Familial Mediterranean Fever Associated with Ankylosing Spondylitis: A Case Report

Ailevi Akdeniz Ateşi ve Ankilozan Spondilit Birlikteliği: Olgu Sunumu

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Abstract
Familial Mediterranean fever (FMF) is an autosomal recessive disease of unknown origin. A less common manifestation of joint involvement in FMF is spondylartropathy, which is almost always HLA B27 negative. In these patients, along with sacroiliitis, inflammatory neck and back pain is also present producing minimal or no radiographic changes on plain films of lumbar and cervical spine. In this report, we presented a patient with FMF whom had been diagnosed to be ankylosing spondilitis (AS) in the light of some clinical and radiological findings. *(Rheumatism 2006; 21: 31-3)*

Key Words: Familial mediterranean fever, ankylosing spondylitis, seronegative spondylarthropaty

Özet
Ailevi Akdeniz Ateşi (AAA), etiyolojisi bilinmeyen otozomal resesif bir hastalıktır. AAA'de eklem tutulumunun nadir görülen bir formu genellikle HLAB27 negatif spondilartropatidir. Bu hastalarda sakroiliit ile birlikte inflamatuvar boyun ve bel ağrısı da gözlenir ve lumber ve servikal direk grafiler normal olabilir veya minimal değişiklik saptanabilir. Bu olgu sunumunda, klinik ve radyolojik bulgularına göre ankilozan spondilit (AS) tanısı koyduğumuz AAA'li bir hasta sunulmuştur. *(Romatizma 2006; 21: 31-3)*

Anahtar Kelimeler: Ailevi akdeniz ateşi, ankilozan spondilit, seronegatif spondilartropati

Introduction

Familial Mediterranean fever (FMF) is an outosomal recessive disease of unknown origin. It's characterized by recurrent attacks of fever and inflamation of serous membranes including the peritoneum, synovium and pleura (1). The joint involvement is the second most common manifestation after abdominal pain (1, 2). On the other hand a less common manifestation of joint involvement in FMF is spondyloartropathy, which is almost always HLA B27 negative (3). Sacroiliitis is a common manifestation of a number of diseases including ankylosing spondylitis (AS). To reach a diagnosis of AS, the patient should have some clinical and radiologic evidence along with sacroiliitis (4).

In this report, we presented a patient with FMF whom had been diagnosed to be AS light of some clinical and radiological findings.

Case Report

A 31-years-old Turkish woman with back and neck pain was admitted to our inpatient clinic. In the history, she had had arthritis in knee and ankle joints when she was 10 years old. This arthritic episode was than diagnosed as acute rheumatic fever. She had had recurrent attacks of arthralgia, fever and abdominal pain until she was 27. Four years ago, she was diagnosed as FMF after an attack of abdominal pain, fever and arthritis of left knee and right shoulder. She was started on 1 mg of colchicine daily and symptoms subsided. For the last two years she had been also suffering from back pain and morning stiffness with a duration of two hours. Her back pain worsened at rest and improved during activity. The analysis of MEFV gene mutation M694V homozygote mutation had been detected before she admitted our clinic.

On examination, movement cervical and lumbar spine was limited in all directions. Finger to floor distance was 28 cm and back pain emerged during examination. Sacral compression, Gaeslen, Mennel and Patrick-Fabere tests were positive, Schober's test and chest expansion were 1.5 cm both. Occiput to wall distance was 4 cm and chin to chest distance 8 cm.

Erythrocyte sedimentation rate was 34 mm/h (0-15 mm/h). Hemoglobin was 10.2 g/dl(12-18 g/dl) , fibrinogen was 384 mg/dl(180-350 mg/dl). Antinuclear antibodies, CRP and rheumatoid factor were all negative. Roentgenograms of sacroiliac joints revealed bilateral grade 2 sacroiliitis. Sacroiliac jo-

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int computed tomograpy revealed as bilateral narrowing and irregularity of articular surface and subcondral sclerosis (Figure 1). Decrease in lordosis and squaring of the vertebral bodies was found in plain lumbar radiographs (Figure 2). The patient was HLA B27 negative.

With the history of FMF and in the light of our examination suggesting a sacroiliac joint pathology, we considered AS and seronegative spondyloarthropathy (SNSA) in differential diagnosis. The patient was diagnosed as AS associated with FMF.

