely recognised complication of slipped capital femoral epiphysis and has also been associated with several other factors including trauma, infection, and immobilisation [3].

However, a significant proportion of cases will have no identifiable disease mechanism [1]. ICH was first described in the mid-seventies [4,5]. Since this time, less than 130 cases have been established in the medical literature and as a result its true incidence remains unknown [3]. Cases are most frequently reported in young females and are typically unilateral. The most common age at presentation is 11, and all confirmed cases have begun before the end of adolescence [3]. Wada and colleagues [6] previously reported on suspected adult-onset ICH (aged 28 and 37 at onset, respectively) but, the authors concede that whilst clinically similar to ICH, the histopathological findings from each case were characteristic of osteoarthritis. Similarly, Sivananthma and Kutty [7] described the case of a 20 year old male thought to have clinical findings highly suggestive of ICH. However, the case they described had a relapsing-remitting course and subsequent pathology showed evidence of avascular necrosis, making the diagnosis of chondrolysis unlikely.

Diagnosis and classification of chondrolysis is challenging as the majority of reported cases lack clear documentation of initial insult, aetiology, treatment and outcome. Thus, differentiating true chondrolysis from other pathologic conditions of articular

cartilage has been noted as a genuine problem within the current medical literature as many reports are affected by so-called ‘pooling effects’ [1]. Provencher et al. recommended that the term ‘chondrolysis’ be applied to patients who are seen within twelve months of an initial insult which results in cartilage damage – such as surgery, slipped capital femoral epiphysis or infection – and who present with pain, stiffness, reduced range of joint motion and severe loss of articular cartilage which is present radiologically and macroscopically [1]. However, this definition fails to take into account so-called ‘idiopathic’ chondrolysis - a condition reported to affect almost one third of all cases involving the hip [1, 3].

Although rare, there are subtle differences which may assist in the clinical differentiation of chondrolysis of the hip from osteoarthritis. Fundamentally, ICH is characterised by worsening hip or groin pain, which occurs over a shorter time span (months rather than years) and affects a younger patient cohort who have no background underlying skeletal dysplasia [1, 3, 8]. The clinical course is rapidly progressive and associated with global joint stiffness. Examination findings are similar to severe osteoarthritis and an antalgic gait with fixed flexion contractures of the hip is frequently noted. Range of motion is reduced in all planes and leg length differences may be observed with subsequent compensation at the pelvis and lumbar spine.

Bleck first documented premature physeal fusion, protrusion acetabuli and widening of the femoral head and neck as the common radiographic changes in ICH [9]. Joint space narrowing to within less than 3mm, regional osteopenia, and the absence of osteophyte formation (differentiating ICH from osteoarthritis) has also been noted

[10]. Bone scintigraphy shows normal bone with diffuse periarticular uptake suggestive of an underlying inflammatory reaction [11]. Reported MRI findings in ICH are variable and may change throughout the course of this disease. Early reported findings include abnormal signal intensity in the proximal femoral epiphysis, acetabular bone marrow oedema, mild synovial hypertrophy, and minimal joint effusion [12]. Contrast-enhanced imaging has identified mild to moderate synovial enhancement in this group. Extensive marrow oedema present in the femoral head, neck and acetabulum have been described by Johnson et al. and may reflect later progression of this disease process [12, 13].

Stanescu et al. reported on a case of 'precocious bilateral hip arthrosis' in a female whose symptoms began at the age of 12 years. These authors identified chondrolysis as a form of cartilaginous dysplasia which typically affects weight bearing areas and spreads peripherally in the hip [14]. Specimens showed a loss of articular cartilage in the hip associated with non-specific inflammation and superficial chondrocyte necrosis. Korula et al. performed synovial biopsies on a cohort of twenty patients with ICH and identified hyperplastic synovium with non-specific chronic inflammation and occasional perivascular infiltration by lymphocytes and plasma cells in 95% [15]. We believe that the current case fulfils the accepted criteria for ICH and, as such, is the oldest recorded presentation.

Difficulties in ascertaining the true incidence of ICH have precluded any form of consensus regarding the best treatment of this condition. As it arises in a young patient population some centres favour conservative management over more invasive interventions, however a solid evidence base for such a strategy is currently lacking

[3, 10]. It could be argued that total joint arthroplasty ultimately remains the gold standard for treatment of extensive articular cartilage damage, regardless of the underlying pathogenesis, with other centres finding that surgery can have a profoundly positive impact by reducing pain and improving overall range of motion in cases of ICH [8, 15].

Although rare, ICH is an extremely debilitating condition which should be considered in the differential diagnosis of adolescent and young adult hip pain. In the event of rapidly progressive disease specialist opinion should be sought when more common conditions have been excluded.

CONFLICT OF INTEREST: The authors declare that they have no conflict of interest.

