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CASES WITH A MESSAGE

Idiopathic hip chondrolysis: a case report of a Caucasian HLA-B27 positive adolescent with a history of long walking

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Abstract

Idiopathic hip chondrolysis is a rare disorder, the pathophysiology of which has not been fully elucidated. Several theories have been proposed regarding the cause of the disease with some of them involving autoimmune-mediated cartilage destruction. There are several similar features between idiopathic hip chondrolysis and rheumatologic diseases such as juvenile idiopathic arthritis, so whether these two disorders are different or not is still debatable. This case report aims to help comprehending this complex disorder by presenting a case of idiopathic hip chondrolysis with apparent risk factors, such as repetitive microtrauma and presence of HLA-B27 antigens. A 15-year-old HLA-B27 positive male presented with idiopathic hip chondrolysis after excessive walking. Initial treatment consisted of medications including corticosteroids, protected weight bearing and surgical soft tissue release. After failure of all these modalities in restoring the decreased range of motion of the hip, a course of a TNF-inhibitor, etanercept was tried. Alleviation of pain achieved early in the treatment period, but range of motion remained mainly unchanged. Although there was a brief improvement of stiffness for a short period after surgery which lasted for about 3 months, stiffness came back afterwards. Administration of a TNF inhibitor in the following period significantly improved his range of motion. The presence of laboratory findings indicating an autoimmune tendency in this patient supports the hypothesis of susceptibility of these patients to autoimmune reactions, while excessive walking was an apparent trigger factor. In future, traditional treatments may be abandoned in favor of novel medications targeting immunologic pathways.

Keywords Idiopathic hip chondrolysis · Surgical treatment · HLA-B27 · Etanercept

Introduction

Idiopathic chondrolysis of the hip (ICH) is an uncommon disease characterized by rapid and extensive loss of the articular cartilage of the femoral head and acetabulum, without any identifiable cause. It is seen predominantly in adolescent girls, with a mean age of 11 and a range of 3–20 years of age [1]. Its true incidence may be underestimated since many cases of ICH may be undiagnosed and patients may be treated for juvenile idiopathic arthritis (JIA) or post-traumatic arthritis. Due to the non specific initial symptoms, physicians must be aware of this disorder to diagnose and treat any case of ICH in time. The similar features of ICH with certain types of juvenile idiopathic arthritis (JIA) may result in wrong diagnosis, although there may be a common field between ICH and JIA and in many cases patients fulfill criteria for both diseases.

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The true cause and pathogenesis of idiopathic hip chondrolysis has not been elucidated yet. Many risk factors have been proposed for the development of the disease, including repetitive microtrauma, an acute minor trauma, a positive autoimmune background or even bacteria originating from remote areas [2, 3]. A thorough examination of the complex autoimmune mechanisms that may be implicated in the pathogenesis of the disease may be the key for the development of successful treatment strategies [3–5].

This is a case report of a patient who presented with symptoms of ICH, the pathogenesis of which may be justified by the presence of certain risk factors such as the constant repetitive trauma due to excessive walking and the positive autoimmune background as indicated by the presence of HLA-B27 antigens. The presence of these two commonly suggested causative factors in this patient further supports the theory that there is a genetic tendency for autoimmune reactions in these patients, and that repetitive trauma may trigger development of the disease. Moreover, successful results after administration of etanercept, a TNF inhibitor further justifies this hypothesis.

Case report

A 15-year-old male presented to the emergency department of our hospital with complaints of left (dominant side) hip pain and limp for the past 2 months. The patient was a refugee from Syria and for the past 3 months he had been walking extensively as he had to cover a long distance from Syria to Greece. He described a constant pain 6–9/10 that was exacerbated after walking roughly 2000 m. During flare-ups which usually lasted for 1–2 days, the patient had to stop for the following days to alleviate pain. The patient denied any history of injury or infection during the past months. He had no other comorbidities and his family history was negative except from hypertension and dyslipidemia in his father's history. On physical examination there was severe restriction of both active and passive hip range of motion (10°–60° flexion, up to 30° abduction, 0°–15° internal rotation and almost zero external rotation), but no signs of infection or inflammation (tenderness, warmth, erythema and swelling or effusion). Lumbar examination was also normal. Preliminary diagnoses included ICH, JIA, and atraumatic osteonecrosis of the femoral head.