Discussion

FMF is a multisystemic disease with an autosomal recessive pattern of inheritance primarily affecting persons of Mediterranean origin (1). As there is no specific test for FMF, the diagnosis is based on the following clinical criteria (1, 5). Tel-Hashomer criteria is the most widely used in the recent years. It is contains major ve minor criteria and its sensitivity and specivity are very high (5). The joint symptoms are the second most common presentation of the disease after abdominal pain. It occurs in 70-75 % of the patients. The most common musculoskeletal presentation of FMF is acute recurrent monoarthritis of short duration. Although it is uncommon sacroiliac joints may be also be affected (6). The exact prevalence of SNSA in patients having FMF is still obscure. Spondylarthropaty in FMF patients is determined as unilateral or bilateral sacroiliitis, recurrent enthesitis and inflammatory back or neck pain with minimal radiologic spinal involvement (6, 7). The relationship between FMF and SNSA was investigeted by Langevitz, who studied the features of SNSA in 3000 FMF patients (7). According to our knowledge this study is most extensive one involving FMF patients. Eleven of these patients were accomplishing the criteria for SNSA. In this study authors stated that despite colchicine therapy for FMF, SNSA might still be a presentation. Our patient accomplished European Spondyloarthropathy Study Group criteria for SNSA and 1984 modified New York criteria for AS. This produced some obscurity about final diagnosis. However, the physical findings revealing 1.5 cm of chest expantion, 1.5 cm of lumbar schober's test, extended occiput to wall and chin to chest distances along with squaring of lumbar vertebra, decrease in lumbar lordosis, bilateral grade 2 sacroiliitis in plain films concluded our final diagnosis as AS. Aganist the knowledge of HLA B27 positivity in most of the AS patients she was HLA B27 negative, an insufficient criterion to rule out AS. There are plenty of studies investigating the presence of SNSA in FMF patients (3, 6-8). However the association of AS with FMF was evaluated in relatively few studies (9-11). In the study of Langevitz, apart from eleven patients with SNSA three patients who had had bilateral sacroiliitis, bamboospine and HLA B27 positivity were diagnosed as FMF together with AS (7). This finding was considered as a coincidence. In another case report an FMF patient of armenian origin was also diagnosed as AS whose HLA B27 positive too (12). In our case, both limited cervical and lumbar spine movements and decrease in chest expantion were noted. These physical findings warned us about a different presentation of AS.

Association of HLA B27 positivity with AS and other diseases causing sacroiliitis is more prevalent than the normal population. But an increase in HLA B27 positivity has not been shown in FMF patients having sacroiliitis (6, 13). Moreover, no established relationship between FMF and HLA B27 has been stated up to date. HLA B27 shows a strong association with AS and its prevalence differs extensively among different ethnic groups (14). This condition may explain HLA B27 negativity in our case.

At present, sacroilitis is considered as a musculoskeletal presentation of FMF. But a question still exists: Is association of FMF and AS just a coincidance or an unexplained common etiology exist for both of them?

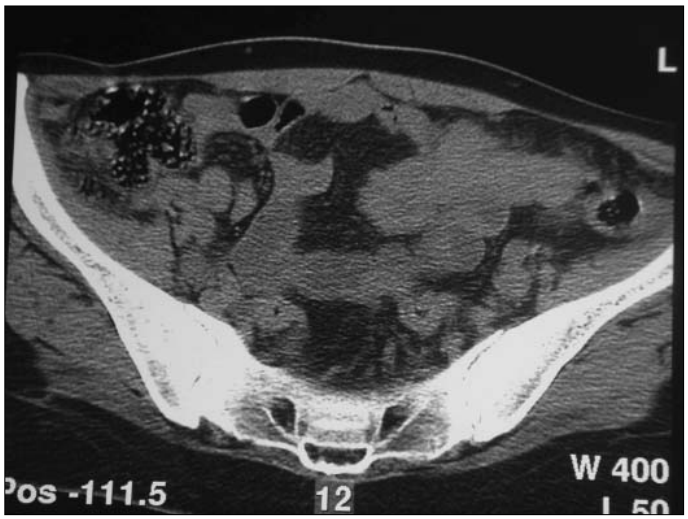

Figure 1. Sacroiliac joint computed tomography of patient : Bilateral narrowing and irregularity of articular surface and subcondral sclerosis

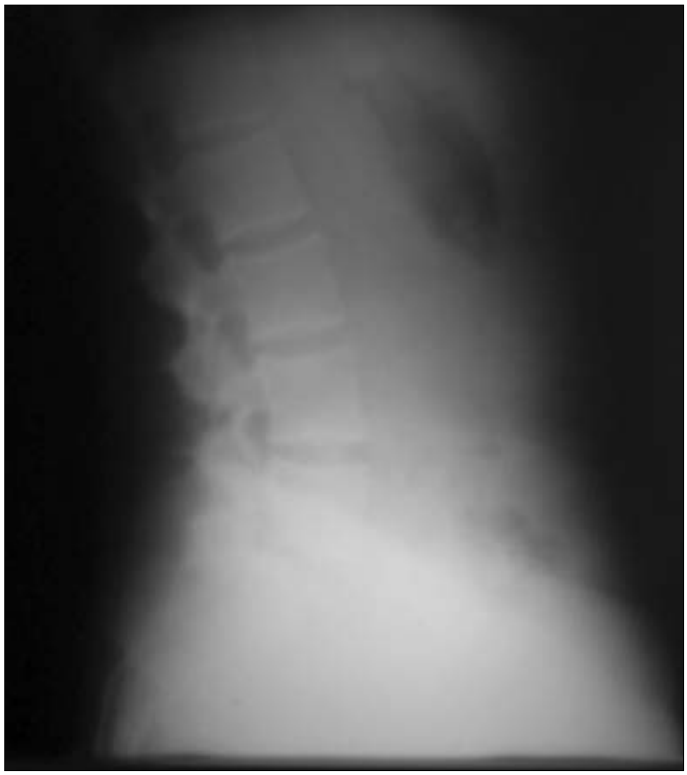

Figure 2. Decrease in lordosis and squaring of the vertebral bodies in the plain lumbar radiographs

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