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Liposynovitis prepatellaris in a child (Hoffa's syndrome): Lessons from MRI

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KEYWORDS

Hoffa's syndrome; Knee injuries; Liposynovitis; Prepatellaris; Juvenile idiopathic arthritis (JIA) **Abstract** *Background:* Liposynovitis prepatellaris (Hoffa's syndrome) is a rare condition in children and rarely discussed in the literature. Hoffa's syndrome can lead to an obscure anterior knee pain resulting from impingement and inflammation of the infrapatellar fat pad.

Aim of the work: The aim of this case report is to increase awareness among rheumatologists about this condition among children to avoid erroneous diagnosis of juvenile idiopathic arthritis (JIA) and unnecessary treatment with disease-modifying antirheumatic drugs (DMARDs).

Case report: In this report we presented a 12 year-old child with this condition who presented with chronic pain and intermittent swelling involving his right knee. The patient was wrongly diagnosed as a case of JIA and wrongly treated with DMARDs for three years duration. The report will shed light on the characteristic MRI features of this condition and the value to order MRI in such atypical presentation.

Conclusion: Hoffa's syndrome can present with chronic arthropathy in children that can mimic mono-articular JIA presentation and eventually unnecessary treatment with DMARDs. MRI is generally very helpful from the diagnostic point of view, it clearly depicts Hoffa's infrapatellar fat pad entrapment and its findings may suggest Hoffa's syndrome.

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1. Introduction

Liposynovitis prepatellaris, Launois-Bensaude's syndrome, fat pad syndrome, and lipoid dermatoarthritis are all names used to describe Hoffa's disease. Fat pad syndrome was first described in the literature by Hoffa in 1903, to describe a condition where the infrapatellar fat pad was impinged between the patella and the femoral condyle. Hoffa's fat pad is an intracapsular, extrasynovial structure that fills the anterior knee

1110-1164 © 2013 Egyptian Society for Joint Diseases and Arthritis. Production and hosting by Elsevier B.V. Open access under CC BY-NC-ND license. http://dx.doi.org/10.1016/j.ejr.2013.01.007 compartment, and is richly vascularized and innervated. Its degree of innervation, the proportion of substance-P-containing fibers and close relationship to its posterior synovial lining implicates infrapatellar fat pad (IFP) pathologies as a source of infrapatellar knee pain [1]. Moreover Hoffa's fat pad contains residual synovial tissue, meaning that primary neoplastic conditions of the synovium may originate and be confined to the fat pad [2].

Hoffa's fat pad separates the patellar tendon from the top end of shin bone (tibia) acting as a shock absorber to protect the knee. The patellar tendon connects the kneecap to the shin bone and is just below the kneecap. Fat pad impingement is when the fatty soft tissue gets pinched between the patella tendon and the end of the shin bone and impingement is usually caused by overextending the knee joint, where the knee is forced beyond its fully straightened normal position.

2. Case presentation

A 12 year-old male patient presented with chronic right knee pain and intermittent swelling for a 3 year duration. The patient used to play football regularly and reported previous trauma to the right knee joint with forced hyper extension. The parents of the child seek second opinion in the rheumatology clinic in our facility (Dr. Erfan and Bagedo General Hospital, Jeddah, KSA). At that time the patient was previously diagnosed as a case of juvenile idiopathic arthritis (JIA) in spite of normal ESR, normal CRP levels, additionally the patient lacks any constitutional manifestations usually dominating the course of JIA with negative rheumatoid factor (RF), negative antinuclear antibodies (ANA) and anti-cyclic citrullinated peptide (anti-CCP) antibodies. Clinically the patient has only mild knee effusion with pain and tenderness mainly under the kneecap (anterior knee pain), while other joints are totally free. Positive Hoffa's test was observed, with the patient in lying with his knees bent, the examiner presses both thumbs along either side of the patellar tendon, just below the patella. The patient experienced pain and apprehension after he was asked to straighten the affected leg, the latter considered a positive sign for IFP impingement.

The patient was wrongly receiving disease-modifying antirheumatic drugs (DMARDs) in the form of oral methotrexate (10 mg/week) for a three year duration and showed no clinical improvement on treatment relative to pain and/or swelling.

Gadolinium enhanced MRI was performed to revaluate the diagnosis and to search for any associated MRI signs like synovial thickening, bone marrow edema or soft tissue edema suggestive of inflammatory joint disease and importantly to exclude any other pathology that can mimic chronic arthropathy [3].

MRI showed a typical pattern of Hoffa's syndrome with heterogeneous dark signal lesion in the IFP on Sagittal T2 sequence (Fig. 1a, with heterogeneous enhancement of the lesion on post contrast T1 fat saturation (Fig. 1b). The later findings are consistent with the chronic stage of Hoff's syndrome which precluded the need to perform diagnostic biopsy.

The parents of the child were reassured and unnecessary DMARDs were stopped and the patient underwent arthroscopic removal of infra patellar fat pad followed by knee rehabilitation and showed marked improvement and stayed pain free after the procedure.



Figure 1 (a) Sagittal T2 showing heterogeneous dark signal lesion in the infrapatellar fat pad (white arrow), (b) Sagittal post contrast T1 fat sat showing heterogeneous enhancement of the lesion (white arrow).

To the authors' knowledge, this condition is infrequently discussed in both the rheumatology and the radiologic literature.