Initial blood work-up including CBC, ESR, CRP, basic metabolic panel, blood cultures and immunology tests (RF, HLA-B27 antigens, ANA, anti-CCP, C3 and C4 complement) revealed positive HLA-B27 and moderately elevated CRP and ESR levels (5 mg/dL and 15, respectively) which were attributed to a viral pharyngitis that was present at that time. Radiographs demonstrated diffuse narrowing of the joint space, while magnetic resonance imaging (MRI) showed altered signal intensity due to marrow edema, marked cartilage thinning and low-volume joint effusion (Figs. 1, 2). The tuberculin skin test was falsely positive due to recent vaccination since subsequent Interferon Gamma Release Assay (IGRA test) was negative. Rheumatology consultation was also obtained. JIA was not considered a likely diagnosis since there were no imaging findings demonstrating inflammation in the sacroiliac joints or involvement of spine. Moreover, the presence of certain clinical features indicated that ICH was much more likely than JIA. These features included the location and course of symptoms which remained confined around hip, while isolated hip arthritis is rare as a presenting feature of JIA. Additionally, the severe and rapid restriction of movements, the absence of back and sacroiliac pain or tenderness and the fact that stiffness was not more prominent in mornings, further justified diagnosis of ICH. Ophthalmologic examination was also performed looking for any signs of iridocyclitis or uveitis indicating JIA, but no such findings were identified. The patient was treated with methylprednisolone (16 mg/day) and NSAID, and skin traction was applied. Few days later, he reported significant improvement of pain (4/10).

A month later the patient returned for re-evaluation. He reported decreased pain (3/10), but there was no improvement on physical examination regarding range of motion. A treatment consisting of physical therapy (including continuous passive motion exercises), protected weight bearing and corticosteroids at the same dose (16 mg/day) was advised. The next follow-up was scheduled for 3 months later. On the next follow-up (4 months after the initial evaluation), pain remained at low levels (2–3/10) but the patient reported increased difficulty during walking and a more severe limp. Physical examination revealed severe restriction of motion

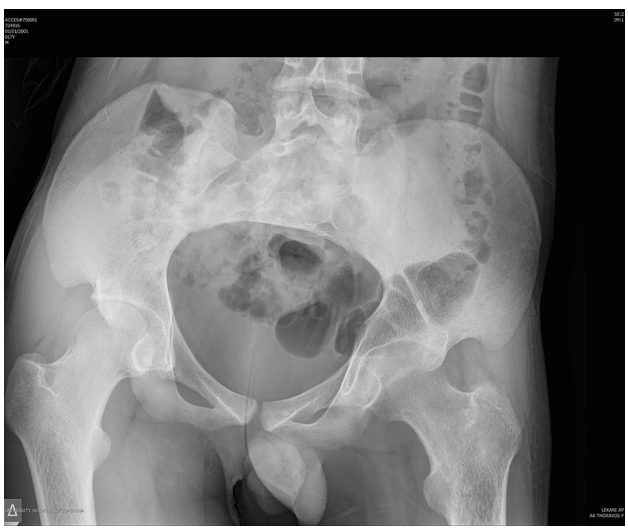

Fig. 1 Pelvis X-ray at initial presentation showing left hip chondrolysis with joint space narrowing. Pelvic obliquity is also apparent

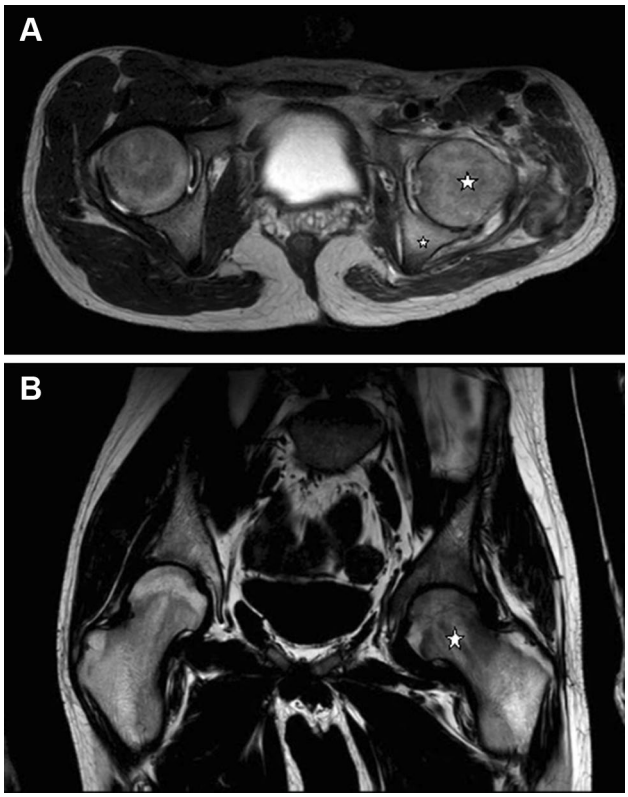

Fig. 2 **a** Axial T2-weighted MRI image at initial presentation. Note the high signal intensity within the femoral head and acetabulum (asterisks), the cartilage thinning, the small joint effusion and the marked gluteal atrophy. **b** Coronal T1-weighted MRI image also showing the altered signal intensity within the femoral head (asterisk)