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SUPPLEMENTARY MATERIAL

TABLES: Nil

FIGURE LEGENDS:

1. (A) Initial antero-posterior (AP) radiograph of right hip showing loss of joint space and juxta-articular osteopenia in a 23 year old male. (B) AP Radiograph of pelvis taken six months later shows progressive loss of joint space and intense osteopenia. (C) Magnetic resonance imaging (STIR sequence) showing synovial thickening and bone marrow oedema in the proximal femur and acetabulum. (D) Magnetic resonance imaging (fat-suppressed proton density image) showing synovial thickening and bone marrow oedema in the proximal femur and acetabulum.
2. (A) Histology of the femoral head showing chondrocyte loss in the superficial and mid-zones of the articular cartilage (#). Haematoxylin & Eosin stain (H/E); magnification x10; (B, C) Areas of articular cartilage showing intrinsic cartilage repair with chondrocyte cluster formation (block arrows). (B) H/E; magnification x10, (C) Toluidine blue x4; (D) Vascular penetration of subchondral bone plate into articular cartilage H/E; magnification x20; (E) The articular cartilage surface (+) has been replaced by a fibrovascular pannus (**) containing small numbers of inflammatory cells. H/E; magnification x2. (F) Formation of a new articular cartilage and subchondral bone plate (#) separated from the pre-existing articular cartilage (*) by trabecular bone and fatty marrow (##). H/E original magnification x2.

FIGURES:

Figure 1

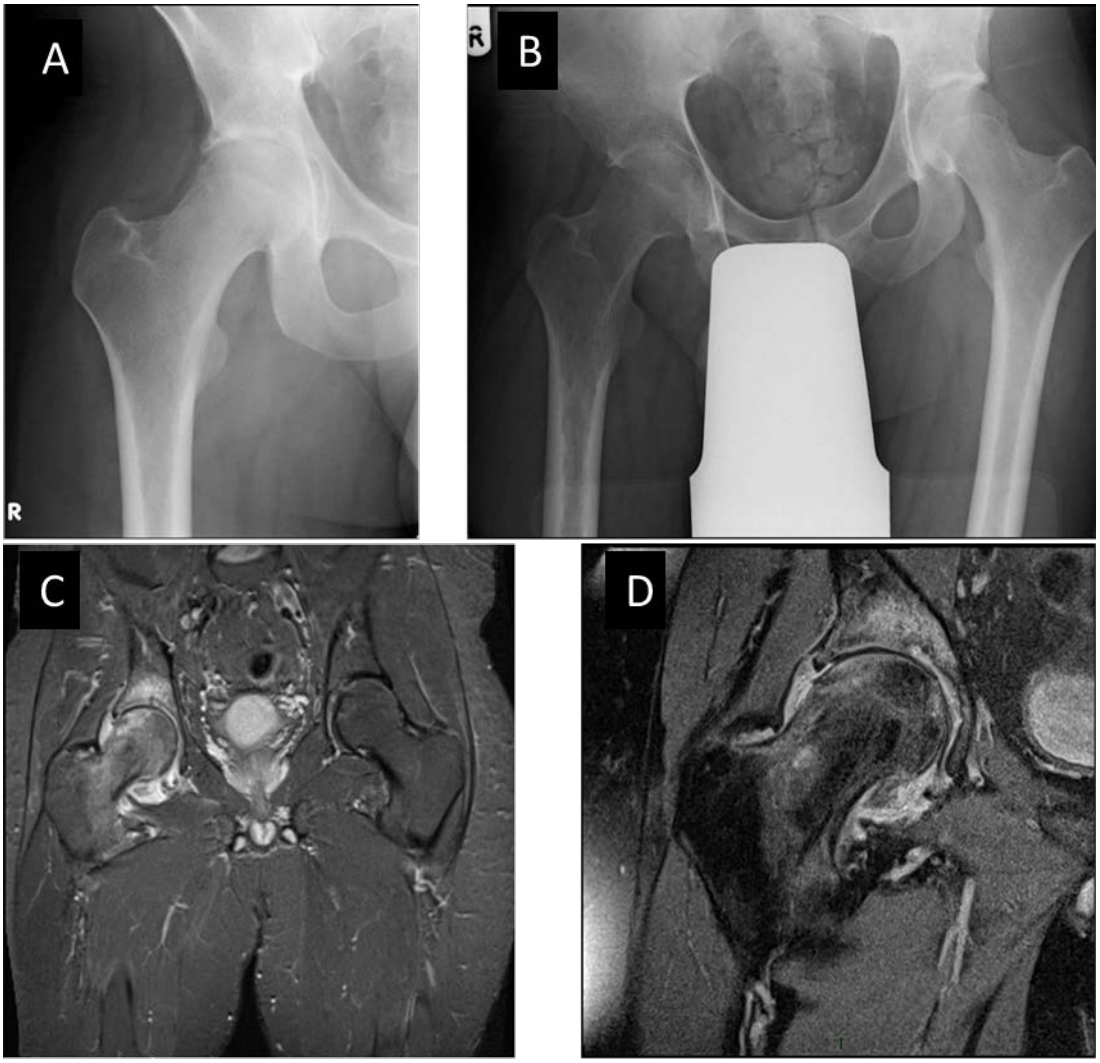

Figure 2