

## Coexistent with ankylosing spondylitis and familial mediterranean fever

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Familial Mediterranean fever (FMF) is an auto-inflammatory disease characterized by recurrent attacks of fever and serositis. Limited data suggest that the prevalence of sacroiliitis is increased in patients with FMF. Ankylosing spondylitis (AS) is a common inflammatory rheumatic disease characterized by inflammation of the axial joints, peripheral oligoarthritis, and enthesitis. Symptoms related to back pain should be investigated thoroughly in order to diagnose AS and distinguish symptoms related to it from FMF. We presented a 46 year old man who diagnosed FMF and AS.

**Key words:** familial Mediterranean fever, ankylosing spondylitis, inflammation

J Clin Med Kaz 2014; 3(33): 29-31.

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### БЕХТЕРЕВ АУРУЫНЫҢ ЖЕРОРТА ТЕҢІЗІ ОТБАСЫЛЫҚ ҚЫЗБАСЫМЕН ҚОСАРЛАСЫП КЕЗДЕСУІ

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Жерорта теңізі отбасылық қызбасы жедел дамиды қабынумен сипатталатын ауру. Бұл қызбаға дене қызуының қайталанбалы ұстамалары мен серозит тән. Шектелген мәліметтердің нәтижесі бойынша, Жерорта теңізі отбасылық қызбасы бар науқастарда сакроилеиттің таралуы жиі кездеседі. Бехтерев ауруы шеткі олигоартрит және энтезитпен көрініс табады, осыған байланысты қабынумен сипатталатын ревматикалық ауру. Арқа тұсында болатын ауру сезіміне шағымдалатын науқастарды Бехтерев ауруын диагноздау үшін мұқият тексеру керек. Алайда, осы тұста Жерорта теңізі отбасылық қызбасына тән симптомдарды да есепке алудың маңызы зор. Біз Жерорта теңізі отбасылық қызбасы мен Бехтерев ауруы диагноздары қойылған 46-жастағы ер адамның клиникалық жағдайын ұсынып отырмыз.

**Маңызды сөздер:** Жерорта теңізі отбасылық қызбасы, Бехтерев ауруы, қабыну.

### СЛУЧАЙ СОЧЕТАНИЯ БОЛЕЗНИ БЕХТЕРЕВА С СЕМЕЙНОЙ СРЕДИЗЕМНОМОРСКОЙ ЛИХОРАДКОЙ

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Семейная средиземноморская лихорадка является воспалительным заболеванием, которая возникает внезапно. Для семейной средиземноморской лихорадки характерны периодически повторяющиеся приступы лихорадки и серозит. Ограниченные данные свидетельствуют, что распространенность сакроилеита увеличивается у пациентов с семейной средиземноморской лихорадкой. Болезнь Бехтерева - это ревматическая болезнь, которая характеризуется воспалением осевых суставов с периферическим олигоартритом и энтезитом. Пациенты с симптомами, связанными с болью в спине должны быть тщательно дообследованы для диагностики болезни Бехтерева. Здесь также важно отличить симптомы, вызванные семейной средиземноморской лихорадкой. Мы представили случай 46-летнего мужчины, которому выставлен диагноз семейная средиземноморская лихорадка и болезнь Бехтерева.

**Ключевые слова:** семейная средиземноморская лихорадка, болезнь Бехтерева, воспаление.

## INTRODUCTION

Complaints of joint disease is seen in 70%–75% of Familial Mediterranean fever (FMF) patients. These joints are usually lower limbs such as hips or knees. Insomuch that arthritis may be the first sign in one-third of FMF patients. The arthritis of FMF usually presents as recurrent and self limited acute attacks. Protracted arthritis is seen only 5% of patients. Increased risk of sacroiliitis in FMF patients is controversial.

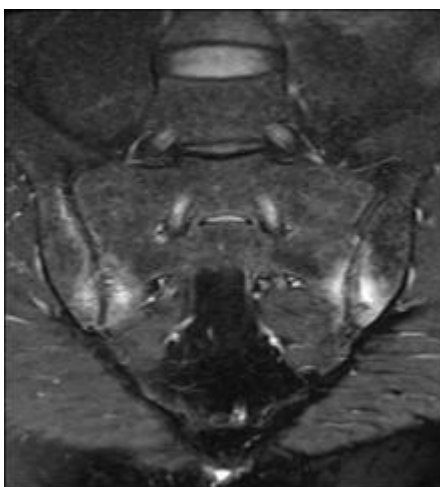
FMF is an autosomal-recessive disease generally seen in Jewish, Armenian, Turkish, and Middle Eastern Arab

populations. The disease is caused by MEFV gene mutation which is located on the short arm of chromosome 16 and encodes an immunoregulatory protein known as pyrin or marenostin. This gene may change the inflammatory response leading to inflammatory diseases. Thus it may contribute to the development of AS. The M694V polymorphism of the MEFV gene is more likely to be associated with arthritis and may be with sacroiliitis [1]. We presented a 46 year old man who diagnosed FMF and ankylosing spondylitis (AS) simultaneously.

## CASE

46 year old man who is under colchicine threatment for 20 years due to FMF, was also having complaints related to backache, showing characteristic of inflamatory diseases in last 3 years. Patient was also suffering from peripheral

joint swelling. Regarding the inflamatory characterised backache sacroiliac magnetic rezonans imaging (MRI) planned. Findings related bilateral sacroiliitis were seen in MRI results (figure 1).



Patient is found HLA B27 positive in adition to heterozygous M694V positive results. In the light of findings patient was considered as having both FMF and

AS. Indomethacin and sulfasalazine were added to ongoing colchicine regime.

## DISCUSSION

FMF is an autosomal-recessive disease characterised by typical acute attacks of serositis, arthritis or skin rash. Joint disease occurs in approximately 75% of patients and generally resolves within a few days without damage. Spondyloarthritis (SpA), usually associated with a lack of HLA-B27, is seen rarely in FMF patients. In FMF patients, SpA usually involves unilateral and bilateral sacroiliitis with inflammatory low back pain and recurrent enthesitis

[2]. Some cases with coincidence of AS and FMF who had a positive HLA-B27 were presented as case reports in the literature. They were diagnosed as coincidence of FMF and AS [3]. As the frequency of M694V is higher among FMF patients with radiographic sacroiliitis than those without this disorder, MEFV gene variations may be the potential pathogenic link between FMF and AS.

## CONCLUSION

The exact relationship between FMF and AS remains obscure. However, SpA was proposed as one of the causes of the possible joint involvement in FMF and the pathogenetic relationship between these two conditions remains unknown. Recent data suggest that pro-inflammatory cytokines including IL-10, IL-12, IL-17, IL-18 contribute to the cytokine network in the inflammatory cascade of FMF and indicates a relationship between chemokines and

subclinical inflammation. A sustained inflammatory reaction is observed in the disease course, and cytokine levels such as IL-1, IL-6 and TNF-alpha have been shown to increase during and between the attacks. As these cytokines are also known to contribute to the pathogenesis of AS, a possible connection between these two diseases can be considered to exist. There is no clear data on the relationship between the attacks of peripheral arthritis and SpA, and the cytokine

network, as well as the gene expression profile in patients with FMF.

We believe that due to highened risk of sacroiliitis

among FMF patients, ankylosing spondylitis should not be overlooked among FMF patients and inflamatory back pain should be investigated thoroughly.

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