PROTRACTED ATHRITIS OF FAMILIAL MEDITERRANEAN FEVER WITH BILATERAL COXARTHROSIS

A CASE REPORT

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ÖZET

AİLEVİ AKDENİZ ATEŞİNE BAĞLI UZAMIŞ ARTRİT VE BİLATERAL KOKSARTROZ: Olgu sunumu.

Ailevi Akdeniz Ateşi hastalığında uzamış eklem tutulumu nadirdir ve genellikle tam fonksiyonel ve anatomik iyileşme ile sonuçlanır. Burada, 34 yaşında Ailevi Akdeniz Ateşi sonucu bilateral kalça eklemi dejenerasyonu, eklem hareket kısıtlılığı, radyolojik olarak ileri koksartroz ile fonksiyonel yetersizlikle sonuçlanan bir olgu sunulmuştur. Hasta kalça artroplasti operasyonu önerisini reddetmiştir.

Anahtar Kelimeler: *Ailevi Akdeniz Ateşi, Koksartroz.*

SUMMARY

Protracted articular attacks in Familial Mediterranean Fever (FMF) are uncommon and they end with almost complete functional and anatomical recovery. Here we report a case of 34 –year- old man with FMF who has bilateral degenerative joint damage of hips with limitation of motion, radiological appearance of severe bilateral coxarthrosis leading to residual incapacity. Although total hip replacement is recommended to the patient, he refuses it.

Key Words: *Familial Mediterranean Fever, Coxarthrosis.*

INTRODUCTION

Familial Mediterranean Fever (FMF) is an uncommon disease that predominantly affects Mediterranean people mostly Armenian, Turkish and Arab populations. It is inherited as an autosomal recessive trait. The characteristic symptoms include intermittent fever and pain in abdomen, chest or joints.

The musculoskeletal attacks in FMF occur together with or without of other manifestations and arthritis is present in almost half of the patients[1]. Asymmetric arthritis in the larger joints of the lower extremity is most typical. The affected articulations in order of decrease in frequency are the knees, ankles, hips, shoulders, feet, elbows, hands and wrists. Arthritis is commonly monoarticular, consists acute attacks of pain and swelling of one articulation at a time, sometimes related to minor trauma and subside in 2 to 3 days[2]. A small percentage of arthritic episodes develop into protracted arthritis with effusions persisting for months[3]. They end with almost complete functional and anatomical recovery except hips[4].

Hereby, we present a case with bilateral degenerative joint damage of hips secondary to FMF with protracted arthritis.

CASE REPORT

A 34-year-old man with a history of recurrent fever and pain in the abdomen and joints for twenty eight years was admitted to our hospital. The attacks occurred two times a month, lasting 3-4 days in the early years of the disease. When he was 16 years old he had his first acute arthritis attack in his knees, lasting two months which repeated several times. The attacks of arthritis occurred together with or without abdominal attacks and fever. When he was 17 years old he had been admitted to a hospital because of prolonged arthritis in his left knee. Synovectomy was applied but a rehabilitation program didn't follow it. The attacks in the left knee subsided but he couldn't bend his left knee in full range of motion. In a six-month period intermittent bilateral hip pain developed and the attacks continued with an increase of frequency and duration. At the age of 22, he was diagnosed as FMF and colchicine therapy was started. Following this treatment the

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number and severity of the attacks decreased. After six years the patient omitted colchicine therapy. During the last five years, his daily living activities was restricted due to the limitations of the hip joints. His mild abdominal pain and arthritis attacks continued infrequently.

When he was admitted to our hospital, he had difficulty in walking and sitting. He showed marked limitation in range of motion of both hip joints. Knee examination disclosed bilateral quadriceps atrophy marked in the left side with no effusion. The left knee had synovectomy scar and flexion was limited at 70°. Ophthalmologic examination was normal.