(10°–30° flexion, 20°–30° abduction and no internal or external rotation), thus surgical treatment was decided. The patient underwent a hip arthroscopy for soft tissue release along with partial capsulectomy. During the procedure, the approach was extremely difficult due to extensive contractures, while assessment of the central part of the joint was impossible due to almost complete obliteration of the joint space. A sample of the capsule and articular cartilage was also obtained for histological evaluation and tissue culture. Pathology report mentioned non-specific signs of chronic inflammation and features consistent with cartilage degeneration (Fig. 3), while tissue culture was also negative. Postoperatively, the patient was placed on a continuous passive motion machine and an intense physiotherapy protocol was followed.

Further, follow-up 1 month after the surgery (5 months after the initial evaluation) was done during which the patient reported no pain. Additionally, physical examination showed slight improvement of his hip range of motion: 10°–50° flexion, 0°–30° abduction, 0°–15° internal rotation, 0°–10° external rotation. Next evaluation 2 months later (7 months after the initial evaluation) showed that

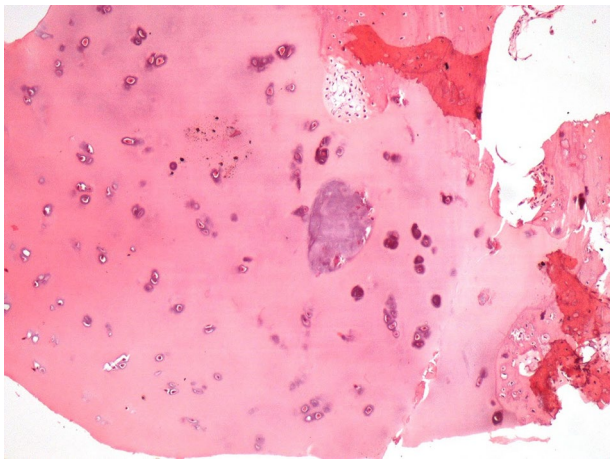

Fig. 3 A microphotograph showing cartilage with considerable loss of chondrocytes and early degenerative changes (haematoxylin-eosin × 100)

the improved range of motion that was achieved after surgery was not maintained and the patient returned to his preoperative status, having a stiff joint with limited range. At that time, a second rheumatology consultation was obtained and a course of etanercept (TNF inhibitor, 50 mg once weekly) for the following months was decided. Etanercept was decided based on the fact that although ICH was diagnosed there were several features commonly found in seronegative arthropathy (i.e., HLA-B27 antigens, young male patient) and that there were other reports of improved outcomes with etanercept in such patients. The next follow-up was 3 months later (10 months after the initial presentation) during which the patient remained free of pain (0–1/10) but with no improvement regarding range of motion, so the patient was scheduled for reevaluation 3 more months later. Physical examination during that evaluation (13 months after the initial presentation and 6 months after initiation of etanercept) showed for the first time marked improvement of his hip range of motion: flexion from 10° to 80°, abduction from 0° to 30°, 0° to 20° of internal rotation and 0° to 15° of external rotation. An additional MRI at the time of that follow-up depicted similar findings to the previous MRI, but with slightly decreased bone marrow edema. The last follow-up was 16 months after initial presentation and 9 months after first administration of etanercept. The patient reported that stiffness was significantly improved during the past 6 months and that limp had also subsided. Physical examination showed that the improved range of motion that had already been observed during the previous follow-up was still maintained (flexion from 10° to 80°, abduction from 0° to 30°, 0° to 20° of internal rotation and 0° to 15° of external rotation).

Discussion

The first description of the disease was back in 1971 by Jones who reported on a series of cases with loss of articular cartilage with no underlying cause [6]. The natural history of the disease has been described by Segaren et al. who reported that there are three possible clinical outcomes: complete resolution of the symptoms, painless but stiff joint or in the worst case painful and malpositioned hip ankylosis [1]. Medications include NSAIDs, analgesics and corticosteroids, while disease modifying agents or even botulinum toxin injections have been recently used [7].