3. Discussion

In this report we described a 12-year old child who presented with chronic Hoffa's disease and wrongly diagnosed as a case of JIA due to a chronic course of knee pain with intermittent attacks of joint pain and swelling. Unfortunately the patient wrongly received DMARDs for a three year duration. The lack of any constitutional manifestations in our case and normal values of inflammatory mediators like ESR and CRP levels were strongly against the diagnosis of JIA.

Hoffa's syndrome occurs as a result of an awkward fall or a knock on the knee joint and usually affects athletic individuals. Clinically tenderness and/or swelling around the bottom and under the kneecap are the common signs at presentation. MRI clearly depicts Hoffa's infrapatellar fat pad, and its findings may suggest the frequently ignored diagnosis of Hoffa's syndrome [2].

Because disease in this region is not uncommon, it is important to be familiar with the various pathologic entities that may occur here. Abnormalities that are intrinsic to this fat pad include Hoffa disease, intracapsular chondroma, localized nodular synovitis, postarthroscopy and postsurgery fibrosis, and shear injury. In addition, the IFP may be involved secondarily from extrinsic processes, including articular disorders, synovial abnormalities (e.g., pigmented villonodular synovitis; synovial hemangioma; primary synovial chondromatosis; chondrosarcoma; lipoma arborescens) and anterior extracapsular abnormalities. The approach to pathologic processes involving the IFP of Hoffa is simplified when one is familiar with regional anatomy and possible differential diagnostic considerations [4].

Hoffa's disease causes an obscure cause of anterior knee pain resulting from impingement and inflammation of the IFP. This condition is characterized by traumatic and inflammatory changes occurring in young athletes, which may lead to pain, swelling and restricted motion of the knee. Hypertrophy of the fat pad may cause it to become trapped between the tibia and femur when the flexed knee is extended suddenly and eventually fibrosis may ensue [2].

On MR images, acute findings indicate the presence of fluid and chronic findings resembling those of scarring after knee arthroscopy. In some patients, rupture of the overlying synovial fold and haemarthrosis may occur after an injury to the IFP [4].

Despite reports of eventual fibrocartilaginous transformation and ossification of the fat pad, no relation has been described between chronic impingement and the development of ossifying chondroma. In previous report Krebs and Parker [5], described extrasynovial ossifying chondroma of the IFP and they consider it as end-stage Hoffa's disease. The authors explained a possible relationship between impingement of the IFP and the development of ossifying chondroma may exist [5].

In our report the patient presented with chronic knee pain and intermittent knee swelling and wrongly diagnosed as a case of JIA. The clinical suspicion and the typical MRI features clearly lead to the appropriate proper diagnosis in our case. Importantly the MRI features also excluded other MRI signs of inflammatory joint disease usually existing in JIA like synovial enhancement and hypertrophy, bone marrow edema and/or soft tissue edema which are common MRI signs that can be observed in early undifferentiated cases of inflammatory arthritis [3].

MR imaging is also important as routine arthroscopy does not visualize the fat pad well enough and it does not give sufficient morphological and intrinsic information [2]. Furthermore in patients who are suspected of having IFP impingement, such MRI findings should be considered and distinguished from other causes of anterior knee pain [6]. Sagittal MRI is the most common imaging technique used to assess the IFP pathology including fibrosis, inflammation, edema, and mass-like lesions [2]. The MRI typically reveals the following signs: a hypointense lesion in the infrapatellar pad of fat in T2-weighted images (WI), and in post contrast T1 WI with fat saturation the lesion demonstrate no enhancement. In Short TI Inversion Recovery (STIR) sequence the lesion demonstrates slightly hyperintense lesion with or without knee effusion. Additionally in patients with Hoffa's fat pad impingement, morphologic changes such as localized edema of the superior and/or posterior part of the fat pad, a deep fluid-filled infrapatellar bursa, non-visualization of vertical and/or horizontal clefts, fibrosis, and calcifications were noted on MR imaging with remarkable frequency [7]. Moreover a high-riding patella, a short distance between the patellar ligament and the lateral trochlear facet, and an increased distance from the tibial tubercle to the trochlear groove are associated with superolateral Hoffa's fat pad edema at MR imaging [8]. Nevertheless patients with symptomatic IPF impingement often exhibit fat pad edema on MRI in the superolateral region of Hoffa's fat pad [9]. Finally edema in the superolateral portion of IFP is usually associated with patellar maltracking [10]. Arthroscopic partial resection for IFP impingement and Hoffa's disease has showed favorable results. Additionally arthroscopic debridement of IFP fibrosis has been successfully used to treat extension block following anterior cruciate ligament reconstruction [1].

In conclusion Hoff's syndrome is not an uncommon cause of knee pain and should be considered in the differential diagnosis of knee injuries especially in sports-related injuries or after neglected anterior knee trauma. MRI is quite helpful for the optimum diagnosis and to exclude other diagnoses. Arthroscopic removal of the inflamed pad of fat is an essential way of managing the disease, together with intensive postoperative rehabilitation. The case presentation is intended to increase awareness among rheumatologists to put in mind this entity that can present with chronic knee arthropathy, and may lead to wrong diagnosis and eventually unnecessary treatment with DMARDs.

4. Conflict of interest

All the authors responsible for this work declare no conflict of interest of any kind.

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