In the laboratory examination, acute phase reactants were normal with erythrocyte sedimentation rate 20 mm/h, serum fibrinogen level was 3.5gr/L. All biochemistry results were normal with no sign of amyloidosis. Rheumatoid factor, ANA, Anti-DNA, HLA-B27 tests were negative. Radiographic studies showed bilateral severe total hip joint space narrowing, osteophytosis, cyst formations, sclerosis, flattening of the femoral heads (Figure 1) and minimal degenerative changes in the knee joints.

Physiotherapy and exercise therapy was applied but the patient had little benefit. As the joint damage was marked total hip replacement (THR) was recommended, but the patient refused the treatment. Colchicine therapy of 1 mg/d is initiated in order to prevent from complications. Prominent changes in clinical findings hasn't been noted in the four year follow-up.

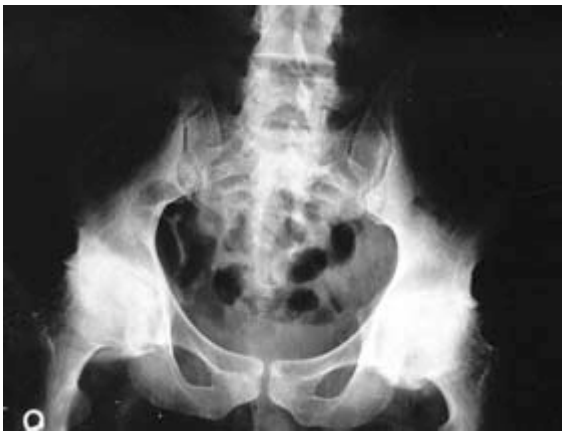

Figure 1: Bilateral severe joint space narrowing, osteophytosis, cyst formations, sclerosis and flattening of the femoral head.

DISCUSSION

Involvement of joints in FMF is reported by various investigators. The arthritis attack might present as either: a) short period attacks of up to 72 hours which subsides gradually with no articular residue, b) protracted attacks with sudden onset but failing to resolve and resulting in chronic arthritis persisting for months which occur 5% of the cases with arthritis, c) abortive attacks consisting of transient artralgia not accompanied by swelling which often result in failure to seek medical attention[3]. Our patient had abortive attacks at the first years of the disease, but after the age of 16 the attacks had protracted course so synovectomy was performed. It has been reported that synovectomy might control articular disease as it did in our patient[5,6] The reason of flexion limitation in the knees might be due to the lack of rehabilitation programme, as these patients require protracted rehabilitation[7].

Protracted arthritis develop in 5% of FMF patients with arthritis and mostly knee, hip, ankle joints are affected. Although the main feature of the protracted arthritis is the full recovery of the patient without any sequel, hip joint is the exception[4]. 84% of the affected hips during protracted arthritis develop residual incapacity, ranging from limitation of motion to complete ankylosis of the joint[8]. In the hip joint chronic effusion may cause avascular necrosis of the femoral head. Our patient had bilateral ankylosis of the hips due to protracted arthritis which restricted his daily living activities.

Zemer et al[9], reported that colchicine therapy prevents not only progression of protracted attacks but amyloidosis as well. Colchicine was administered to our patient when FMF was diagnosed at the age of 22. Though he used the drug regularly for six years and had benefit, he omitted the treatment, and his symptoms progressed in years. Protracted arthritis and the degenerative joint damage can also be attributed to colchicine discontinuation. When he was admitted to our hospital, colchicine treatment was administered on regular basis.

In patients who have restricted daily living activities due to degenerative arthritis, THR can be treatment of choice. THR, preferably cementless is recommended by Salai et al[10]. Rehabilitation after surgery is important. Our patient is also a candidate of THR but he hasn't accepted the operation yet.

Protracted arthritis in the hip joint of FMF patients may result with incapacitating damage. Therefore, when hip involvement develops preventive measures of arthrosis such as body weight control, therapeutic exercise, arrangement of daily living activities and occupation should be considered.

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