Idiopathic chondrolysis is mainly a diagnosis of exclusion and other disorders including septic arthritis, aseptic osteonecrosis, chondrolysis secondary to slipped capital femoral epiphysis and JIA must be ruled out [7, 8]. In our case, aseptic femoral head osteonecrosis was one of our preliminary diagnoses. The possibility of this disorder was ruled out by the absence of risk factors and by the MRI findings which were not consistent with those of osteonecrosis (single-density "band-like" lesion on T1-weighted images, and a "double-line" sign on a T2-weighted image) [9]. Idiopathic hip chondrolysis and certain types of JIA (oligoarticular or enthesitis related arthritis [ERA]) have almost identical features thus differential diagnosis between these two diseases is quite difficult, while some others believe that these two entities are the same and must be treated as such [10]. Main clinical features of ICH include severe restriction of hip movements in all directions, insidious onset of hip pain and limp. Flexion contracture along with muscle wasting develops shortly in the course of the disease resulting in fixed deformities, pelvic obliquity and apparent limb length discrepancy. Pain, stiffness and limp are also the main symptoms of JIA. Although stiffness is a typical symptom of JIA, there are some distinctive features of stiffness that help differentiating ICH from JIA: morning stiffness is more prominent in cases of JIA while in cases of ICH there is no association with time. Additionally, in cases of ICH stiffness progress rapidly, leading to fixed deformity early in time, while this is not usually evident in JIA. Even though four or fewer joints are involved in oligoarticular JIA, monoarthritis is extremely rare thus monoarticular involvement favors ICH [11, 12]. In cases of ERA, sacroiliac joints are usually involved, whereas these joint are unaffected in ICH. Moreover, iridocyclitis, uveitis, enthesitis and back pain are common manifestations of JIA but not of ICH.

Typically, the patient presented in this study fulfilled three criteria for diagnosis of enthesitis-related arthritis (ERA): arthritis, the presence of HLA-B27 antigens and onset of disease in a male over 6 years of age, thus ERA cannot be excluded [13, 14]. However, the absence of SI joint tenderness, enthesitis, inflammatory lumbosacral pain or acute anterior uveitis, and the fact that he had no first degree relatives with ankylosis spondylitis, ERA, sacroiliitis with IBD, Reiter's syndrome or acute uveitis make diagnosis of ERA debatable and ICH remains the most likely diagnosis in our opinion. Certain laboratory findings in our patient favor diagnosis of ICH over oligoarticular JIA. ANA were absent, whereas HLA-B27 antigens were present, while in oligoarticular JIA there is a high frequency of positive ANA and negative HLA-B27 antigens. Moreover, imaging findings in JIA and ICH are not similar and they may help when clinical and laboratory findings are equivocal. MRI in ICH demonstrates a more rapid and extensive destruction of the articular cartilage than in JIA, while a hypervascular synovium with significant contrast enhancement is a prominent feature of JIA [15, 16]. In our case, MRI did not demonstrate contrast enhancement of synovium, while sacroiliac joints and spine were not involved.

It has been suggested that development of the disease is the result of an autoimmune-mediated cartilage destruction triggered by an unknown event in a genetically susceptible individual [1]. This hypothesis is further supported by the fact that there are similar case reports with HLA-B27 positive patients [2, 3]. Since HLA-B27 antigens are commonly found in certain autoimmune diseases such as ankylosing spondylarthritis, the presence of these antigens in patients with ICH indicates that probably similar autoimmune reactions are involved in the development of ICH or even that there is a common field between ICH and seronegative arthropathies. Furthermore, Appleyard et al. mentioned that a 9-month treatment with a TNF inhibitor (which is commonly used in HLA-B27 positive patients with ankylosing spondylitis) in a patient with ICH resulted in significant improvement of range of motion [5]. Etanercept was decided in our patient only after all other surgical and conservative means of treatment had failed and resulted in significant improvement of hip range of motion as well.

The case described herein has several strengths. A wide set of laboratory tests, imaging studies and biopsy of the joint capsule were performed, with serial imaging evaluation. In addition, both surgical and medical treatments were tried (including disease modifying agents), thus a more intimate assessment of the existing treatment options can be done. There are also certain limitations of the study that must be addressed. Evaluation of treatment was mainly based on volumetric movements of hip to assess progression of stiffness, while more subjective methods of evaluation such as serial ultrasound would be preferred. Additionally, the results may be limited by the relatively short follow-up.

This case report aids in comprehension of the pathogenesis of the disease and of its natural history when both

surgical and medical treatment are used. In a susceptible to autoimmunity patient as indicated by the presence of HLA-B27 antigens, a repetitive trauma such as excessive walking may incite a hyper-inflammatory response resulting in rapid destruction of the articular cartilage. This is the third positive HLA B27 case of ICH in the literature. The growing number of such findings raises more interest about the immunologic background of the disease. In this direction, more studies are needed to further assess the association between the disease and a possibly disoriented immunology system. Whether ICH is a subtype of JIA or it is a totally different disorder must be considered given that there is a gray zone between these two diseases. However, the answer to that question is worthy only because in those cases diagnosed as ICH novel treatment modalities that are used for JIA including disease-modifying agents and biologic factors may prove to be more efficient than the current treatment options for ICH.

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Compliance with ethical standards

Ethical approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflict of interest All authors declare no conflicts of interest.

Informed consent The patient's parents have given their informed consent to use the patient's medical data and photos in the publication of this case study.

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